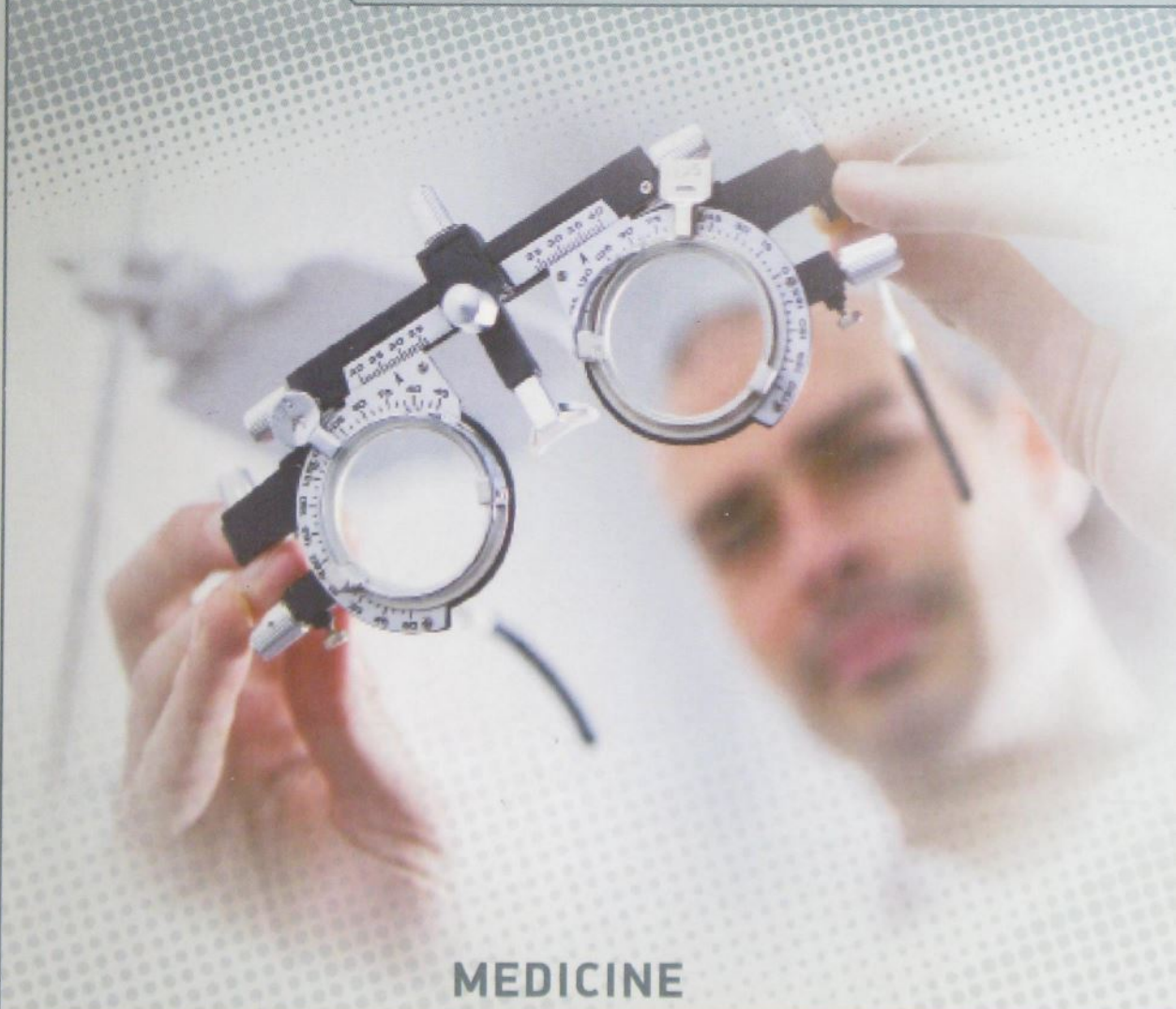
OPHTHALMOLOGY

Edited by Professor **O.P. VITOVSKA**

SECOND EDITION



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TEXTBOOK

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APPROVED
by the Ministry of Education and Science of Ukraine as a textbook for students of higher medical educational establishments

PUBLISHED
pursuant to the Order of the Ministry of Health of Ukraine No. 502 as of 22 June 2010 as a national textbook for students of higher medical educational establishments

RECOMMENDED
by the Academic Council of Bogomolets National Medical University as a textbook for students of higher medical educational establishments

Kyiv
AUS Medicine Publishing
2020

UDC 617.7
LBC 56.7ya73
O-91

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Acknowledgments:
We are extremely grateful to our colleagues and associates Iryna V. Deryapa, Volodymyr V. Kireev, Dmytro S. Churiumov, Stanislav G. Saksonov, Pavlo M. Salata, Oleg G. Parkhomenko, Anna A. Chmil, Olena V. Antonenko for the assistance, support, and invaluable suggestions given to us during preparation of this textbook

Ophthalmology : textbook / O.P. Vitovska, P.A. Bezditko, I.M. Bezkorovayna et al. ; O-91 edited by O.P. Vitovska. — 2nd edition. — Kyiv : AUS Medicine Publishing, 2020. — 648 p.
ISBN 978-617-505-829-9

The textbook Ophthalmology is intended to provide medical students, interns, ophthalmology residents, as well as primary care physicians with a general approach to eye exam and treatment of the most common ocular diseases and emergencies.

In connection with the health care reform in Ukraine that includes, inter alia, the adaptation of achievements of modern European and American ophthalmology in Ukrainian practical medicine, the authors included data on the latest world achievements and guidelines for ophthalmic diseases treatment.

The book contains the latest information on the eye anatomy, thorough ophthalmic examination, concepts of diagnosis, and recent advances in the treatment of the most common ocular disorders and emergencies. Most chapters are illustrated with color clinical photographs or illustrations. At the end of each chapter, key points, schemes, control questions, multiple choice tests, and clinical cases are given to help students prepare for practical lessons and future exams. The appendix provides ophthalmic pharmaceuticals sorted by classes.

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CONTENTS

ABBREVIATIONS

Ad-Cs – adenoviral conjunctivitis
AIDS – acquired immune deficiency syndrome
AKC – atopic keratoconjunctivitis
ALT – argon laser trabeculoplasty
AMD – age-related macular degeneration
ART – antiretroviral therapy
BAK – benzalconium chloride
CDCR – conjunctivodacryocystorhinostomy
CIC – conjunctival impression cytology
CMV – cytomegalovirus
CNS – central nervous system
CPC – cyclophotocoagulation
CRA – central retinal artery
CRAO – central retinal artery occlusion
CRV – central retinal vein
CRVO – central retinal vein occlusion
D – diopter
DCR – dacryocystorhinostomy
DCT – dacryocystectomy
DES – dry eye syndrome
DM – diabetes mellitus
DNS – deviated nasal septum
DR – diabetic retinopathy
EBMD – epithelial basement membrane dystrophy
ECCA – extracapsular cataract extraction
EFA – essential fatty acids
GPC – giant papillary conjunctivitis
HIV – human immunodeficiency virus
HSV – herpes simplex virus
ICCE – intracapsular cataract extraction
i/m – intramuscular
IOFB – intraocular foreign body
IOL – intraocular lens
IOP – intraocular pressure
IR – infrared
KCS – keratoconjunctivitis sicca
KPs – keratic precipitates
MS – multiple sclerosis
NPDR – non-proliferative diabetic retinopathy
NLD – nasolacrimal duct
NO – nitric oxide
NPDS – non-penetrating deep sclerectomy
NSAIDs – non-steroidal anti-inflammatory drugs
OD – right eye
ONA – optic nerve atrophy
OS – left eye
OTCS – ocular trauma classification system
OU – both eyes
PAC – perennial allergic conjunctivitis
p/b – parabulbar
PDR – proliferative diabetic retinopathy
PHA – persistent hyaloid artery
PHPV – persistent hyperplastic primary vitreous
PK – penetrating keratoplasty
PMMA – polymethylmethacrylate
POAG – primary open-angle glaucoma
PTK – phototherapeutic keratectomy
PVD – posterior vitreous detachment
r/b – retrobulbar
RGP contact lenses – rigid gas permeable contact lenses
RP – retinitis pigmentosa
RPE – retinal pigment epithelium
SAC – seasonal allergic conjunctivitis
SLT – selective laser trabeculoplasty
SPK – superficial punctate keratitis
SRAT – slow releasing artificial tear
TB – tuberculosis
TBUT – tear film break-up time
UV – ultraviolet
VA – visual acuity
VKC – vernal keratoconjunctivitis
WHO – World Health Organization

CHAPTER 1

Anatomy of the Eye and Visual Functions

OBJECTIVES

To know:
— the structural features of the eye — the globe and ocular adnexa;
— the structure, basic properties, and functions of each part of the eye;
— the physiology of vision;
— the main visual functions.

Plan:

1. **THE ORBIT AND OCULAR ADNEXA**
   - **1.1. The Orbit and Its Content**
   - **1.2. The Eyelids**
   - **1.3. The Lacrimal System**
   - **1.4. The Conjunctiva**
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     - The Iris
     - The Ciliary Body
     - The Choroid
   - **2.3. The Inner Nervous Layer**
     - The Retina
   - **2.4. The Optic Nerve**
3. **THE INTERIOR OF THE EYEBALL**
   - **3.1. The Aqueous Humor**
   - **3.2. The Lens**
   - **3.3. The Vitreous**
4. **SEGMENTS AND CHAMBERS OF THE EYEBALL**

Content:

The eye is the organ of sight or vision. Vision is by far the most used of the five senses and is one of the primary means that we use to gather information from our surroundings. More than 75 % of the information we receive about the world around us consists of visual information.

The organ of vision consists of the eyeball, ocular adnexa and supplying structures.

EYE FACTS

We see with our brain, not our eyes. The eyes function like a camera, capturing light and sending data back to the brain.

1. The Orbit and Ocular Adnexa

1.1. The Orbit and Its Content

Definition

The orbit is the bony socket of the skull in which the eye and its adnexa are situated. It is a cone-shaped cavity formed by the union of cranial and facial bones.

Structure

The orbit is formed by seven different bones:

— the *frontal bone* forms the roof of the orbit,
— the *zygomatic bone* forms the strong lateral wall,
— the *maxillary bone* creates the orbital floor,
— the *lacrimal bone* forms the medial wall,
— the *ethmoid bone* forms the medial wall. The thinnest area in the orbit is a part of the ethmoid bone called the lamina papyracea,
— the greater and lesser wings of the *sphenoid bone* forms the back of the orbit,
— the *palatine bone* is situated at the back part of the nasal cavity between the maxilla and the pterygoid process of the sphenoid and forms the floor of the orbit.

The orbital apex is the posterior end of the orbit and the entry point for the nerves and vessels supplying the eye. Here the four orbital walls converge. It has three openings, the *optic canal* which transmits optic nerve and ophthalmic artery, the *superior* and *inferior orbital fissures* which transmits a number of nerves, arteries and veins.

Properties

The volume of the orbit is approximately 30 cm^3 with an entrance dimension of 35 mm high and 45 mm wide. Behind the orbital margin is the maximum size of 1 cm. Orbital depth in the adult orbit is 40—45 mm from entrance to exit. The eyeball itself occupies about one fifth of the space of the orbit. In addition to the eyeball itself, the orbit contains the lacrimal gland, muscles, blood vessels, nerves, and fatty tissue. This fatty tissue serves as a protective cushion for the eye. The rim of the orbit protects the globe from impact with large objects.

Functions

The main functions of the orbit are:

— to provide protection and support for the globe together with the optic nerve, ocular muscles, nerves, blood vessels, and lacrimal gland;
— to provide attachment for the muscles that control eyeball movement.

1.2. The Eyelids

Definition

The eyelids are two movable protective muscular folds of skin that cover the front of the eyeball when closed. The upper lid ends at the eyebrows and is more moveable of the two; the lower lid merges into the cheek. There are short curved hairs, the eyelashes, situated on their free edges.

EYE FACTS

When you blink, you shut your eyes for about 0.3 sec. That's a total of 30 min each day!

Structure

The eyelids consist of the following layers starting anteriorly:

— the skin;
— the layer of subcutaneous connective tissue;
— the layer of striated muscle fibres of the orbicularis muscle and levator palpebrae superioris muscle;
— the layer of submuscular connective tissue (the nerve and vessels lie in this layer);
— the fibrous layer, including the tarsal plate and meibomian glands laying in the substance of the tarsal plates;
— the thin connective tissue — septum orbitale or palpebral fascia;
— the layer of smooth or non-striated fibres — palpebral Müller's muscle;
— the palpebral conjunctiva.

Properties

When the eye is open, the upper lid covers about 1/6 of the cornea and the lower lid just touches the limbus. The space between the two open lids is called the *palpebral fissure*. It measures about 8—11 mm vertically in the center and about 28—30 mm horizontally. The two lids join each other at the medial and lateral angles (or outer and inner canthi). The medial canthus is about 2 mm higher than the lateral canthus (fig. 1.1). The lid margin is about 2-mm broad and is divided into two parts by the punctum. On the upper eyelid, approximately 150 eyelashes are arranged in three or four rows; on the lower eyelid there are about 75 eyelashes in two rows.

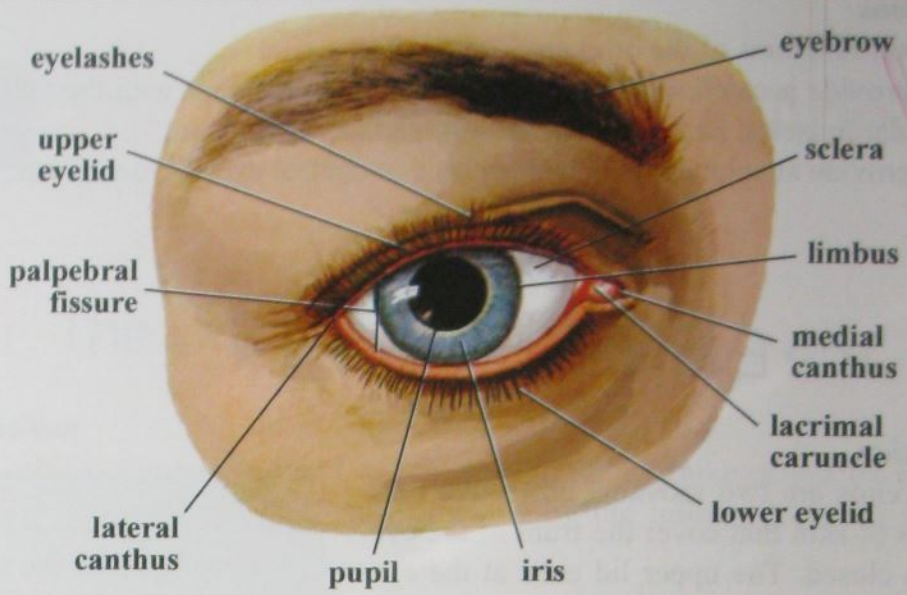

Fig. 1.1. Palpebral Fissure and External Eye Anatomy (right eye)*

The *eyelid skin* is the thinnest, more elastic and mobile than the skin elsewhere in the body. It is well vascularized and has no subcutaneous fat. Just below the skin there is a layer of loose subcutaneous tissue which can easily get filled up with blood or fluid resulting in a swollen eyelid after an injury.

The *tarsal plate* is the main supporting structure of the lids, it gives stiffness to the eyelids and helps maintain their contour. This tarsal plate is also a place for muscles to attach. Its length is 25 mm, the thickness 1 mm, and the maximum (central) height is 10 mm (upper tarsus) and 4 mm (lower tarsus).

The *meibomian glands* are long, thin and run parallel to each other, perpendicular to the eyelid margin, and are located in the tarsal plate of the eyelids. These glands secrete oil into the tear film that keeps the tears from evaporating too quickly.

There are several sets of *smaller glands* situated within the structure of the eyelids. These glands are: the glands of Zeis — sebaceous glands that open into the follicles of the eyelashes, the glands of Moll — modified sweat glands that also open into the eyelash follicles, the glands of Wolfring — these are accessory lacrimal (tear) glands.

The inner surface of the eyelids is lined with the palpebral conjunctiva.

Two *muscles* are responsible for eyelid movement. The *orbicularis oculi*, which forms an oval sheet across the eyelids, closes the eyelids. The *levator palpebrae*, which arises from the apex of the orbit and is inserted by three parts on the skin of the lid, anterior surface of the tarsal plate and conjunctiva of the superior fornix. It elevates the upper lid.

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* All figures used in the Chapter 1 are taken from *Анатомія* людини: у 3 т. Т. 2 / А.С. Головацький, В.Г. Черкасов, М.Р. Сапін та ін. — Вид. 4. — Вінниця: Нова Книга, 2009. — С. 371—397.

Functions

The eyelids' main functions are:

— to protect the anterior eyeball from mechanical trauma, extremes of temperature and excessive light that might damage the eye;
— to secret the oily part of the tear film;
— to spread tears over the ocular surface by blinking so that tears lubricate the cornea and conjunctiva thus protecting them from dehydration;
— to contain the puncta through which tears drain into the lacrimal drainage system;
— the eyelashes help filter out foreign matter, including dust and debris, and prevent these from getting into the eye.

1.3. The Lacrimal System

Definition

The lacrimal system or apparatus contains structures for tear production and drainage. It consists of the lacrimal glands and the drainage system.

Structure

The lacrimal system consists of:

— *the lacrimal gland and its ducts:*
— the upper orbital part;
— the lower palpebral part;
— *the accessory lacrimal glands:*
— the Glands of Krause;
— the Glands of Wolfring;
— *the lacrimal drainage system:*
— the upper and lower lacrimal puncta;
— the lacrimal canaliculi;
— the lacrimal sac;
— the nasolacrimal duct.

The *Tear Film* consists of three layers from anterior to posterior:

1. The lipid layer.
2. The aqueous layer.
3. The mucus layer.

Properties

The *lacrimal gland* is located in a shallow depression within the orbital part of the frontal bone, so it lies above the outer corner of the eye. It consists of an upper orbital and a lower palpebral parts. The orbital part is larger, about the size and shape of a

small almond. The palpebral part is small and consists of only one or two lobules. It is visible when one everts the upper lid and has a patient look inward and downward. The lacrimal gland has ducts that open into the palpebral conjunctiva. The lacrimal gland itself is really only responsible for reflexive tearing. It produces the tears that flow nasally across the eye and enter into the drainage system.

The *accessory lacrimal glands.* The glands of Krause are microscopic glands lying beneath the palpebral conjunctiva between the fornix and the edge of tarsus. These are about 42 in the upper fornix and 6—8 in the lower fornix. The glands of Wolfring are present near the upper border of the superior tarsal plate and along the lower border of the inferior tarsus. They are responsible for basal tearing. They account for about 10 % of the total lacrimal secretory mass.

The *drainage system* includes the conjunctival lake where tears collect, the nasal angle which forms the lacrimal lake, the superior and inferior (upper and lower) puncti into which tears drain. The lacrimal punctum is about 0.3 mm in diameter. The tears then flow into the superior and inferior canaliculi, the common canaliculus, the lacrimal sac and finally drain into the nasolacrimal duct into the nose (fig. 1.2). Tear drainage is an active process. Each blink of the lids helps to pump tears through the system.

Tears are composed of water, sodium chloride and a mildly antibacterial enzyme. It contains electrolytes, proteins, immunoglobulins, glucose and dissolved oxygen (from the atmosphere). Tear flow originates in both the lacrimal gland and accessory lacrimal glands at a rate of approximately 1 microliter per minute. Tears have a pH of 7.6 and an osmolarity similar to sodium chloride at 0.9 percent. Precorneal tear film lies just anterior to the globe, is 6—10 microns thick, and consists of three layers: the outer lipid layer, the middle aqueous layer, and the inner mucus layer.

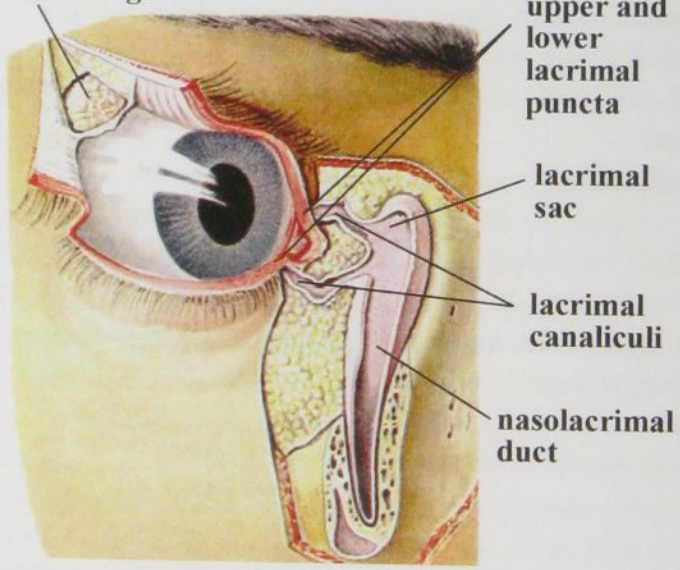


Fig. 1.2. Lacrimal Apparatus of the Eye (anterior view of the right eye)

The outer *lipid layer* (approximately 0.1 μm thick) secreted by the Meibomian glands, prevents evaporation of the tear film and lubricates the eyelid.

The middle *aqueous layer* (approximately 8 μm thick) constitutes 90 % of the thickness of the tear film and provides oxygen and nutrition for the cornea. This layer also has antibacterial properties and washes away debris. It is secreted by 2 kinds of lacrimal glands: the main lacrimal gland and the accessory lacrimal glands of Krause and Wolfring.

The inner *mucus layer* (approximately 0.8 μm thick) is secreted by goblet cells distributed throughout the bulbar and palpebral conjunctiva. This hydrated glycoprotein layer makes the corneal surface hydrophilic and decreases surface tension of the tear film.

Functions

Functions of the tear film are:

— keep the surface of the eye and conjunctiva moistened and lubricated;
— provides oxygen and nutrition to the corneal epithelium;
— washes away debris and noxious irritants;
— prevents infection due to presence of antibacterial substances;
— facilitates movements of the lids over the globe;
— provides a smooth refracting surface over the cornea.

1.4. The Conjunctiva

Definition

The conjunctiva is the thin transparent vascular mucous membrane that covers the inner surface of the eyelids and anterior ocular surface except the cornea. It starts at the limbus, covers the sclera, and lines the inner surface of the eyelids. When the lids are closed it forms the conjunctival sac together with the surface of the cornea. The conjunctiva is normally of shiny appearance.

Structure

The conjunctiva is typically divided into three parts:

1. *The palpebral or tarsal* conjunctiva — the conjunctiva that lines the eyelids.
2. *The fornix or transitional conjunctiva* — the conjunctiva where the inner part of the eyelids and the eyeball meet, the palpebral conjunctiva is reflected at the superior fornix and the inferior fornix to become the bulbar conjunctiva.
3. *The bulbar or ocular conjunctiva* — the conjunctiva covering the eyeball over the sclera. This part of the conjunctiva is bound tightly and moves with the eyeball movements.

Properties

The name *conjunctiva* (to conjoin — to join) has been given to this mucous membrane owing to the fact that it joins the eyeball to the lids.

Histologically, the conjunctiva consists of three layers, namely: epithelium, adenoid layer, and fibrous layer. The conjunctiva consists of stratified nonkeratinizing epithelium.

The conjunctiva contains two types of mucin producing *glands* that secrete mucus, which is essential for wetting the cornea and conjunctiva and accessory lacrimal glands (the glands of Krause and the glands of Wolfring).

The bulbar conjunctiva medially forms the caruncle. The *caruncle* is a small, ovoid, pinkish mass, situated in the inner canthus. It is a piece of modified skin and so is covered with stratified squamous epithelium and contains sweat glands, sebaceous glands, and hair follicles.

Functions

The conjunctiva:

— assists in lubricating the eye by producing mucus and tears along with the lacrimal system;
— prevents the entry of foreign bodies, microorganisms or other noxious agents into the eye and orbit;
— enables smooth movement of eyelids on the eye surface.

1.5. The Extraocular Muscles

Definition

The extraocular muscles are muscles that control eye movements.

Structure

There are six *extraocular muscles* that control the movement of each eyeball (fig. 1.3):

— four *rectus muscles* (which are named according to their insertion into the sclera):
— superior;
— inferior;
— medial;
— lateral;
— two *oblique muscles*:
— superior;
— inferior.

EYE FACTS

The extraocular muscles that move the eyes are the strongest muscles in the human body for the job that they have to do. They are 100 times stronger than they need to be.

Properties

Four rectus muscles attach to the eyeball anterior to the equator and each pulls the eye in the direction of their attachment:

— the *medial rectus muscle*, the strongest extraocular muscle, rotates the eyeball inward toward the nose (adduction);
— the *lateral rectus muscle* rotates the eyeball outward toward the temple (abduction);
— the *superior rectus muscle* rotates the eyeball upwards (elevation) and assists in intorsion;
— the *inferior rectus muscle* rotates the eyeball downwards (depression) and assists in extorsion.

The oblique muscles attach the globe posterior to the equator. The insertion of the muscles determines the direction of their pull:

— the *superior oblique muscle*, the longest and thinnest eye muscle, rotates the eyeball downward and inward directions (intorsion).

— the *inferior oblique muscle*, shortest of the extraocular muscles, rotates the eyeball upward and outward directions (extorsion).

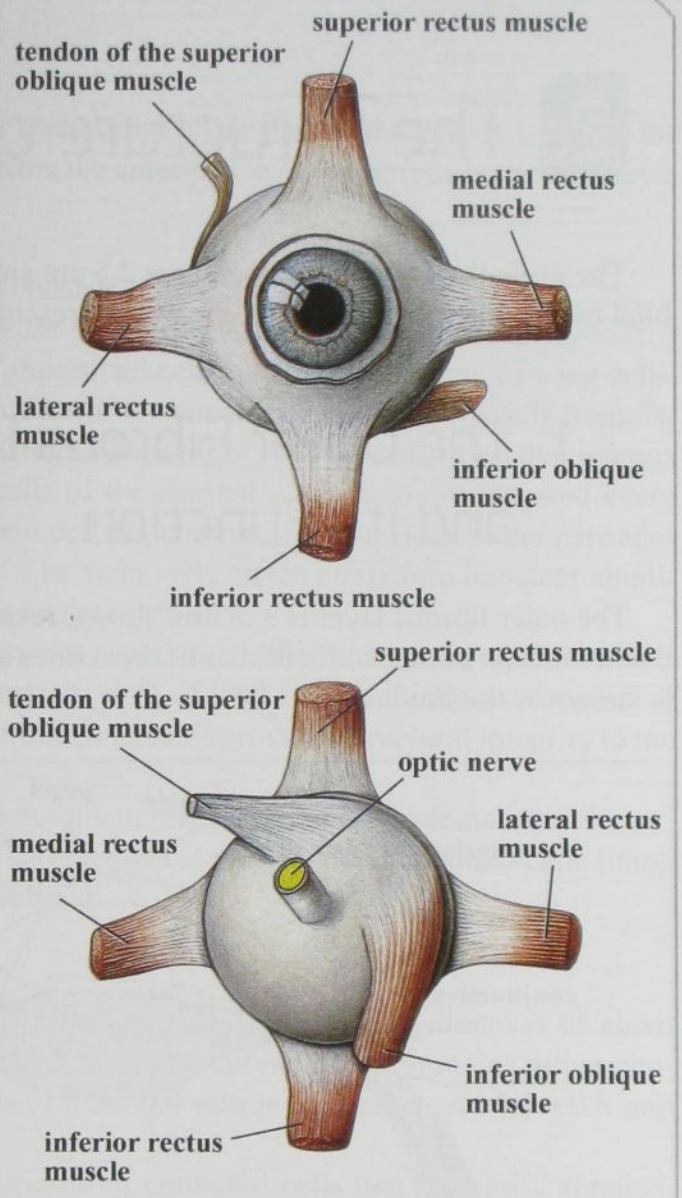


Fig. 1.3. Extraocular Muscles (anterior and posterior view of the right eye)

Movement of the eyes is complex because in order to look in a specific direction the muscles of one eye must coordinate with the other. For example in order to look up and to the right, the lateral rectus and superior rectus of the right eye must coordinate with the medial rectus and superior rectus of the left eye.

All the extraocular muscles except the inferior oblique originate from a fibrous ring around the optic nerve (annulus of Zinn) at the orbital apex. The muscles fan out towards the eye to form a "muscle cone". The optic nerve, the ophthalmic blood vessels, and the nerves to the extraocular muscles (except the fourth cranial nerve) are contained within the muscle cone.

The connective tissue between the individual ocular muscles is incorporated into the fascial sheath of the eyeball (Tenon's capsule).

Functions

The primary function of the extraocular muscles is to control the eyeball movements from left to right and up and down, inward and outward or even around in circles when one wishes.

2. The Structure of the Eye

The eyeball weighs 7.5 g, measures 2.5 cm and rests on a pad of fat lining the orbital cavity. Internal anatomy of the eye is presented on the fig. 1.4.

2.1. The Outer Fibrous Layer and Its Function

The outer fibrous layer is a dense, strong, resistant coat or capsule which protects the intraocular contents. It consists of the cornea and the sclera, the junction of which is known as the limbus.

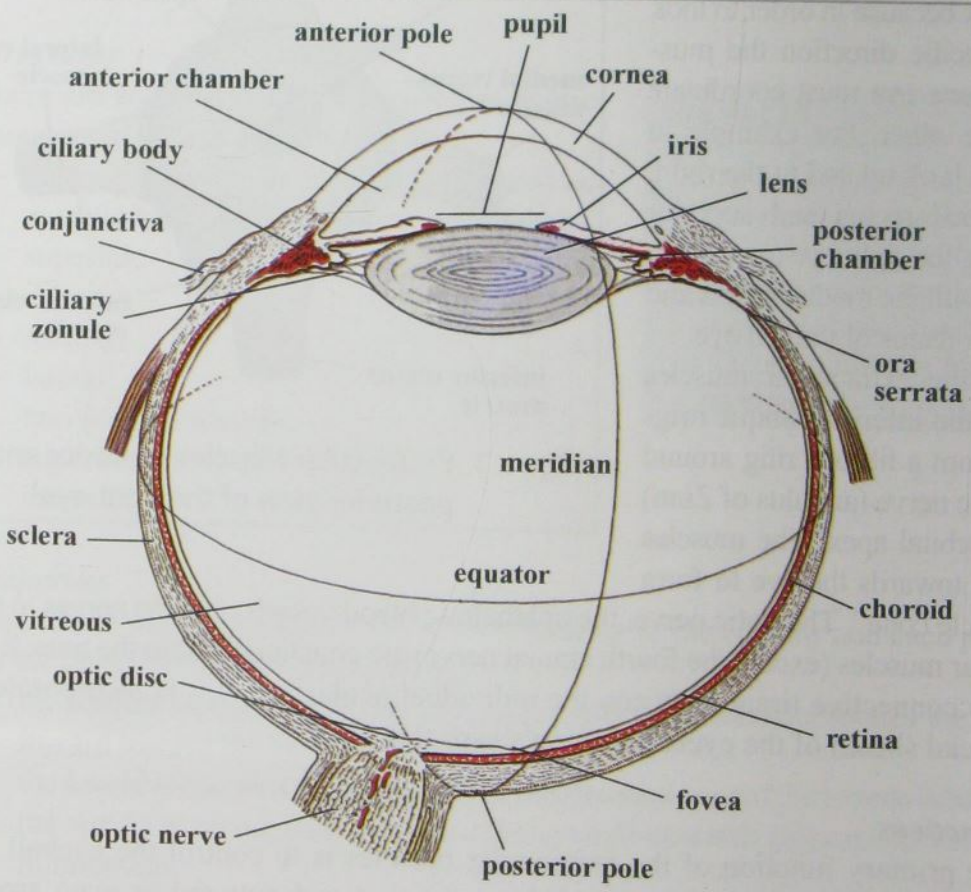


Fig. 1.4. Internal Anatomy of the Eye (sagittal section)

The Cornea

Definition

The cornea is the most anterior transparent, clear part of the eye. It is called the "optical window" into the eye. It forms the anterior 1/6 of the fibrous layer of the eye.

Structure

On the cross section, the cornea consists of five layers:

1. *The epithelium and its basement membrane* — the most superficial layer. It is composed of two to three layers of superficial cells, two to three layers of wing cells, and one layer of basal cells. The surface of the superficial epithelial cells is irregular due to the presence of microplicae (ridge-like folds of the plasmalemma) that interact with the overlying tear film. The cells of the corneal epithelium are renewed every 7—10 days from the pluripotent stem cell population, which resides in the palisades of Vogt at the corneoscleral limbus. The stem cells differentiate into transient amplifying cells when they migrate to the central cornea.
2. *Bowman's layer* — a homogeneous sheet of modified stroma, an acellular zone.
3. *The stroma* forms of approximately 90 % of total corneal thickness. Consists of lamellae of collagen, cells and ground substance. It provides structural integrity to the cornea.
4. *Descemet's membrane* — the basement membrane of the endothelium.
5. *The endothelium* — a single layer of five- to seven-sided cuboidal cells lining the inner surface of Descemet's membrane.

Properties

The cornea of an adult human eye has an average horizontal diameter of about 11.5 mm and a vertical diameter of 10.5 mm, and curvature that remains rather constant throughout life. Its thickness is of 0.5—0.6 mm in the center and 0.6—0.8 mm at the periphery.

The outside surface layer is composed of epithelial cells that are easily abraded. Though epithelial injuries are painful, this layer heals quickly and typically does not scar. If the stroma is damaged, this can lead to scar formation.

The inner endothelium works as a pump to keep the cornea dehydrated. If the endothelium becomes damaged (during surgery or by degenerative diseases), aqueous fluid can flow unhindered into the stroma and cloud up the cornea with edema. Endothelial cell count is very important as these cells don't regenerate when destroyed. If the cell count gets too low, the endothelial pump can't keep up and the cornea swells with water, possibly necessitating a corneal transplant to regain vision.

The cornea is avascular and gets its nutrition from tears on the outside, aqueous fluid on the inside, and from blood vessels of the limbus.

The cornea is the most densely innervated tissue of the body. For comparison, the cornea has 300—600 times the sensory innervation density of the skin. This dense innervation is necessary since the cornea is the first line of defence against injury to the eye.

The cornea is:
— transparent;
— smooth and regular surface;
— reflective;
— avascular;
— highly sensitive.

Functions

The functions of the cornea are:
— transmission of light;
— the main refracting surface of the eye (2/3 of the total refractive power; the dioptric power is +40 to +45 D), focuses the light onto the retina;
— support and protective function.

The Sclera

Definition

The sclera is the opaque, white fibrous outer protective coat of the eye. It almost completely surrounds the eye except on the anterior surface of the eye ball. It is about 1 mm thick. A few strands of scleral tissue pass across the anterior portion of the optic nerve as the lamina cribrosa.

Structure

The sclera consists of collagen fibrils maintained by fibroblasts that run parallel to the surface of the eye.

The sclera can be subdivided into 3 layers:

1. *episclera* — external layer, loose connective tissue adjacent to the periorbital fat, which contains blood vessels to nourish the sclera;
2. *sclera proper*, also called Tenon's capsule — the dense investing fascia of the eye composed of dense type I and III collagen; avascular;
3. *lamina fusca* — the inner layer of the sclera, located adjacent to the choroid and containing thinner collagen fibres and pigment cells.

Properties

The sclera forms the posterior opaque five-sixths part of the external fibrous tunic of the eyeball. It is the thickest posteriorly and the thinnest beneath the insertions of the rectus muscles. The thickness is 1 mm posteriorly near the optic nerve and 0.3 mm anteriorly.

Functions

The functions of the sclera are:
— forming — maintains the shape of the eye;
— protective — being the support to the inner eye coats, acts as a skeleton to the eye;
— the place of the extraocular muscles attachment.

The Limbus

The limbus is the border zone, about 1.5 mm wide, between the transparent cornea and opaque sclera where the conjunctival epithelium joins with the corneal. There is a minute arcade of blood vessels about 1 mm broad present at the limbus.

2.2. The Middle Vascular Layer

The middle layer of the eye, the uvea, is highly vascular and supplies nutrition to the various structures of the eyeball. It consists of three parts — iris, ciliary body and choroid. However, the entire uveal tract is developmentally, structurally and functionally one indivisible structure.

The Iris

Definition

The iris is the most anterior part of the uvea, shaped like a disc with a central opening — the pupil, which rests on the anterior surface of the lens.

Structure

The iris consists of four layers and two muscles:

1) *the endothelium* — a single layer of flat endothelial cells continuous with the corneal endothelium;
2) *the stroma* — a connective fibrovascular loose tissue rich in melanocytes. Posteriorly it contains two muscles:
   — *the sphincter muscle* — circularly arranged, encircling the pupillary margin causes constriction of the pupil under parasympathetic stimulation;
   — *the dilator muscle* — less distinct muscle fibres which run radially around the pupil and cause dilation in response to sympathetic stimulation.
3) *the basement membrane* — strong connective tissue, which forms the support and innermost layer of the iris;
4) *the pigment epithelium* — two-layered cuboidal posterior cells continuous with that of the ciliary body. This pigment layer serves to prevent the penetration of light through the iris into the inner dark chamber of the eyeball.

The iris is divided into *two zones*:

— *the pupillary zone* — the inner part of the iris that forms the pupil boundary;
— *the ciliary zone* — the remaining part of the iris that extends into the ciliary body.

Properties

The iris is the colored part of the eye. The color of the iris is determined by a dark pigment called melanin. The more melanin present, the darker the iris, whereas fewer pigment cells give rise to green or blue eyes.

EYE FACTS

Some people are born with two differently colored eyes. This condition is heterochromia.

The iris has rings of two muscle fibres that regulate the size of the pupil. When they contract, it causes the pupil to constrict (become smaller). This occurs in bright light. A second set of muscle fibres radiate outward from the pupil. When these muscles contract, the pupil dilates (becomes larger). This occurs under reduced illumination or in darkness. The pupil size can change from 2 mm to 8 mm. This means that by changing the size of the pupil, the eye can change the amount of light that enters it.

Functions

The primary function of the iris is to regulate the amount of light that enters the eye.

The Ciliary Body

Definition

The ciliary body is a part of the uvea between the iris and the choroid.

Structure

The ciliary body consists of three zones. They are:

— the *ciliary ring* — here the ciliary body is firmly attached to the choroid and retina (ora serrata);
— the *ciliary processes* — which attach to the lens;
— the *ciliary muscles* — which control the curvature of the lens.

Properties

The ciliary body is a ring-like part of the uvea, triangular in section. It contains the ciliary muscle, vessels and fibrous connective tissue. The folds on the inner epithelium are called ciliary processes.

The ciliary processes secrete the aqueous fluid which fills the posterior and anterior chambers and provides nutrition for avascular tissues in the eye such as the cornea.

The ciliary body is attached to the lens by connective tissue called the zonular fibres (fibres of Zinn). Relaxation of the ciliary muscle puts tension on these fibres and changes the shape of the lens in order to focus light on the retina.

Functions

The main functions of the ciliary body are:

— accommodation (focusing on near and distant objects);

— aqueous humor production

— supporting the lens through its suspensory ligaments (zonular fibres).

The Choroid

Definition

The choroid is the posterior part of the uvea. It extends from the optic disc to the ora serrata. Its inner surface is smooth, brown and lies in contact with the pigment epithelium of the retina. The outer surface is rough and lies in contact with the sclera.

Structure

The choroid consists of three layers:

1. the *layer of large vessels* or the stroma contains the larger arteries and veins of the choroid and the ciliary nerves. A varying number of melanocytes are also present;
2. the *layer of small vessels* or the capillary network of the choroid. It is the densest capillary network of the body. When usually individual red blood cells must squeeze through capillaries, in the choroid several erythrocytes fit side by side. Therefore, they have time to release only a part of the oxygen they transport. The capillaries of the choroid are fenestrated;
3. *Bruch's membrane* is the outermost layer which is the basement membrane of the capillaries of the choroid and that of the pigment epithelium of the retina.

Properties

The human choroid is the thickest at the far extreme rear of the eye (0.2 mm), while in the outlying areas it narrows to 0.1 mm.

The choroid is attached to the sclera only around the optic nerve and where blood vessels and nerves pass through the sclera. Otherwise the choroid — like the ciliary body — is only apposed to the sclera. The choroid separates easily from the sclera if exudate or blood gains access between them. The choroid is, however, tightly attached to the pigment epithelium of the retina.

Pigment cells within the choroid containing melanin pigment absorb light after it has passed through the photoreceptor cells so that it will not reflect back into the retina from outer layers so it helps to lessen glare within the eye.

Functions

The main functions of the choroid are:

— to supply nutrients to the retina;

— to support the retina;

— to prevent reflection of light within the eye.

2.3. The Inner Nervous Layer

The inner nervous layer is concerned with visual functions.

The Retina

Definition

The retina is the innermost layer of the eye. It is sensitive tissue that is a thin, transparent, net-like complex membrane. It converts light signals into electrical impulses that are transmitted to the brain, which finally interprets them as visual images. It lines about 3/4 of the eyeball.

Structure

Functionally the retina consists of two parts — light-sensitive (outer or neuro-epithelial) and light-conductive (cerebral) which are composed of three major classes of neurons: 1) neuro-epithelium (rods and cones); 2) bipolar cells and 3) multipolar, ganglion cells (fig. 1.5). Together they make ten layers:

1. the internal limiting membrane;
2. the basement membrane formed by Müller cells;
3. the ganglion cell layer;
4. the inner plexiform layer;

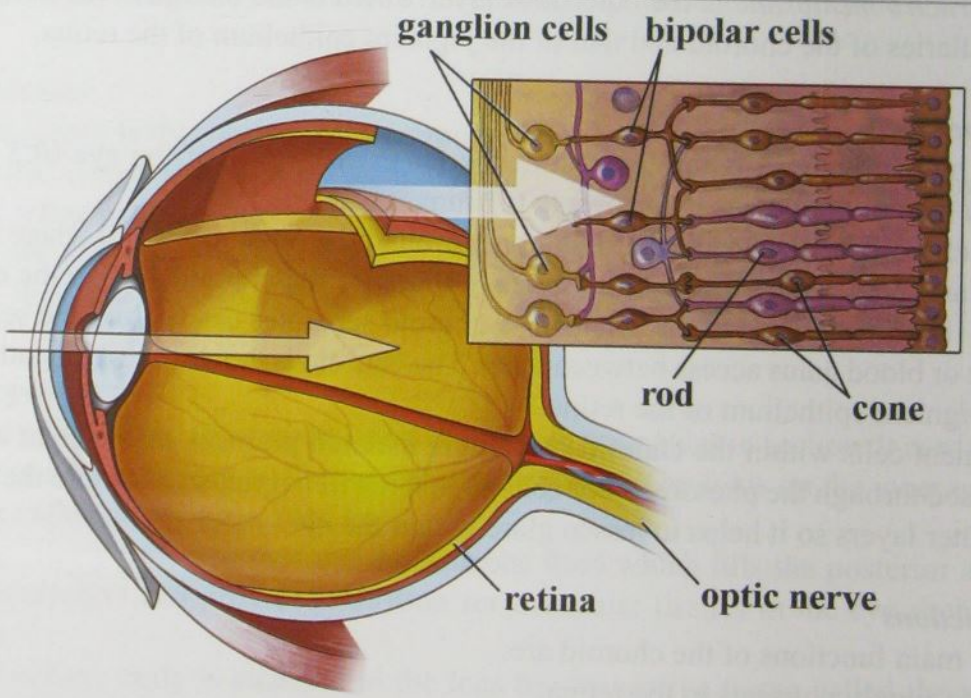


Fig. 1.5. Three major classes of retinal neurons

5. the inner nuclear layer of bipolar, amacrine and horizontal cell bodies;
6. the outer plexiform layer;
7. the outer nuclear layer of photoreceptor cell nuclei;
8. the external limiting membrane;
9. the layer of rods and cones;
10. the retinal pigment epithelium.

Properties

The retina contains two kinds of photoreceptors — cones and rods, which differ in function.

EYE FACTS

Human eyes contain around 107 million light sensitive cells — 7 million cones which help us see color and details, as well as 100 million rods which help us see better in the dark.

The *cones* are sensitive to bright light and color. Pigments of the cones have absorption peaks in the blue, green or yellow parts of the spectrum. The cones are found primarily in the center (or fovea) of the retina.

The *rods* are more sensitive than the cones. They are stimulated by low intensity of light and function with reduced illumination (dark adaptation). The rods are found primarily in the periphery of the retina. Pigments of the rods have an absorption peak in the blue-green part of the spectrum.

In humans, the rods are approximately 20 times more abundant than the cones. The density of the rods and cones varies between different regions of the retina.

The two most significant parts of the retina are central-placed *macula lutea* and the *optic disc* located toward the nose.

The *macula lutea* is also called the yellow spot. It is about 5.5 mm in diameter. Histologically, the macula has several layers of ganglion cells, whereas in the surrounding peripheral retina the ganglion cell layer is only one-cell thick. The depression near the center of the macula is called the *fovea*. It is about 1.5 mm in diameter and is the most sensitive part of the retina. In its center there is a shining pit called *foveola* (0.35 mm in diameter), the thin, avascular bottom of the fovea. It is a point of the sharpest vision. This area of the retina is responsible for central vision and contains the largest concentration of the cones in the eye.

The *optic disc* is also known as the optic nerve head. It is a pink-colored, well-defined circular area of 1.5 mm in diameter. The optic disc is where the optic nerve attaches to the eye. The central retinal artery and vein emerge through the center of this disc. The absence of visual cells in the optic disc causes the appearance of a blind spot in the field of vision.

Functions

The main function of the retina is transducing light into nerve impulses that travel via the optic nerve to the lateral geniculate nucleus of the thalamus and finally to the visual cortex where the sensation of vision is created.

2.4. The Optic Nerve

Definition

The optic nerve is the large bundle of nerves that connects the eye to the brain that transmits the impulses from the retina. It is also known as cranial nerve II.

Structure

The optic nerve can be divided into 4 parts:

1. *intraocular part*;
2. *intraorbital part* extending from the globe to the apex of the orbit;
3. *intracanalicular part* within the optic canal;
4. *intracranial part* that merges into the optic chiasm and then optic tract.

Properties

The optic nerve is the only nerve in the body that we can actually see (using our ophthalmoscope) in vivo. It meets the posterior part of the globe slightly nasally to the posterior pole and slightly above the horizontal meridian. The front surface of the optic nerve, which is visible on the retina, is called the optic disk or optic nerve head. There are no light-sensitive cells on the optic disc — and hence the blind spot that anyone can find in their field of vision.

The trunk of the optic nerve is formed by the axons of the 1.2 million ganglion cells of the retina and they will not regenerate if severed. The nerve fibre layer of the retina is comprised of these axons and they converge to form the optic nerve. The optic nerve is made up of visual fibres (80 %) and afferent pupillary fibres (20 %).

The orbital portion of the nerve travels within the muscle cone to enter the bony optic foramen to gain access to the cranial cavity.

The optic nerve contains within its fibres the central retinal artery and the central retinal vein.

Functions

The optic nerve is responsible for transmitting nerve signals formed by the retina to the brain, which interprets them as images.

3. The Interior of the Eyeball

3.1. The Aqueous Humor

Definition

The aqueous humor (or aqueous fluid) is the clear, colorless watery fluid that fills the front part of the eye, between the cornea and the lens.

Structure

The aqueous humor is an optically clear solution of electrolytes (in water). It normally contains a low concentration of proteins, but a higher concentration of ascorbic acid compared with plasma.

It consists of 80 % of plasma glucose, 1 % of plasma, mostly albumins.

Properties

The aqueous humor is produced by the ciliary processes, then seeps through the pupil into the anterior chamber and leaves the eye through the trabecular meshwork into the canal of Schlemm, the aqueous veins and the conjunctival episcleral veins.

The aqueous humor in the anterior chamber is a component of the optical system of the eye. It has an index of refraction of 1.336, slightly lower than that of the cornea.

The rate of aqueous humor outflow is normally equal to the rate of aqueous secretion. If the rate of outflow is lower than the rate of secretion, intraocular pressure increases. Normal volume is 0.3 ml.

Functions

The functions of aqueous fluid are:

— to supply the cornea and the lens with nutrients and oxygen;
— to maintain the intraocular pressure, giving the front of the eye its shape;
— to contribute to light refraction.

3.2. The Lens

Definition

The lens is a biconvex, transparent, flexible, crystalline structure located behind the iris and the pupil suspended by the zonules.

Structure

The basic lens structure consists of a central nucleus surrounded by the cortex. The nucleus and cortex are contained within the lens capsule. The capsule is held in place by suspensory ligaments called zonules that insert around the periphery and connect to the muscular ciliary body.

Properties

It is avascular. It has not nerves or blood vessels. The lens derives its metabolic needs from the aqueous and vitreous. The lens also has the highest protein concentration of any tissue in the body (65 % water, 35 % protein).

Its diameter is 9—10 mm and thickness varies with age from 3.5 mm (at birth) to 5 mm (at extreme of age). Its weight varies from 135 mg (0—9 years) to 255 mg (40—80 years of age).

Functions

The main functions the lens are:

— accommodation depending on the flexibility of the lens;
— refraction, helps to focus light rays onto the retina.

3.3. The Vitreous

Definition

The vitreous is a clear, avascular gel-like body filling the posterior segment of the eye, the space between the lens and the retina.

Structure

The outer surface of the vitreous — the hyaloid membrane — is normally in contact with the posterior lens capsule and zonular fibres.

Properties

The vitreous comprises 2/3 of the volume and weight of the eye. It is about 4 ml in volume. Its water content is 98 %.

It is normally firmly attached to optic disc and pars plana and apposed to the retina.

It mechanically stabilizes the volume of the globe, helps to maintain the shape of the posterior chamber of the eyeball and is a pathway for nutrients to reach the lens and retina.

Functions

The vitreous body serves as:

— a buffer to protect the retina against transmitted forces from external blows and pressure;

— a bridge for metabolite transfer between the anterior and posterior chambers of the eye;
— a light refractor.

4. Segments and Chambers of the Eyeball

Clinically, the eye can be considered to be composed of **two segments**:

1. The *anterior segment* — all structures from (and including) the lens forward — the lens, iris, cornea, aqueous humor.

2. The *posterior segment* — all structures posterior to the lens — the vitreous, retina, choroid, and optic disc.

The **three ocular chambers** are:

1. The *anterior chamber* — the space between the cornea and the iris. It is filled with the aqueous humor.

2. The *posterior chamber* — the small space between the iris and the lens. It is filled with aqueous humor behind the iris and in front of the anterior lens capsule.

3. The *vitreous chamber* — the space behind the lens with its gel-like vitreous body.

5. Blood Supply and Innervation

5.1. Arterial Blood Supply

The ***ophthalmic artery*** is the major blood supply to the eye. It is the first major branch of the internal carotid artery. The ophthalmic artery has many branches which supply oxygen and nutrients to the orbit, the eyeball, its muscles, the lacrimal gland, the eyelids and the skin of the forehead.

The ophthalmic artery enters the orbit through the optic foramen with the optic nerve below and lateral to it. As the artery turns medially in the orbit, it gives off branches which are divided into *an orbital group* — distributing vessels to the orbit and surrounding parts; and *an ocular group* — distributing vessels to the muscles and eyeball.

The orbital group consists of the following:

— the lacrimal artery;
— the supraorbital artery;
— the ethmoidal arteries;
— the internal (medial) palpebral arteries;
— the supratrochlear (frontal) artery;
— the dorsal nasal artery.

The ocular group consists of the following (fig. 1.6):

— the central retinal artery;
— the long posterior ciliary arteries;

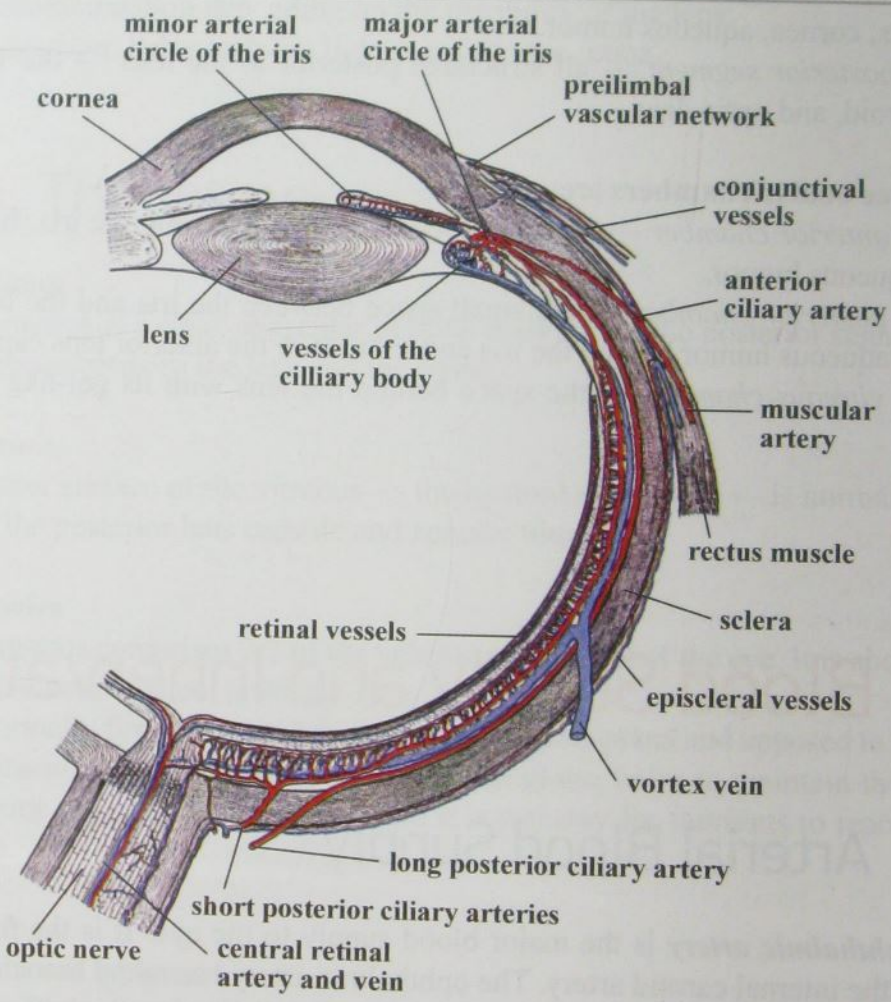


Fig. 1.6. Blood Supply of the Eyeball

— the short posterior ciliary arteries;
— the anterior ciliary artery;
— the muscular artery.

The *central retinal artery* is the first artery branch, as well as one of the smallest branches, of the ophthalmic artery, which passes through the optic nerve 1 cm. behind the eye, enters the eyeball through the center of the optic disc and provides blood flow to the inner retinal layers.

The ciliary arteries are divided into 3 groups: long posterior, short posterior, and anterior ciliary arteries.

The *long posterior ciliary arteries*, two in number, pierce the sclera posteriorly at short distance of the optic nerve, run forward between the sclera and choroid as far forward as the ciliary body and iris. They supply the iris, ciliary body, and choroid.

The *short posterior ciliary arteries,* from 6 to 12 in number, travel anteriorly around the optic nerve to the posterior part of the eyeball, piercing the sclera around the entrance of the nerve. They supply the optic nerve head, the choroid and ciliary processes.

The *anterior ciliary arteries* are derived from the muscular branches of the ophthalmic artery. There are about eight of them. They pass forward and run beneath the conjunctiva, then pierce the sclera a short distance from the corneal limbus. They terminate in the great arterial circle around the iris and in the ciliary processes. They supply the anterior vascular layer of the eye: the iris, ciliary body, and conjunctiva, sclera, rectus muscles.

The *muscular arteries* are numerous small branches that arise in two groups; superior muscular branches and inferior muscular branches. They supply the extraocular muscles.

The *lacrimal artery* is one of the largest branches of the ophthalmic artery, and arises just as the latter enters the orbit. It runs along the superior edge of the lateral rectus muscle and supplies the lacrimal gland, eyelids, and conjunctiva.

The *supraorbital artery* springs from the ophthalmic artery as it crosses the optic nerve. It passes anteriorly along the medial border of the superior rectus muscle and the levator palpebral muscle, then through the supraorbital foramen to supply the muscles and skin of the forehead.

The *ethmoidal arteries:* there are two of them — anterior and posterior. They pass through the anterior and posterior ethmoidal canals, enter the nose to supply the ethmoidal sinuses as well as the frontal sinus and also enter the cranium to supply the meninges.

The *internal (or medial) palpebral arteries* — the superior and inferior — arise from the ophthalmic artery below the trochlea of the superior oblique muscle, descend behind the lacrimal sac to the upper and lower eyelid. They anastomose with the palpebral branches of the lacrimal artery and form a vascular arch in each eyelid. They supply the eyelids, the lacrimal duct and sac, and also the caruncle and the palpebral conjunctiva.

The *supratrochlear artery,* also known as the frontal artery, is one of the terminal branches of the ophthalmic artery, arising at the point where the latter travels posterior to the trochlea. The supratrochlear artery leaves the orbit at the inner canthus of the eyelids by piercing the palpebral fascia. It travels alongside the supratrochlear nerve and ascends to the forehead. Here it supplies the forehead and the scalp.

The *dorsal nasal artery* is the lower terminal branch of the ophthalmic artery. It leaves the orbit in the medial corner by piercing the palpebral fascia, and then it descends to the nose, gives branches to the lacrimal sac and anastomoses with the facial artery. It supplies the upper part of the lacrimal sac, outer surface of the nose.

5.2. Venous Drainage

Venous drainage of the orbit occurs through two major veins — the superior and inferior ophthalmic veins, which pass through the superior orbital fissure into the cavernous sinus.

The central vein of the retina may join an ophthalmic vein or enter the cavernous sinus directly.

The vortex veins drain the uvea (the choroid, ciliary body, iris) into the superior orbital veins and thence to the cavernous sinus encircling the anterior chamber of the eyeball.

The veins of the external orbit drain through the angular vein of the face into the facial veins.

5.3. Innervation

The eye is supplied by three types of nerves: motor, sensory, and autonomic.

The Motor Nerves

The *oculomotor nerve* (cranial nerve III) innervates four extraocular muscles (superior, inferior and medial rectus muscles, inferior oblique muscle), and a muscle of the upper eyelid (levator palpebrae). It also controls pupil constriction (light reflex, pupillary reflex) and thickening of the lens of the eye (accommodation reflex).

The *trochlear nerve* (cranial nerve IV) innervates only one muscle — the superior oblique muscle. Thus it controls eye movements brought about by this muscle.

The *abducens nerve* (cranial nerve VI) supplies a single muscle — the lateral rectus muscle. Thus it has a role in the eyeball movements.

The Sensory Nerve

The *ophthalmic nerve* (cranial nerve V) is one of the branches of the trigeminal nerve that supplies the whole eye and adnexa. This nerve has three branches:

— the *frontal nerve* enters through the superior orbital fissure and provides sensory innervation to the skin of the forehead and upper eyelid, scalp, and mucosa of the frontal sinus;

— the *lacrimal nerve* provides sensory innervation for the lacrimal gland and gives off branches to the conjunctiva and skin of the lateral part of the upper eyelid.;

— the *nasociliary* nerve enters the orbit through the lower part of the superior orbital fissure and gives off a number of branches that innervate the eyeball, orbit, and other parts of the face. One of its branches, the infratrochlear nerve, supplies the eyelids, conjunctiva, and lacrimal sac.

The Autonomic Nerves

The *ciliary ganglion* is a peripheral ganglion of the parasympathetic system of the eye. It is situated in the posterior part of the orbit between the optic nerve and the lateral rectus. It is about the size of a pinhead (1—2 mm) and contains about 2500 neurons. The ciliary ganglion gives rise to the postganglion fibres.

The *sympathetic nerves* originate in the superior cervical ganglion in the cranial portion of the sympathetic chain. They form the sympathetic root of the ciliary ganglion and supply the following smooth muscles — dilator pupillae muscle and Müller's muscle in the lids.

The *parasympathetic nerves* originate from the oculomotor nerve. They form the short oculomotor root of the ciliary ganglion and innervate the ciliary muscle of accommodation and the sphincter pupillae muscle.

6. Visual Pathways

Light that enters the eye via its anterior components travels through different layers of transparent neurons of the retina where it is captured by the photoreceptors at the back of the retina. The neurons of the neural retina convert visual information into nerve impulses, which travel through the optic nerve to the brain. Image on the retina is inverted. The brain translates the image into its upright position.

Information flows from the eyes, crossing at the optic chiasm, joining left and right eye information in the optic tract. Information from the right visual field (now on the left side of the brain) travels in the left optic tract. Information from the left visual field travels in the right optic tract. Each optic tract terminates in the lateral geniculate nucleus (LGN), which is a sensory relay nucleus in the thalamus of the brain.

The LGN consists of six layers: layers 1, 4, and 6 correspond to information from one eye; layers 2, 3, and 5 correspond to information from the other eye. The neurons of the LGN then carry the visual image through the optic radiation to the primary visual cortex (V1) which is located at the back of the brain (caudal end). The visual cortex is the most massive system in the human brain and is responsible for higher-level processing of the visual image. It lies at the rear of the brain above the cerebellum.

The visual pathways include the following (fig. 1.7):

- the retina (rods and cones; bipolar cells, ganglion cells);
- the optic nerve;
- the optic chiasm;
- the optic tract;
- the lateral geniculate nucleus;
- the optic radiation;
- the visual cortex.

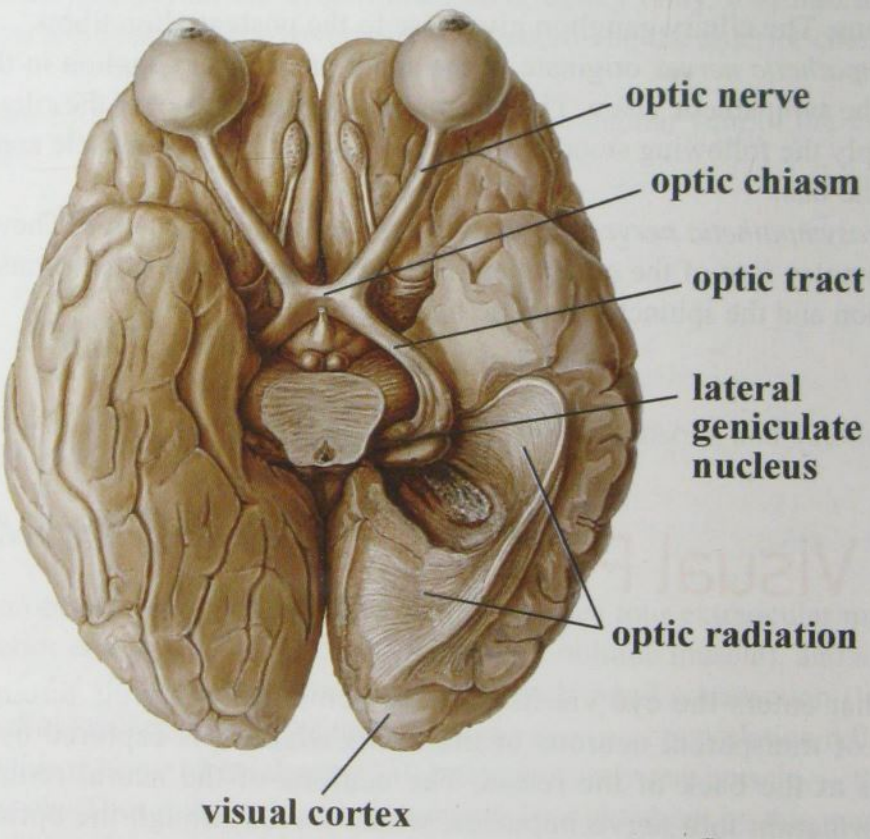


Fig. 1.7. The Visual Pathways

7. The Physiology of Vision

The eye is a complex biological device. The functioning of a camera is often compared with the workings of the eye, mostly since both focus light from external objects in the visual field on to a light-sensitive medium. In the case of the camera, this medium is film or an electronic sensor; in the case of the eye, it is an array of visual receptors.

The primary function of the eye is to form a clear image of objects in our environment. These images are transmitted to the brain through the optic nerve and the posterior visual pathways. The various tissues of the eye and its adnexa are thus designed to facilitate this function.

Light entering the eye is refracted (*refraction*) as it passes through the cornea. It then passes through the pupil (controlled by the iris) and is further refracted by the lens. The cornea and lens act together as a compound lens to project an inverted image onto the retina.

The eye can adjust (*accommodation*) to seeing objects at various distances by flattering or thickening of the lens. Accommodation is also facilitated by changing the size of the pupil. The pupil also constricts with bright light to protect the retina from intense stimulation.

Light rays are absorbed by the photoreceptors on the retina and are changed by electrical activity to transmit the image to the cortex. *Bilateral vision* provides depth perception.

8. Basic Visual Functions

Central vision (or visual acuity) is the central part of visible space. It's main meaning is perception of small details.

Color perception is the ability of the eye to distinguish between different colors as excited by light of different wavelengths.

Peripheral vision (visual field) — is an area which is perceived by the eye while it looks straight ahead. It is not a flat plane but a three-dimensional structure.

Photoperception (or light sense) is the ability of the eye to perceive light as such and in all its gradation of intensity.

Binocular vision (or stereoscopic vision) is vision by two eyes. Normal binocular vision is attained when both eyes are directed towards the object so that the image is projected on the fovea. Although both eyes receive their own images of the object, only one visual object is perceived by the observer. Binocular vision provides a much more accurate assessment of one object relative to another, e.g. its distance, depth, height and width.

Review:

1. Key Points

The *eye* is the organ of vision or sight. It consists of the eyeball, ocular adnexa and supplying structures.

The *ocular adnexa* include the orbit, eyelids, lacrimal apparatus, conjunctiva, and extraocular muscles.

The *eyeball* has three layers, namely:

1. The outer fibrous layer — the cornea and sclera.

2. The middle vascular layer — the iris, ciliary body, and choroid.

3. The inner nervous layer — the retina.

The interior of the eyeball — the aqueous humor, lens, vitreous.

The *segments of the eyeball* — anterior and posterior.

The chambers of the eyeball — anterior, posterior, and vitreous.

Ocular muscles:

— the extraocular muscles — the superior rectus, inferior rectus, medial rectus, lateral rectus, inferior oblique, superior oblique muscles;

— the intraocular muscles — the ciliary muscle, sphincter muscle, dilator muscle;

— the eyelid muscles — the orbicularis oculi, levator palpebrae, Müller's muscle.

The *ophthalmic artery* is the major blood supplier to the eye. It is the first major branch of the internal carotid artery.

Venous drainage of the orbit occurs through two major veins — the superior and inferior ophthalmic veins, which pass through the superior orbital fissure into the cavernous sinus.

Innervation of the eye is provided by three types of nerves — motor, sensory, and autonomic nerves.

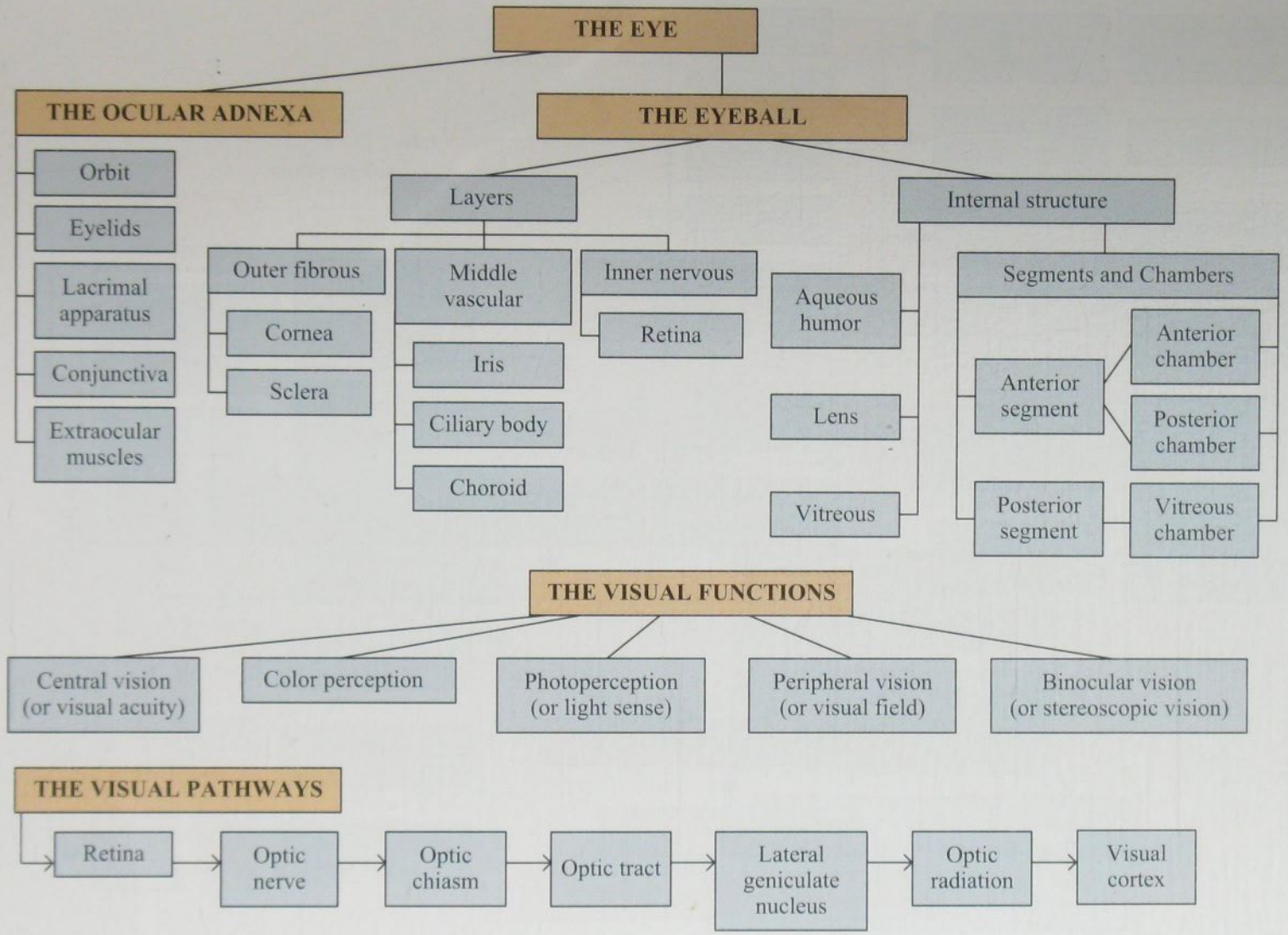
The *visual pathways* are the retina, optic nerve, optic chiasm, optic tract, lateral geniculate nucleus, optic radiation, visual cortex.

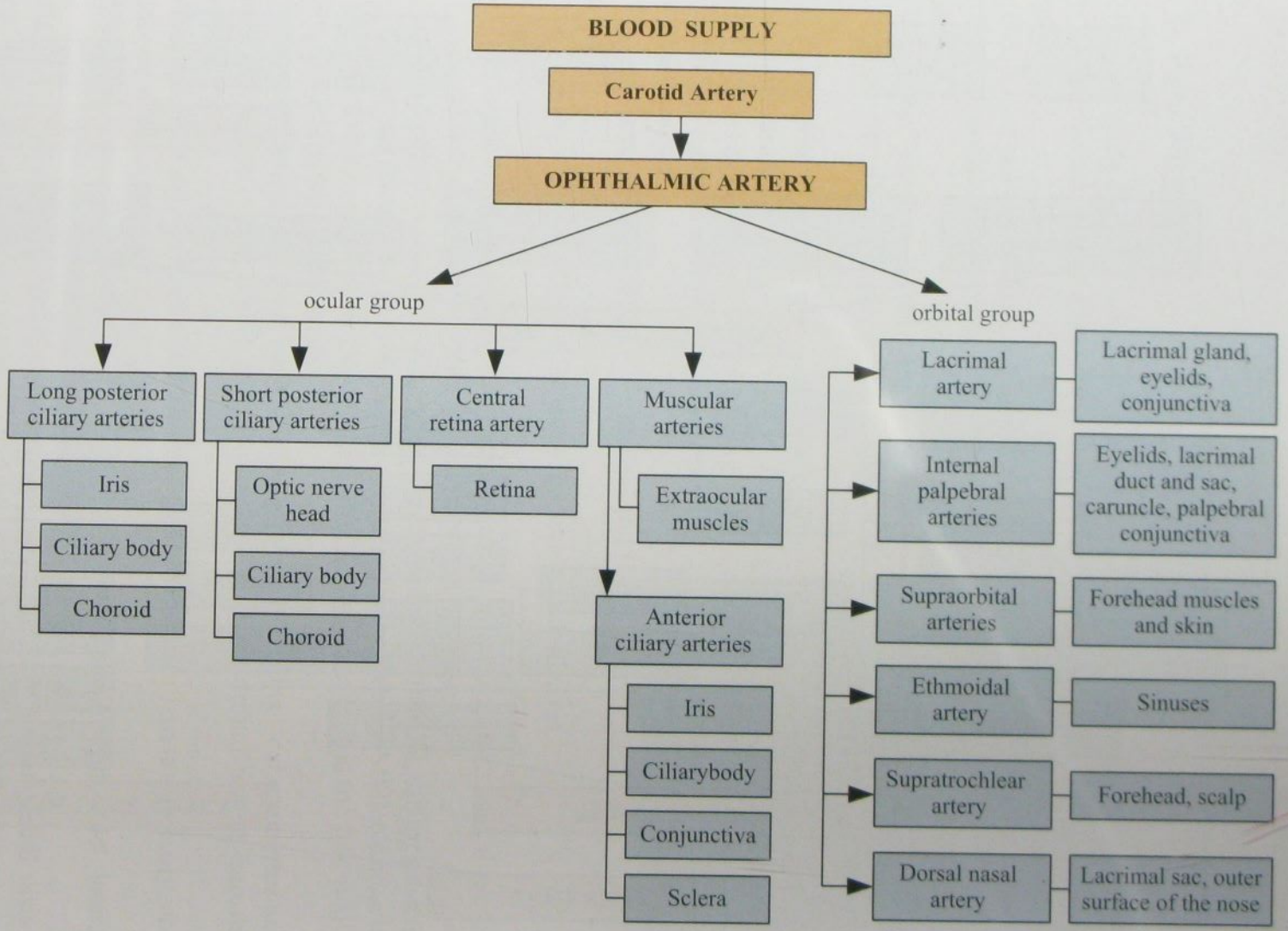
The *basic visual functions* — central vision, color perception, peripheral vision (visual field), photoperception (or light sense), binocular or stereoscopic vision.

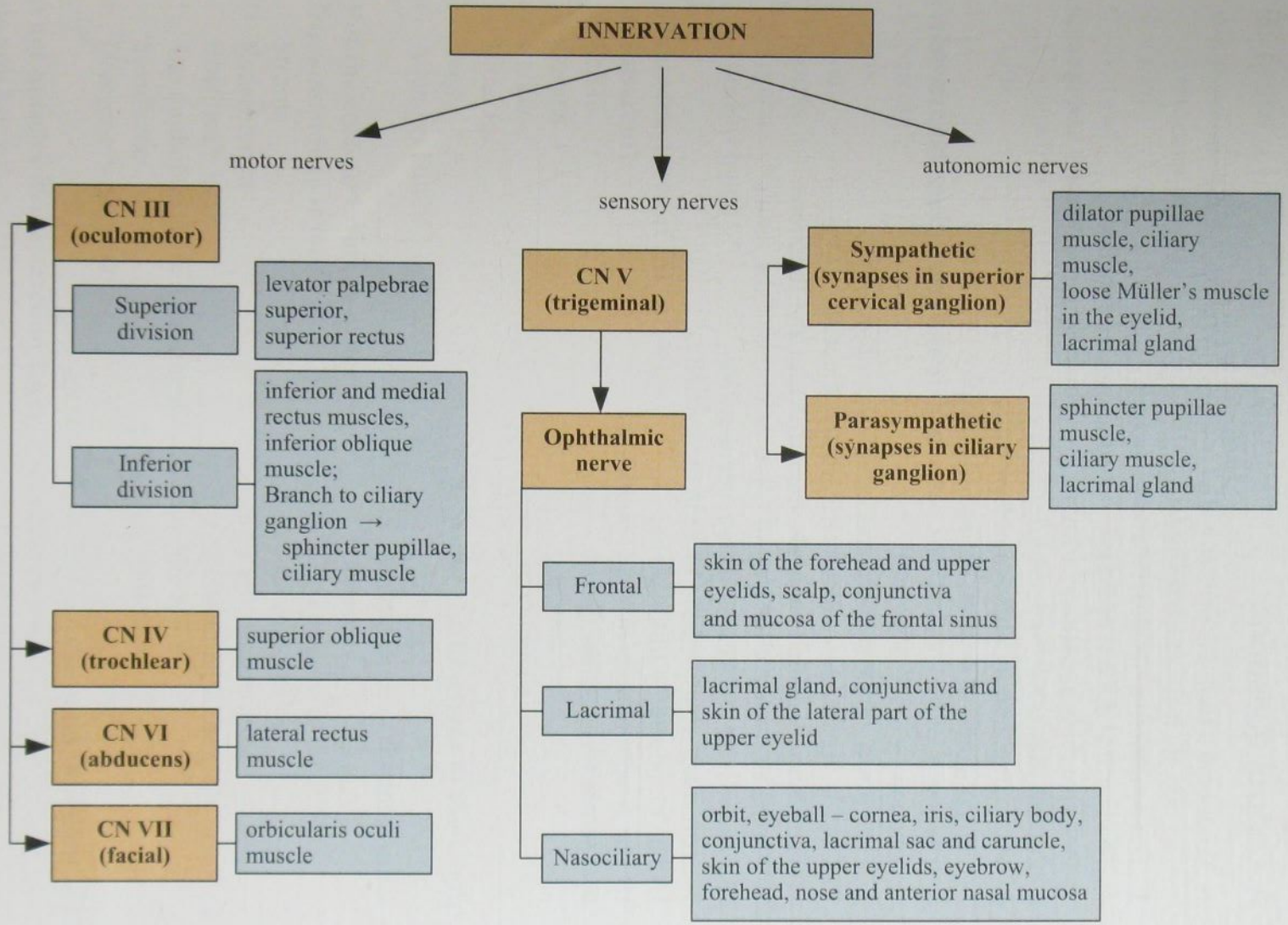
EYE FACTS

The eye is the only part of the human body that can function at 100 % ability at any moment, day or night, without warm-up, without rest. Your eyelids need rest, the external muscles of your eyes need rest, the lubrication of your eyes requires replenishment, but your eyes themselves never need rest. But please rest them!

2. Diagrams


THE EYE
THE OCULAR ADNEXA
Orbit
Eyelids
Lacrimal apparatus
Conjunctiva
Extraocular muscles
THE EYEBALL
Layers
Outer fibrous
Cornea
Sclera
Middle vascular
Iris
Ciliary body
Choroid
Inner nervous
Retina
Internal structure
Aqueous humor
Lens
Vitreous
Segments and Chambers
Anterior segment
Anterior chamber
Posterior chamber
Posterior segment
Vitreous chamber
THE VISUAL FUNCTIONS
Central vision (or visual acuity)
Color perception
Photoperception (or light sense)
Peripheral vision (or visual field)
Binocular vision (or stereoscopic vision)
THE VISUAL PATHWAYS
Retina
Optic nerve
Optic chiasm
Optic tract
Lateral geniculate nucleus
Optic radiation
Visual cortex


BLOOD SUPPLY
Carotid Artery
OPHTHALMIC ARTERY
ocular group
orbital group
Long posterior ciliary arteries
Iris
Ciliary body
Choroid
Short posterior ciliary arteries
Optic nerve head
Ciliary body
Choroid
Central retina artery
Retina
Muscular arteries
Extraocular muscles
Anterior ciliary arteries
Iris
Ciliarybody
Conjunctiva
Sclera
Lacrimal artery
Lacrimal gland, eyelids, conjunctiva
Internal palpebral arteries
Eyelids, lacrimal duct and sac, caruncle, palpebral conjunctiva
Supraorbital arteries
Forehead muscles and skin
Ethmoidal artery
Sinuses
Supratrochlear artery
Forehead, scalp
Dorsal nasal artery
Lacrimal sac, outer surface of the nose


INNERVATION
motor nerves
sensory nerves
autonomic nerves
CN III (oculomotor)
Superior division
levator palpebrae superior, superior rectus
Inferior division
inferior and medial rectus muscles, inferior oblique muscle; Branch to ciliary ganglion → sphincter pupillae, ciliary muscle
CN IV (trochlear)
superior oblique muscle
CN VI (abducens)
lateral rectus muscle
CN VII (facial)
orbicularis oculi muscle
CN V (trigeminal)
Ophthalmic nerve
Frontal
skin of the forehead and upper eyelids, scalp, conjunctiva and mucosa of the frontal sinus
Lacrimal
lacrimal gland, conjunctiva and skin of the lateral part of the upper eyelid
Nasociliary
orbit, eyeball – cornea, iris, ciliary body, conjunctiva, lacrimal sac and caruncle, skin of the upper eyelids, eyebrow, forehead, nose and anterior nasal mucosa
Sympathetic (synapses in superior cervical ganglion)
dilator pupillae muscle, ciliary muscle, loose Müller's muscle in the eyelid, lacrimal gland
Parasympathetic (synapses in ciliary ganglion)
sphincter pupillae muscle, ciliary muscle, lacrimal gland

3. The Review Questions:

A. Control questions:

1. What bones form the orbital walls?
2. The lids, their muscles and function.
3. The conjunctiva, its structure and function.
4. The lacrimal system of the eye, its structure and function.
5. Describe the tear pathway form start to finish. Name the layers and function of the tear film.
6. The extraocular muscles, their function.
7. The optic nerve, its structure and topography.
8. The layers of the eyeball. The outer, middle and inner coats, their sections and function.
9. The cornea, its structure, characteristics and function.
10. The iris, its structure, characteristics and function.
11. The ciliary body and choroids, their structure and functions.
12. The peculiarities of the structure and functions of the retina.
13. The lens, its function, nutrition, peculiarities.
14. How many chambers are there in the eyeball?
15. The blood supply of the globe.
16. The structural parts of visual pathways.
17. The basic visual functions.

B. Tests

1. What are the ocular adnexa?
- A. Extraocular muscles
- B. Orbit
- C. Tear sac
- D. Aqueous humor
- E. Conjunctiva
- F. Lids

2. What are the eyeball contents?
- A. Sclera
- B. Iris
- C. Lens
- D. Cornea
- E. Vitreous
- F. Aqueous humor

3. What structures of the eye are transparent?
- A. Sclera
- B. Cornea
- C. Retina
- D. Vitreous
- E. Lens
- F. Aqueous humor

4. Put the corneal layers in order from outside to inside:
- A. Stroma
- B. Endothelium
- C. Bowman's layer
- D. The epithelium with a basement membrane
- E. Descemet's membrane

5. What muscles of the vascular layer do you know?
- A. Sphincter muscle
- B. Superior rectus muscle
- C. Levator muscle
- D. Ciliary muscle
- E. Orbicularis muscle
- F. Dilator muscle

6. Enumerate the ocular chambers:
- A. Vitreous
- B. Anterior chamber
- C. Lens
- D. Posterior chamber
- E. Aqueous humor
- F. Retina

7. Name the functions of the cornea:
- A. Protecting
- B. Trophic
- C. Light refraction
- D. Light transmission
- E. Maintenance of the intraocular pressure
- F. Accommodation

8. Name the functions of the sclera:
- A. Forming
- B. Maintenance of the intraocular pressure
- C. Participation in the act of accommodation
- D. Light-transmitting
- E. Formation of the clear image on the retina
- F. Protecting

9. What are the main functions of the ciliary body?
- A. Refraction
- B. Accommodation
- C. Aqueous absorption

D. Aqueous production
E. Aqueous outflow
F. Trophic

10. Name the functions of the uvea:
A. Formation of the clear image on the retina
B. Trophic
C. Forming
D. Light refraction
E. Absorption of light rays
F. Maintenance of the intraocular pressure

11. Functions of the aqueous humor of the anterior chamber are:
A. Trophic
B. Mechanical
C. Light transmission
D. Accommodation
E. Light refraction
F. Maintenance of the intraocular pressure

12. Where is the aqueous humor produced?
A. Schlemm's canal
B. Uvea
C. Ciliary processes
D. Retina
E. Zonular fibres
F. Iris

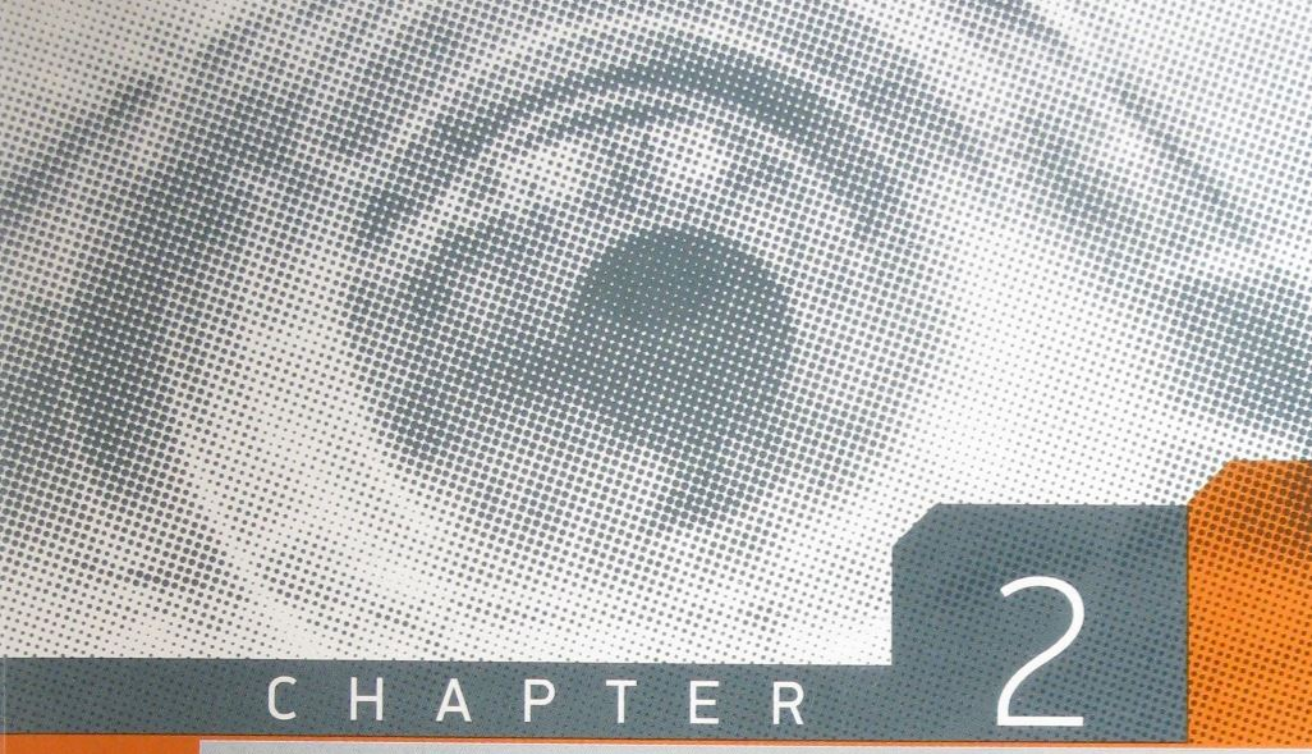
13. Functions of the lens are:
A. Light refraction
B. Light transmission
C. Protection
D. Accommodation
E. Light perception
F. Trophic

14. What is the physiological meaning of the retina?
A. Light perception
B. Forming
C. Light transmissin
D. Trophic
E. Participation in accommodation
F. Transformation of light energy into nerve impulse

15. What arteries are supplying the eyeball?
A. Central retinal artery
B. Lacrimal artery
C. Short posterior ciliary arteries
D. Supraorbital artery
E. Long posterior ciliary arteries
F. Anterior ciliary arteries

16. Put the elements of visual pathways in order:
A. Visual cortex
B. Optic nerve
C. Lateral geniculate nucleus
D. Retina
E. Optic radiation
F. Optic chiasm
G. Optic tract

17. Name the main functions of the organ of vision:
A. Light perception
B. Color vision
C. Accommodation
D. Binocular vision
E. Convergence
F. Central vision



CHAPTER 2

The Ophthalmic Examination

OBJECTIVES

— to facilitate understanding of the importance of complete objective examination of the patient for precise diagnostics and determination of indications for following treatment;
— to be able to take an effective eye history to make a correct diagnosis;
— to know how to perform a basic eye examination.

Plan:

1. BASIC EXAMINATION

1.1. History Taking

- Chief Complaint
- History of Present Illness
- Medical History
  - Past Ocular History
  - Past Medical History
  - Drug History
  - Family History
  - Social History
  - Allergy History

1.2. External Examination

- State of Periorbital Area, Lids and Lid Margins
- Ocular Motility and Binocular Alignment
- Pupillary Size and Response
- Corneal Examination by Side Illumination

1.3. Visual Functions Examination

- Visual Acuity
  - Distance VA Testing
  - VA in children
  - Near VA Testing
- Refraction Test
  - Subjective Refraction
  - Objective Refraction
- Visual Field
  - Confrontation Visual Field Testing
  - Perimetry

1.4. Eye Exam

- Slit Lamp Examination
- Tonometry
  - Palpation
  - Applanation Tonometry
  - Non-Contact Tonometry
- Ophthalmoscopy
  - Red Reflex and the Lens or Lens Examination by Trans Illumination

- Direct Ophthalmoscopy
- Indirect Ophthalmoscopy
- Slit Lamp Ophthalmoscopy

2. ADDITIONAL METHODS OF EXAMINATION

2.1. Exophthalmometry
2.2. Upper Lid Eversion
2.3. Tear / Lacrimal Function Test
- Tear Volume Test
- Tear Film Stability Test

2.4. Cornea Examination
- Corneal Size Measurement
- Corneal Sensitivity
- Fluorescein Staining of the Cornea
- Pachymetry

2.5. Ciliary Body Sensitivity
2.6. Gonioscopy
2.7. Campimetry
2.8. Amsler Grid Test
2.9. Examination of Contrast Sensitivity
2.10. Color Vision Testing
2.11. Examination of Binocular Vision
2.12. Examination of Light & Dark Adaptation

3. INVESTIGATIVE TECHNIQUES

3.1. Keratometry
3.2. Corneal Topography
3.3. Pentacam
3.4. Aberrometry
3.5. Ultrasonography
3.6. Fundus Photography
3.7. Fluorescein Angiography
3.8. Optical Coherence Tomography
3.9. Heidelberg Retinal Tomography
3.10. The GDx Nerve Fibre Analysis
3.11. Visually Evoked Response
3.12. Radiological Imaging Techniques

Content:

As in all clinical medicine, an accurate history and examination are essential for correct diagnosis and treatment. Most ocular conditions can be diagnosed with a good history and simple examination techniques. Conversely, the failure to take a history and perform a simple examination can lead to conditions being missed that pose a threat to sight, or even to life.

1. Basic Examination

1.1. History Taking

The ophthalmic examination is always preceded by a detailed history, which can help to determine any symptoms the individual is experiencing, when they began, the presence of any general health problems, medications taken and occupational or environmental conditions that may be affecting vision.

The history narrows down the differential diagnoses and helps suggest the disease and its cause. A good history can also identify the parts of the clinical examination that need special attention and indicate a need for particular additional investigations.

An ophthalmic history as any medical history starts from taking a *Personal Data* — name, age, sex, occupation (type of work and industrial hazards).

Chief Complaint (CC)

The patient history continues with the chief complaint that is the focus of the exam. It is the primary or most significant symptom(s) that a patient states as the reason for seeking medical care.

Patients with eye disorders commonly present with one or more of following complaints:

- visual loss;
- pain;
- itching, foreign body sensation;
- a change in the appearance of the eye or ocular adnexa (redness, growth on the eyeball, swollen eyelids, etc.);
- discharge from the eye, tearing.

A patient should be asked what exactly is worrying him/her. Sometimes a doctor should help a patient by asking general questions, for example: What is the problem with your eyes? Which eye is affected? Is this symptom/problem constant or occasional? How does this problem interfere with your work or other activities?

If the patient has several complaints, document them in order of highest to lowest medical risk. For example, consider the elements of the exam performed when the patient complains of red eyelids that itch, and compare them to the elements of the exam performed when the second complaint is that the vision in the left eye has become progressively worse over the past month. Because the second complaint might carry the greatest medical risk, it should be listed first.

History of Present Illness

The history of the present illness (HPI) provides a chronological description of how the patient's present illness developed, from the first sign or symptom to the present. You should explore every complaint with the "basic questions" — What? When? Where? Why? How?

Getting a history of the present illness means getting as much information you can about the chief complaint. You are trying to "qualify" and "quantify" the problem. Think of yourself as a detective.

You sometimes see the HPI elements listed in terms of the acronym "SOCRATES", as follows:

Site (location of the problem): Where exactly is the pain? Right or left eye? What part of the eye or vision?

Onset (onset of the problem): When did it start? Was it constant/intermittent, gradual/sudden?

Character (quality of the symptom): How is it affecting you? Bothersome, aware or painful?

Radiation (where the pain moved to): Does it radiate/move anywhere?

Associations (associated symptoms and associations with any conditions or problems): Is it associated with a headache or light sensitivity? Is there anything else associated with the pain e.g. sweating, vomiting? Do you associate the problem with infection, injury, medical conditions, etc.?

Time course: How long have you noticed the symptoms? How long does it last? Is the vision blurry all the time, or during a certain time of day?

Exacerbating/relieving factors (what makes the problem better or worse): Have you done anything to treat the problem? Did the treatment help? Does anything make the symptoms better or worse?

Severity (severity of the symptom): How severe is the pain (consider using the 1—10 scale)? How does it affect daily work/physical activities?

Medical History

This includes the past ocular history, past (systemic) medical history, family and social history, drug history and presence of allergies. Currently treated and past medical conditions, illnesses, injuries, and surgeries should be recorded. You should record the year of onset or occurrence of older incidents, and the month and year of more recent entries. As far as the eyes are concerned, high blood pressure and diabetes are of particular importance.

Past Ocular History

Ocular history should inquire about past clinic visits and surgeries. Ask about previous ophthalmological problems including: poor vision since birth or during childhood; recurrent ocular problems, particularly inflammatory; problems associated with contact lens wear.

Specifically ask about cataract and glaucoma surgeries, refractive surgery, and previous history of eye trauma. You can often piece together your patient's ocular history by examining their eye drops.

For example, the patient's red eye may be associated with complications of contact lens wear, allergy or a corneal abrasion or ulcer. A history of severe myopia (shortsightedness) considerably increases the risk of retinal detachment. A history of hypermetropia (longsightedness) and typically the use of reading glasses before the age of 40 increases the risk of angle closure glaucoma.

Past Medical History

Many systemic disorders affect the eye, and the medical history may give clues to the cause of the problem. Past medical history should include the usual health questions, but with the main emphasis on conditions directly contributing to ocular pathology such as diabetes, hypertension, coronary artery disease and autoimmune diseases. Also, ask about thyroid problems and asthma (you might need to prescribe a beta-blocker for glaucoma but it should be avoided in these patients). Enquire about nasal disease such as sinusitis and hay fever, trauma or surgery.

Drug History

Many drugs affect the eye, and they should always be considered as a cause of ocular problems; for example, chloroquine may affect the retina. Steroid drugs in many different forms (drops, ointments, tablets, and inhalers) may all lead to steroid induced glaucoma. Thus find out what medications the patient is taking, including dosage and how often they are taking them, e.g. once-a-day, twice-a-day, etc. Some questions to ask include: Are you taking any tablets, even homeopathic ones or vitamins? Are you taking any other tablets such as hormone replacement therapy? Do you take any tablets for blood pressure or diabetes?

Family History

There are many disorders in ophthalmology that are inheritable. For any disease that has a genetic component, the age of onset and the severity of disease in affected family members can be very useful information. It is particularly important to ask about having glaucoma, macular degeneration, retinal tears or detachment, strabismus, amblyopia, hypertension, and diabetes in family members. A good example of the importance of the family history is primary open angle glaucoma. This may be asymptomatic until severe visual damage has occurred. The risk of the disease may be as high as 1 in 10 in first degree relatives, and the disease may be arrested if treated at an early stage.

Social History

This is the opportunity to find out a bit more about the patient's background, current life and habits that may affect his or her health. Social history is important in

determining the patient's suitability for treatment, and follow-up. Ask about patient's occupation and hobbies, appreciation of these is needed to understand the patient's visual requirements with respect to sport, driving, reading, etc.

Remember to ask about smoking as it has a large impact on macular degeneration, arteriosclerosis, and can cause optic neuropathy. Alcohol and sexual history should also be considered.

Allergy History

It is important to obtain a history of any allergies, since these may limit the therapeutic options; list basic allergies and their reaction. It does make a difference if a patient simply had itching following a fluorescein angiogram, or if the patient went into anaphylactic shock. In general, five types of allergic responses should be inquired about:

— allergy to drugs (taken internally or applied topically);
— allergy to inhalants (dust, pollens, etc.);
— allergy to contactants (cosmetics, woollens and so forth);
— allergy to ingestants (food allergies);
— allergy to injectants (tetanus antiserum).

1.2. External Examination

After history taking the basic ophthalmic examination consists of an external examination, evaluation of extraocular muscle motility and pupil function, followed by specific tests for visual acuity, visual fields, intraocular pressure and ophthalmoscopy.

State of Periorbital Area, Lids and Lid Margins

External examination of eyes consists of *inspection of the eyelids, surrounding tissues and palpebral fissure* (remember the **four L**'s: **l**ids, **l**ashes, **l**acrimal apparatus and **l**ymph nodes). It is carried out in daylight or artificial illumination.

Before the beginning of external examination one should pay attention to the form, size of a head and a facial skull, check whether the left and right sides of the face and its regions are symmetrical, evaluate the eyeball position in the socket.

Ocular Motility and Binocular Alignment

Ocular motility should always be tested, especially when patients complain of double vision or neurologic disease is suspected. Extraocular movements can be tested uniocularly as well as binocularly.

Sit up close, in front of the patient. Ask the patient to focus on an object (such as a pen or a finger) and with the eyes only follow it in each of the 9 diagnostic positions of gaze: 1 — straight ahead; 2 — right; 3 — upperright; 4 — up; 5 — upperleft; 6 — left; 7 — lowerleft; 8 — down; and 9 — lower right. This allows the examiner to diagnose strabismus, paralysis of the ocular muscles, and gaze paresis.

Binocular alignment is evaluated with *a cover test*. During a cover test, one eye is covered while the other eye focuses on an object at distance or near. Cover tests are part of all comprehensive eye examinations given to young children because they are at a higher risk of developing problems with eye alignment. Amblyopia is currently the leading cause of vision loss in children.

There are two types of cover tests performed during eye examinations: the unilateral cover test and the alternating cover test. During the unilateral cover test, a patient is asked to focus on a distant object while the doctor covers each of a patient's eyes in turn. If either of the uncovered eyes has to move to focus on the object, this may be evidence of strabismus. The second part of the exam is the alternating cover test. A patient is asked to focus on an object while the eye cover is switched from one eye to the other. If the doctor detects eye movement after the eye cover is removed, this is an indication of phoria. A significant amount of phoria can lead to eyestrain and/or double vision.

Ocular alignment may be assessed by the *Hirschberg corneal light reflex test.* This test relies on observing the light reflected off the cornea. Normally the corneal reflex is at the center of the pupil and symmetrical in both eyes — orthophoria — correct eye position. If the reflection is displaced, strabismus may be present. Temporal displacement of the light reflex (inward or nasal deviation of the eye) indicates esotropia, nasal displacement (outward or temporal deviation) indicates exotropia, inferior displacement (upward deviation) indicates hypertropia, and superior displacement (downward deviation) indicates hypotropia.

The Hirschberg test estimates the degree of the eyes deviation by determining how far the displaced light reflex is off center. Each mm of deviation of the corneal light reflection is equivalent of 7° turn of the visual axes. When the reflex is situated at the pupil margin the deviation is 15°, in the mid-iris region — about 30°, on the limbus — about 45°, out of the limbus — 60° and more (fig. 2.1). While it is not a very accurate test, nevertheless it is useful in young children and in estimating the deviation if one eye is blind (non-fixing) as the cover test cannot be performed in this situation.

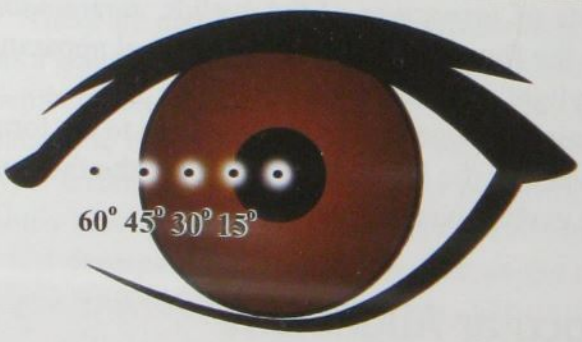


Fig. 2.1. Determination of the eye deviation degree by Hirschberg corneal light reflex test

To perform the *Hirschberg corneal light reflex test,* follow these steps:

— Sit in front of the patient at a distance of about 40 cm.
— Hold a source of light (a penlight or an ophthalmoscope) in front of the patient's eyes at a distance of about 30—33 cm.
— Ask the patient to look directly into the light and assess the location of the corneal light reflex in each eye.

Pupillary Size and Response

The pupils are normally round, black and symmetric to each other. They constrict in reaction to direct illumination (direct response) and to illumination of the opposite eye (consensual response). The pupil dilates in the dark. Both pupils constrict when the eye is focused on a near object (accommodative response).

EYE FACTS

About 20 % of the healthy population has unequal pupils ("anisocoria").

An examination of pupillary function includes inspection of the pupils for equal size, regular shape, reactivity to light, and direct and consensual accommodation. These steps can be easily remembered with the mnemonic PERRLA (D+C): **P**upils **E**qual and **R**egular; **R**eactive to **L**ight and **A**ccommodation (**D**irect and **C**onsensual). Pupillary assessment is an important part of neurological examination because changes in the size, equality and reactivity of the pupils can provide vital diagnostic information in a critically ill patient.

Examination of the pupils begins with the assessment of the *pupillary size and form.* The normal pupil size in adults varies from 2 to 4 mm in diameter in bright light to 4 to 8 mm in the dark. Pupils smaller than 2 mm are said to be miotic; pupils larger than 6 mm are mydriatic. Miotic pupils may be caused by antiglaucoma medications, chronic iris inflammation, age, or a neurologic disorder. Mydriatic pupils are normally more common in children and myopic eyes. Abnormal mydriasis is caused by certain drugs, neurologic disorders, iris injury, or acute glaucoma. The pupils should be equal in size, although a small difference (1 mm) may be normal variation.

Both pupils' shape should be round; abnormal shapes may indicate cerebral damage. For example, oval shape could indicate intracranial hypertension. The pupils are normally centered or a little nasal in the iris. An eccentric pupil may be the result of faulty embryonic development, injury, intraocular surgery, or inflammation.

NOTE!

Dilated and fixed pupils and pupils of unequal size may indicate serious neurologic disorders and conditions, such as head trauma, stroke, brain tumors or brain death. These conditions are medical emergencies!

Pupil size changes may accompany symptoms of drug use, drug overdose, or poisoning.

In some cases, pupil symptoms may be a sign of a serious or life-threatening condition that should be immediately evaluated in an emergency setting.

Measurement of the pupil size should be held under normal lighting conditions using a millimeter ruler (or half circles on the bottom of a ruler). With experience, the examiner can become fairly accurate at estimating this measurement by sight.

To measure the *pupil's size*:

— Ask the patient to sit on a chair.
— Sit in front of the patient and ask him/her to fix on a distant object.
— Place the millimetre ruler on his/her lower eyelid, trying to hold it away from the visual axis of the patient, and measure the pupillary size.
— Repeat for the left eye.
— Compare the sizes of the pupils.
— Note and compare the shapes of the pupils.

The *pupil light response test* assesses the reflex that controls the size of the pupil in response to light. Testing pupillary reactions requires a dimly lit room, to avoid interference from daylight. The patient should focus on a distance target to eliminate the accommodation reflex. A source of light (a penlight or an ophthalmoscope) is used and the pupil should constrict immediately when a light is shone directly into it (direct response); the contralateral (opposite) pupil should also constrict at the same time (consensual response). Both pupils should be equal in their response and should dilate simultaneously when the light source is removed.

If one of the eyes is not detecting light well, then the pupils will not constrict as well when light is shone in that eye. Often, however, this pupil defect is subtle. The *swinging light test* enables to pick this up by comparing the pupil response *between* the eyes. Swing the light back-and-forth between the eyes. A normal response should be brisk constriction of each pupil as light is shone onto the eye, with a consensual response seen in the other eye, i.e. constriction. Things look different if one eye doesn't see well. As before, when you shine the light in the good eye, there is constriction, but when you turn to the bad eye, both eyes seem to dilate a little. The bad eye still senses light and constricts, but not as well. This phenomenon is known as a relative afferent pupillary defect (RAPD) and also called a Marcus Gunn pupil and indicates a possible optic nerve problem.

In order *to test pupil reaction to accommodation and convergence* the patient should be asked to fix first on the distant object and then to look at a near object (a pen or an ophthalmoscope) at a distance of 20—25 cm; the normal pupils constrict. This is termed the near reflex.

To test the *direct pupillary reaction to light*:

— Ask the patient to sit in front of you and look at a distant object.
— Direct the light of a penlight or an ophthalmoscope at the patient's right eye.
— Watch if the pupil constricts (a normal reaction).
— Repeat for the left pupil.

To test the *consensual pupillary reaction to light*:

— Ask the patient to sit in front of you and look at a distant object.
— Direct the light of a penlight or an ophthalmoscope at the patient's right eye.

— Watch the left pupil to see if it constricts along with the right pupil (a normal consensual response).
— Repeat for the left pupil, watching the right pupil for response.

Corneal Examination by Side Illumination

The method of side, or focal, illumination is necessary for examination of the ocular adnexa and anterior part of the eyeball (lids, lacrimal puncti, conjunctiva, sclera, limbus, cornea, anterior chamber, iris and pupil). The eye is examined with a point of light source and a lens or loupe placed at the side of the patient (that is why the method is called side illumination) (fig. 2.2).

Particularly the cornea is examined by side illumination paying attention if it is smooth, transparent and reflective. The reflection is distorted in the presence of corneal disorders. The examination is held in a dim lit room.

To *exam the cornea by side illumination,* follow these steps:

— Ask the patient to sit down in front of you.
— Place the lamp at the left side and in front of the patient at a distance of about 40—50 cm.
— Illuminate the patient's eye by focusing the light beam from the lamp with the help of the lens of dioptric power 20 D placed at a distance of 7—10 cm from the patient's eye.
— For a more detailed examination you can hold the 13 D lens with the left hand in front of the patient's eye.
— Pay attention if the cornea smooth, transparent, reflective and at signs of inflammation, dystrophy, laceration or foreign body presence.

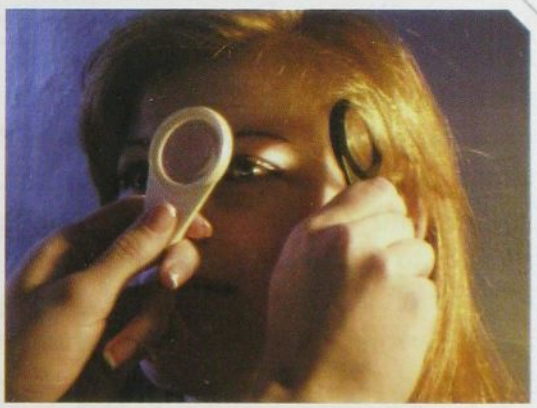


Fig. 2.2. Examination of the cornea by side illumination

1.3. Visual Functions Examination

Visual Acuity

Visual acuity (VA) is the most common measurement of central visual function. The term "vision acuity" was introduced by Dodners to describe the "sharpness of vision", in other words — perception of small details. Therefore, visual acuity, the

sharpness of vision, is a measurement of the eye ability to distinguish two points, located at the minimal distance from one another*.

The following are common abbreviations used to denote visual acuity:

— VA or Visus — visual acuity;
— OD (*oculus dexter*) for the right eye;
— OS (*oculus sinister*) for the left eye;
— OU (*oculi uterque*) for both eyes.

Distance VA Testing

We determine visual acuity with visual charts. They contain letters or symbols which are called optotypes calculated in angular minutes (the principle of Snellen**) and arranged with the larger ones at the top and the smaller ones at the bottom. The eye charts must be clean and well illuminated for the examination. Each eye is tested separately.

In countries of North and South America as well as in Western Europe visual acuity is commonly assessed by the *Snellen chart*. This is a board that consists of high contrast letters, each row is decreasing in size to the bottom. Snellen's optotypes included only 9 letters: C, D, E, F, L, O, P, T, and Z. Each row is denoted a number from 20 to 200 and corresponds to the distance from which a "normal" eye, i.e. one without refractive errors, could read that row of letters.

EYE FACTS

Some birds, such as hawks, are believed to have visual acuity of around 20/4, meaning that the detail that you can see clearly at 20 feet a hawk can see at 100 feet away.

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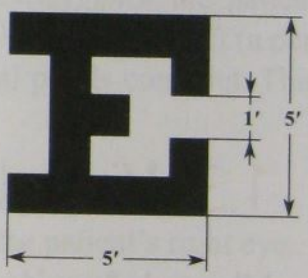
* The smallest viewing angle of the healthy eye at which two distinct points can still be discriminated is approximately 1 angular minute (1'); this corresponds to a diameter of individual cone (4μ). If the images fell on two cones separated by an unilluminated cone, then the points would be perceived as two distinct sources. The dimensions of the visual angle depend upon the size of the object as well as its distance from the eye.

** Herman Snellen, Dutch ophthalmologist (1835—1908), was the first to develop a visual acuity chart. Snellen assumed that the "average" eye could just read a letter if the thickness of the lines and the spaces between them subtended 1 angular minute (1'), and the whole letter — 5 angular minutes (5'). The standard distance for this test is 20 feet or 6 meters.



According to Snellen, vision acuity is calculated as a fraction: Visus = d/D, where d is the distance at which the chart is viewed; D — the distance at which the "normal" eye can read the same row of the chart. Normal vision is therefore recorded as 20/20 or 6/6. This says that one sees as well at 20 feet as an average person would at 20 feet. Vision of 20/100 (6/30) means that at 20 feet (6 m) from the chart a patient can read a letter, which someone with normal vision would be able to read from 100 feet (30 m) away. If a person has a visual acuity of 20/40, he/she is said to see detail from 20 feet (6 m) the same as a person with normal eyesight would see from 40 feet (12 m).

Other charts with a similar principle are used too, e.g. *Landtolt C chart* (broken ring or C in different orientations); *Tumbling E chart* (letter E rotated in various positions) (fig. 2.3). Other charts are usually used if patients cannot read English and are also useful for young children (charts with pictures), to provide an indication of visual acuity without a requirement to read or verbalize.

The *logarithmic chart (LogMAR)* is another commonly used scale, especially for large scale clinical trials and orthoptic childhood screening. The LogMAR scale converts the geometric sequence of a traditional chart to a linear scale. It measures visual acuity loss: positive values indicate vision loss, while negative values denote normal or better visual acuity. This scale is rarely used clinically.

The *Golovin—Sivtsev Table* is a standardized table for visual acuity testing which is used in Ukraine and several countries of Eastern Europe. The table consists of two parts with 12 rows each. A value D indicated to the left of each row gives the distance in meters from which a person with a "normal" vision can read the corresponding row. A value V indicated to the right gives the minimum visual acuity needed to read the row from a distance of 5 meters. VA is denoted in decimal value of the Snellen ratio (0.1, 0.2, 0.3, … 1.0, etc.); normal vision is denoted as VA = 1.0. The rows represent visual acuity values between 0.1 and 2.0. The left part consists of series of the Cyrillic

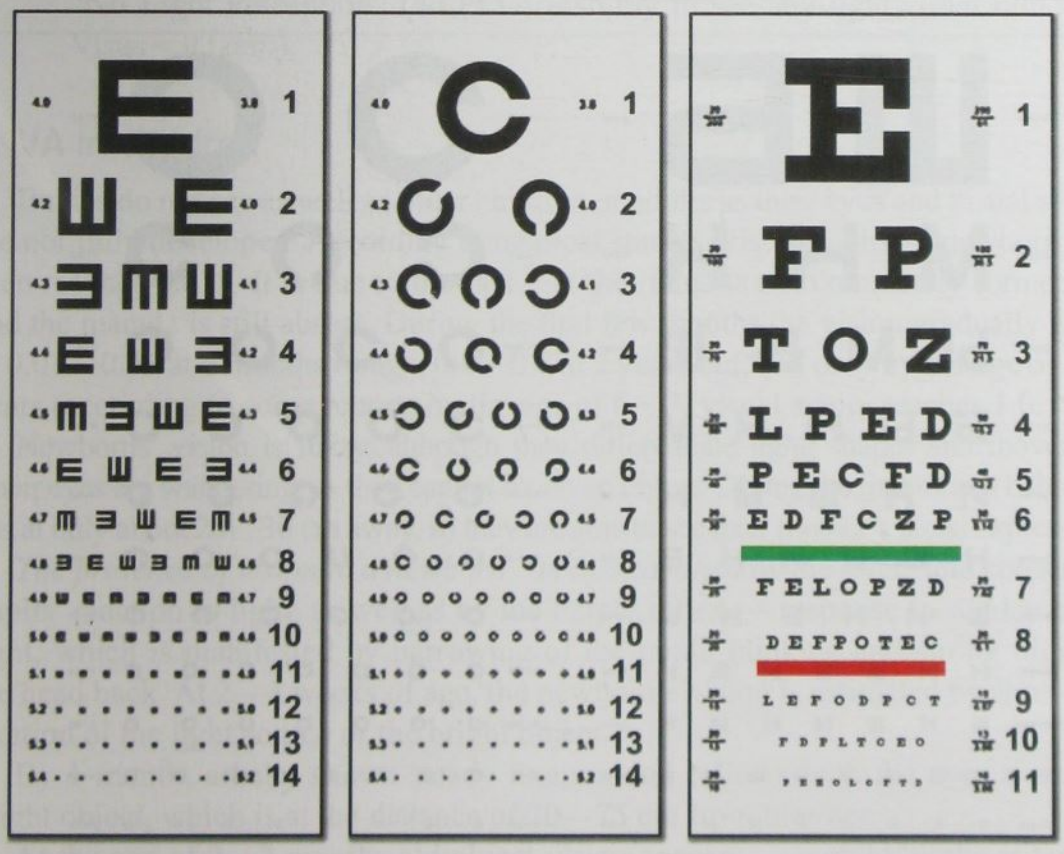


Fig. 2.3. The Tumbling E chart, the Landtolt C chart, and the Snellen chart, used for visual acuity test

letters Ш, Б, М, Н, К, Ы and И in a definite order. The right part of the table consists of Landolt C symbols (fig. 2.4).

To test *distance visual acuity with the Golovin—Sivtsev table*, follow these steps:

— Make the patient sit at the designated distance, usually 5 meters, from the well-illuminated chart.

— Visual acuity is tested separately for each eye (with and without glass), so ask the patient to cover one eye with their hand or by using a hand-held occluder. The fingers should not be used to cover the eye because the patient will be able to see between them. By convention, the right eye is tested and recorded first.

— If a patient wears eyeglasses or contact lenses, then these should be worn.

— Ask the patient to read down to the lowest row of letters that they can see clearly.

— Record the visual acuity according to the row of letters being read (it is accepted to make one mistake in 3—6 rows, and two — in 7—10 rows). For example, a patient reads the 5th line of the Golovin—Sivtsev table with mistakes in 2 letters; the visual acuity is equal to 0.4.

— Repeat the procedure for the other eye.

If visual acuity is 0.5 or less in one or both eyes, test each eye again with the pinhole occluder and record these results. The use of the pinhole should give a more accurate reading of their visual acuity, as any true refractive errors not corrected by

Fig. 2.4. The Golovin—Sivtsev table for visual acuity testing

the patient's eyeglasses will be eliminated by the pinhole, or if a patient cannot use contact lenses because of a red eye or has not brought their glasses.

If a patient is unable to read the largest optotype on the Table, move him/her closer to it. Once the patient is able to read the chart, the letter size and test distance are noted. Visual acuity is calculated in this case by the formula: Visus=d/D, where d — a distance from the chart at which the patient can read; D — a distance at which the "normal" eye can read the same row of the chart. For example, if a patient reads the 2nd line of the Golovin—Sivtsev table from a distance of 2 m, the visual acuity calculated by the formula will be 2/25=0.08.

If the patient is unable to read the chart at any distance, he or she is tested as follows:

— Counting Fingers (CF) — the ability to count fingers at a given distance (1 m). If this is unsuccessful, proceed to elicit whether the patient can perceive hand movements.

— Hand Motion (HM) — the ability to distinguish a hand if it is moving or not in front of the patient's face. If a patient can't detect hand motion, test light perception by use of a pen light or an ophthalmoscope.

— Light Perception (LP) — the ability to perceive any light (correct and incorrect light projection) — Visus = 1/proectio lucis certa; Visus = 1/proectio lucis incerta.

— If the patient sees nothing at all, then the visual acuity should be recorded as "No Light Perception" (NLP) — inability to see any light. Total blindness. Visus = 0 (zero).

VA in children

Babies do not see as well as older children or adults as their eyes and visual system are not fully developed. According to medical studies, visual acuity of newborn children reaches 0.015. It is due to the fact that the retina is not completely formed yet, and the macula is still absent. During the first few months the vision gradually grows to 0.01—0.03 at 6 months of age, 0.4—0.7 at 2 years old, and only by the age of 3—5 years (according to some reports by the age of 6—7) visual acuity reaches 1.0.

Newborns' vision is fuzzy, although they differentiate light, shapes and movement. Their eyes are wandering, as they cannot fix on an object. During the first month babies focus at only about 20—30 cm away, so they are able to see their mother's facial expressions.

The presence of vision in a newborn can be examined by the direct and consensual pupils' reaction to light, as well as by the Peiper reflex — response to sudden bright light, which is manifested by narrowing of the pupil, blinking and strong tilting of the head back. At 2—3 weeks of age, the newborn's vision is estimated by short-term fixation of the light source or the bright object.

By 1 month, a baby shows steady fixation and follows with the eyes a moving bright object, which is at the distance of 20—75 cm from him/her.

At the age of 2—3 months old object vision appears — a child blinks at fast approaching of an object to the eyes (the reflex of danger), examines his/her hands, finds the mother's breast, responds to the appearance of people.

By 3 months, babies are learning how to move their gaze from one object to another without having to move their head. Ocular movements are coordinated.

Between four and six months, a baby recognizes parents and smiles. The baby also shifts from preferring what is familiar to that which is new. The child will also look for objects when they fall from view.

To test baby's vision at 2—5 months a bright red ball with a diameter of 4 cm is used. The ball is placed at different distances from the child and his/her attention is attracted by its movements. The ball is gradually approaching the child and the examiner notes the distance from which the child begins to follow the ball with the eyes, or reaches a hand toward it.

Between 6—12 months, baby's vision acuity improves rapidly. By 1 year, near and distant acuities are good. A child examines objects in hands visually, watches what is going on around him/her and can transfer objects from hand to hand. They can discriminate between simple geometric forms, scribble with a crayon and are visually interested in pictures. A child can visually spot a small (2—3 mm) object nearby; watches faces and tries to imitate expressions; can differentiate between known and unfamiliar people. At this age, a child can recognize him/herself in the mirror and parents in the photos.

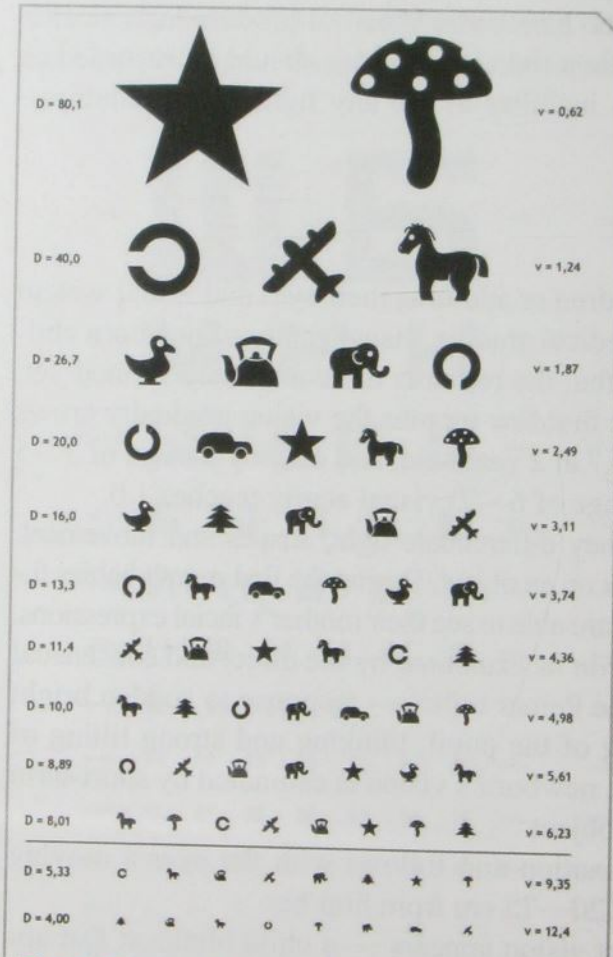


Fig. 2.5. The Orlova table, used for visual acuity test in children

To test baby's vision at 6—12 months we also use a bright red ball only with a diameter of 0.7 cm. If the child begins to distinguish the ball (there are tracking movements) from a distance of 5 meters, the visual acuity is about 1.0, from the distance of 1 m — 0.2, 50 cm — 0.1.

By 2—3 years, myelinisation of the optic nerve is completed and retinal maturity is almost achieved. There is vertical (upright) orientation; all optical skills are smooth and well coordinated. A child can complete a simple formboard correctly (based on visual memory), can do simple puzzles, match objects of the same type by properties (color, shape), and can point at pictures in a book. Visual acuity at this age is examined by the simultaneous tracking of soundless toys of different sizes.

By 4—5 years, there is slower ongoing development of the eye until it

is complete. For vision examination at this age, there are a number of different assessment methods appropriate for young children, varying from matching tests to special charts or tables, in which as optotypes familiar for children objects and animals are shown (the Orlova table) (fig. 2.5).

Visual acuity of children *older than 5 years* can be checked with the help of traditional charts and by the traditional method. The distance from the chart to the patient is 5 m. The child is asked to alternately close the left, then the right eye and name optotypes. At the age of 6—7 years normal visual acuity is considered to be of 1.0. (average VA of children is shown in table 2.1).

Table 2.1

Average Visual Acuity of Children According to Age (www.detskoezrenie.ru/rod)

Child's Age	Average Visual Acuity
New-born	0.002—0.02 (up to 2 %)
1 month	0.008—0.03 (up to 3 %)
3 months	0.05—0.1 (5—10 %)
6 months	0.1—0.3 (10—30 %)
1 year	0.3—0.6 (30—60 %)
2 years	0.4—0.7 (40—70 %)
3 years	0.6—0.9 (60—90 %)
4 years	0.7—1.0 (70—100 %)
5 years	0.8—1.0 (80—100 %)
7 years	0.9—1.2 (90—120 %)
8—15 years	0.9—1.5 (90—150 %)

Near VA Testing

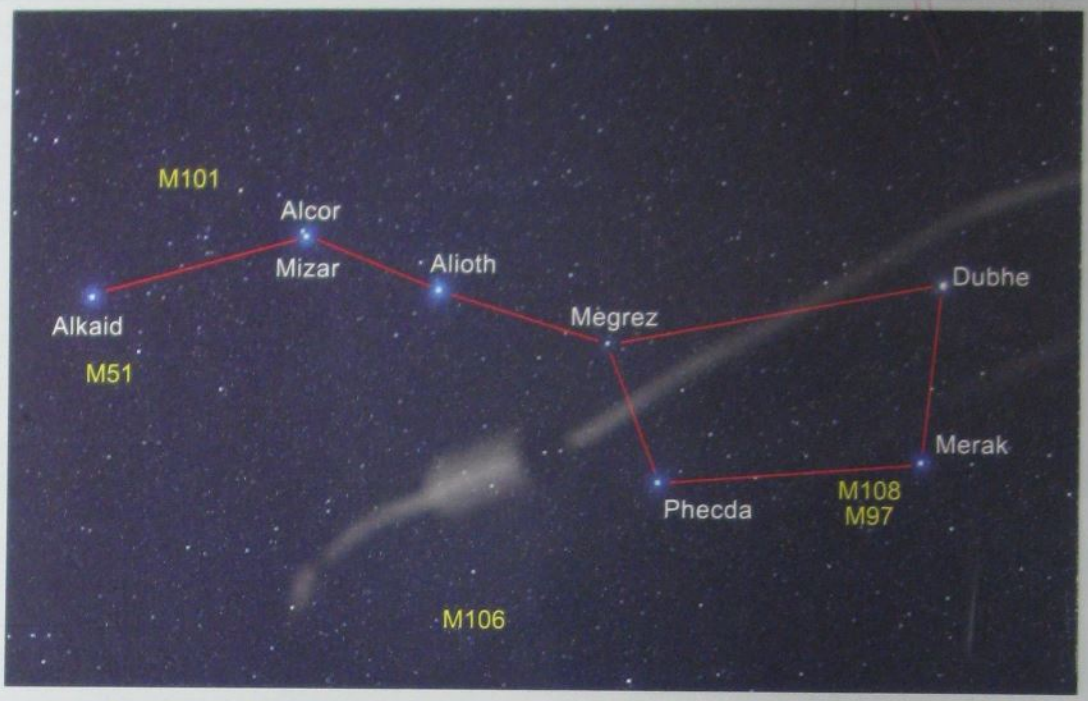
Near vision can be assessed with a reading test-type book or a near-chart. It is important to ensure that the book is adequately illuminated. Ask the patient to wear reading glasses if they usually wear them. Test each eye separately.

The patient should hold the near test chart at about 25—30 cm to read the smallest print that he/she can comfortably see. The size of the test type seen with each eye, and then both eyes simultaneously, is recorded.

Refraction Test

Refraction is a necessary test used to determine a patient's best corrected or potential visual acuity in case of need for prescription of eyeglasses or contact lenses. Usually it is performed after visual acuity test in cases when a patient has less than 1.0 (20/20) vision which detects refractive error or vision problem.

EYE FACTS



Vision testing in ancient times was as important as it is today. A common ancient naked eye test used the double star of the Big Dipper (also known as the Ursa Major or the Big Bear). The second star from the end of the handle of the Big Dipper is an optical double star. It can best be seen with careful observation on a moonless, clear night. The ability to perceive this separation of these two stars, Mizar and Alcor, which are separated by 12 min of arc, was considered a test of good vision and was called the Arab Eye Test. It has been used by the Arabs when choosing their horsemen, by American Indians as a test of visual acuity. Alexander the Great is said to have required all of his generals to be able to resolve this pair.

A refractive error is an optical defect that does not allow light to be brought into sharp focus on the retina, resulting in blurred or distorted vision. Examples of refractive error are myopia (nearsightedness), hyperopia (farsightedness) and astigmatism. During the refractive test a doctor determines what lens power is needed to compensate for refractive errors so that light comes to a sharp focus on the retina.

There are two methods of evaluating the refractive error of an eye:

— Subjective refraction — where the result depends on the patient's ability to discern changes in clarity. His process relies on the cooperation of the patient.

— Objective refraction — where the result depends purely on the examiner's judgment to determine the optimum optical correction.

Subjective Refraction

The subjective refraction test consists of placing various combinations of lenses before the patient's eye until the maximum visual acuity is reached. The trial frames (fig. 2.6) or phoropter are used for this purpose.

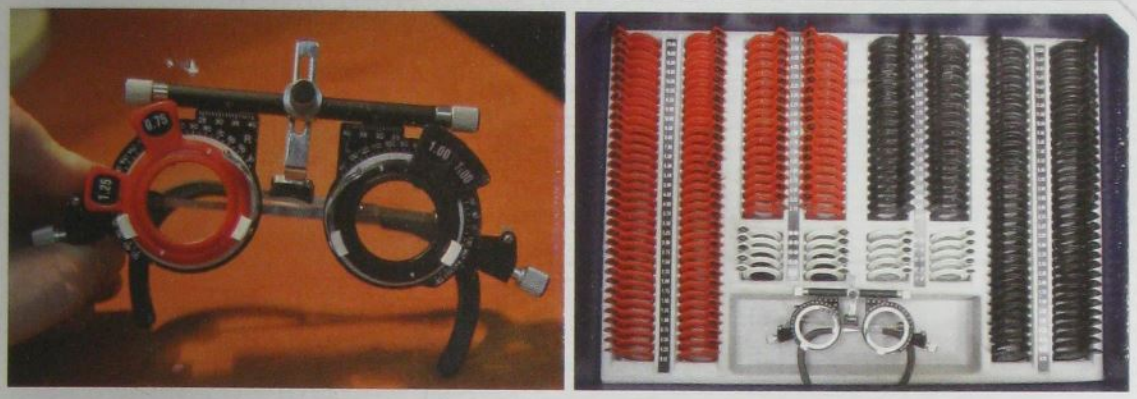


Fig. 2.6. The trial frame with lens and lens set for performing subjective refraction test

A phoropter (also called refractor) is a device commonly used, in conjunction with a Snellen chart, to determine the best corrective lenses for a person being assessed for eyeglasses or contact lenses. The phoropter contains a complete range of corrective lenses, allowing the person to compare different levels of correction while viewing the chart. Typically, the doctor uses the phoropter to refine the information obtained from the autorefractor before prescribing lenses.

To obtain an accurate refraction in young children who may skew refraction measurements by subconscious accommodation that make the result invalid cycloplegic eye drops are used to temporarily relax the ciliary muscle of the eye.

Objective Refraction

Retinoscopy (skiascopy or shadow test) and automated refraction are objective refraction tests used to determine refractive errors without interaction or subjective information from the patient. They are especially useful when examining non-speaking patients or young children.

Retinoscopy is an objective method of investigating, diagnosing, and evaluating refractive errors of the eye. The term retinoscopy literally means "an exam of the retina". A bright streak of light is shone through the pupil and is seen as a red reflex reflected from the retina. The retinoscope streak is moved gently, and the direction of the light reflex from the retina is observed. Based on the movement and orientation of this retinal reflection, the refractive state of the eye is measured. By placing a series of trial lenses of different power in front of the patient's eye, the observer can calculate whether the patient is shortsighted (myopic) or longsighted (hypermetropic) and measure the amount of astigmatism that needs correction.

Automated refractometry is the method of measuring refraction automatically with the aid of light-sensitive detectors and a computer until a focused image appears on the retina. These systems operate with infrared light based on the movement and orientation of this retinal reflection, the refractive state of the eye is measured.

Visual Field

Visual field testing should never be omitted from the basic eye examination. All patients should have their visual fields (peripheral vision) checked. A patient may have great central vision, with perfect eye-chart scores, but suffer from "tunnel vision" resulting from neurological diseases or glaucoma. Your patient may not even be aware of this peripheral visual loss if it has progressed slowly over time.

Visual field is an area which is perceived by the eye while it looks straight ahead. It is therefore the locus of all points in space, which is visible when one point is fixed. The borders of the visual field are measured in angle degrees. The boundaries of normal visual field for a white color object are temporally — 90°, down temporally — 90°, down — 70°, down nasally — 50, nasally — 60°, up nasally — 55°, up — 60°, and up temporally — 70°. In addition, it is important to mention that visual field boundaries depend on anatomical peculiarities of a patient's facial scull constitution and can slightly vary. (Total visual field with both eyes open is approximately 180° horizontally.)

EYE FACTS

Chameleons are best known for their ability to change color. But they also have very unique eyes: the eyelids are fused, covering almost the full eyeball except for a small hole that the pupil can see through. The eyes move independently of one another, which provides a full 360-degree field of vision.

A visual field test is an eye examination that can detect dysfunction in central and peripheral vision which may be caused by various medical conditions such as glaucoma, stroke, brain tumors or other neurological failures. Visual field testing can be performed clinically by keeping the subject's gaze fixed while presenting objects at various places within their visual field.

Confrontation Visual Field Testing

Confrontation testing (Donder's test) is a relatively simple method that doesn't require any instrumental equipment and relies on comparing the patient's visual fields with the examiner's, and hence the test requires that the examiner has a normal visual field.

To perform the *confrontation visual field testing* follow these steps:

— Sit in front of the patient about 1 meter away with his or her eyes at the same level as yours.

— Test each eye separately. Ask the patient to cover one eye with the palm of the hand. If the patient covers their left eye, cover your own right eye i.e. the contralateral eye, with your hand as well and vice versa.

— Ask the patient to focus on your uncovered eye and not move their eyes anywhere else, otherwise the test is void.
— Move a target object (a pen, fingers) from the periphery (start outside the usual 180° visual field) toward the midline in all four quadrants in the nasal and temporal fields and in the superior and inferior fields. Ask the patient to signal when they first see the object and, as you move towards the center, whether it disappears.
— Repeat the test in each quadrant for the other eye.
— A patient with a normal visual field will see the object at the same time as you; a patient with an abnormal or restricted field of vision will see the object later than you.
— The essential requirement for this test is the normal visual field of the examiner himself.

Confrontation visual field testing may be used as a quick screening test in patients to determine obvious visual field defects, but may not be as sensitive as other tests.

Perimetry

Perimetry is the method for determining the peripheral field of vision by means of a perimeter. There are a number of techniques used in ophthalmologic practice. They fall into one of two categories: kinetic or static perimetry that are either operated manually or computer assisted (automatic).

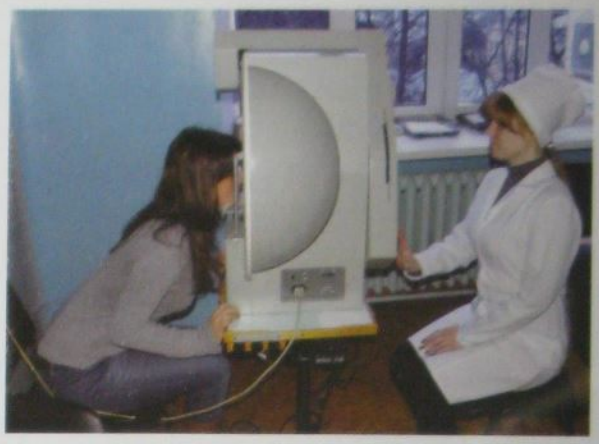
Kinetic perimetry involves detection of moving targets and static perimetry involves detection of a stationary target. Static testing in general is superior to kinetic perimetry in detecting slopes and scotomas (field defects), and tends to be more reliable and consistent, particularly for detecting glaucomatous visual field loss.

Kinetic Perimetry

Kinetic perimetry involves moving of the object from the non-seeing area to the point at which it is first seen. The best known of kinetic perimeters is referred to as the Goldmann perimeter. It is a white hemispheric bowl of even luminance onto which a small bright light stimulus is projected. The light stimulus, whose size and brightness can be varied, is moved manually by the examiner from outside of the bowl towards the center (fig. 2.7).

To perform the *visual field testing with the hemispheric kinetic perimeter* follow these steps:

— Ask the patient to sit facing the perimeter.
— Explain the test to the patient emphasizing the importance of maintaining fixation on the central target and the necessity to inform the examiner about the first instance of seeing the light stimulus.
— Each eye is tested separately so ask the patient to cover one eye with their hand or by using a hand-held occluder.



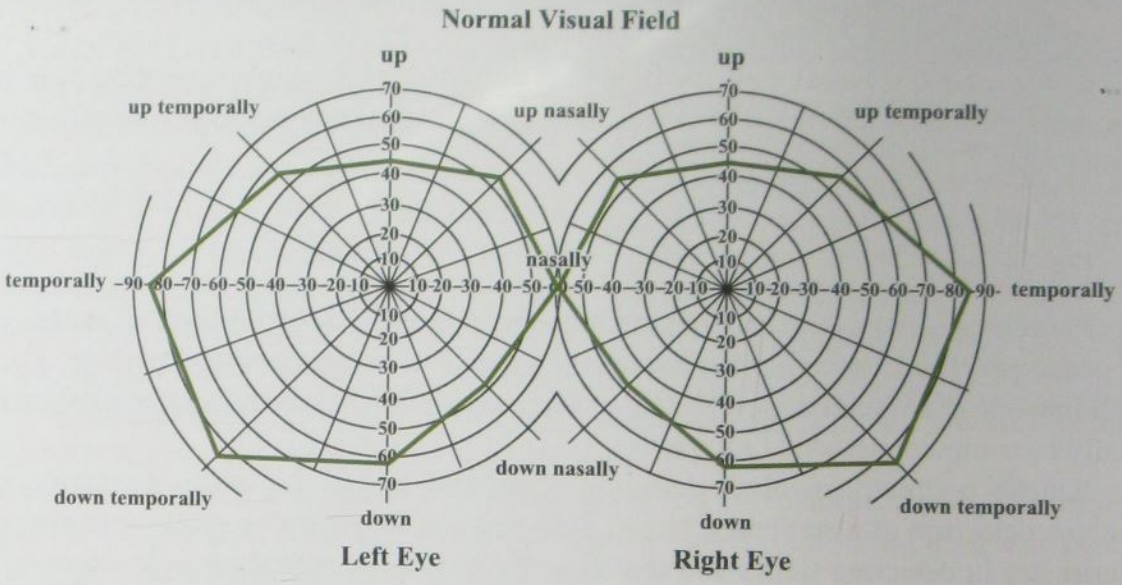


Fig. 2.7. The visual field testing by the hemispheric kinetic perimeter and the chart for the results recording with the normal borders outlined

— Help the patient place their chin onto the chinrest and the forehead against the forehead strap, which allow the head to remain in a steady position during testing.
— Ask the patient to focus on a fixation point at the center of the hemisphere.
— First, a light stimulus of constant size and intensity is used. The light stimulus is presented much like in confrontation testing in the periphery first, and then moved at a constant rate of speed towards the center until the point when it is first detected by the patient.
— Note the location of target detection on a chart and repeat the process for different meridians around the visual field.
— Connect the points of first seeing the stimulus by a line (an isopter) — establish a boundary of vision for that light stimulus and plot the blind spot.
— Repeat the procedure using different test light stimulus (with reduced or increased size and brightness) and create other isopters.

— Ask the patient to remove the occluder to allow the covered eye to adapt to the light before testing the fellow eye.
— Repeat the test for the fellow eye.

Periodically during the test instruct the patient to close his or her eyes and rest, especially when the patient is tired or not responding. You may allow the patient to sit back between isopters and relax.

In this way, kinetic perimetry is useful for mapping visual field sensitivity boundaries. It may be a good alternative for patients that have difficulty with automated perimetry, either due to difficulty maintaining constant gaze, or due to cognitive impairment.

Static Perimetry

Static perimetry uses a stationary light stimulus (constant location), the luminance of which is gradually increased until seen. It is generally done using automated equipment. A few different types of static perimetry tests can be used of which Humphries' is the most common and accepted as gold standard of the automated perimetry (fig. 2.8).

During automated Humphrey visual field testing, a patient is asked to focus on a target and press a button if and when they see a light that shows up in their peripheral visual field. If the patient does not see the light, its intensity is gradually increased

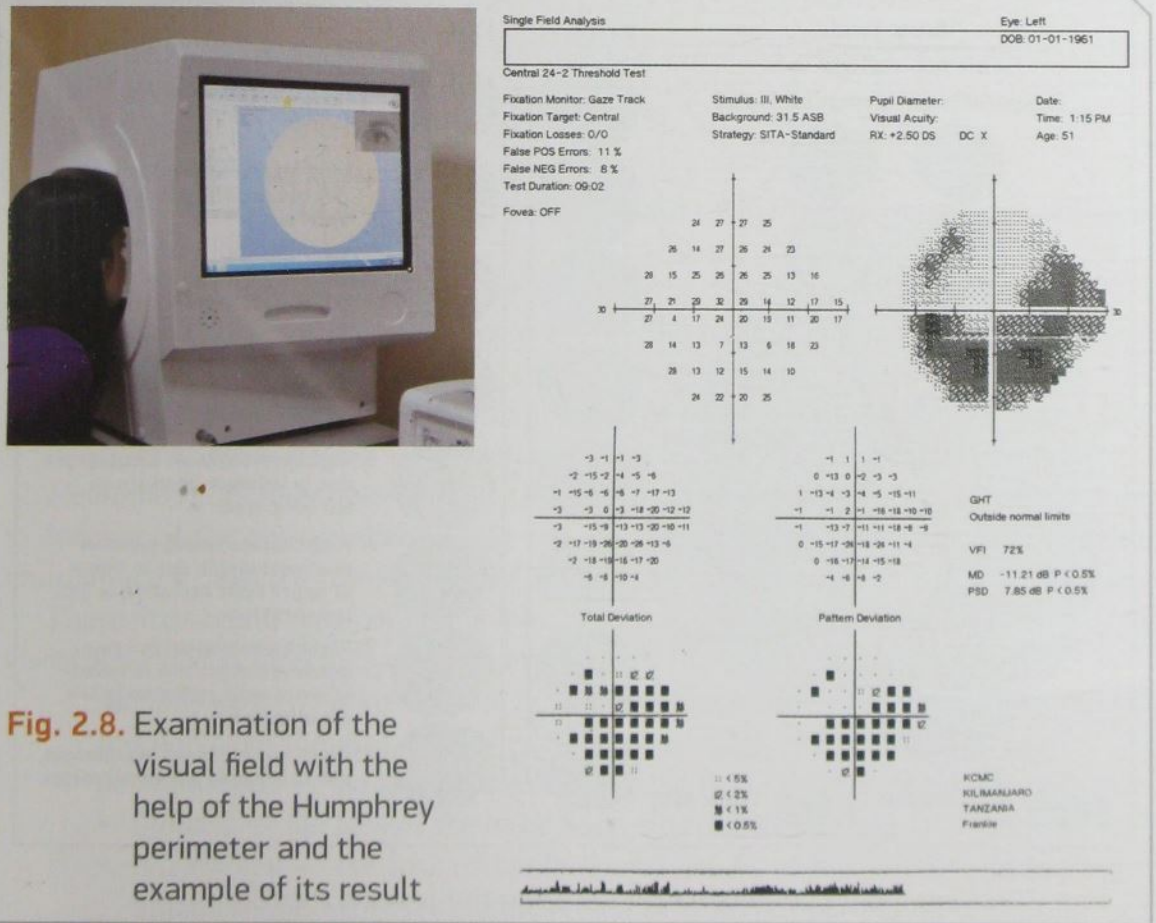


Fig. 2.8. Examination of the visual field with the help of the Humphrey perimeter and the example of its result

until it is seen. The minimum intensity required for the detection of a light stimulus is called the "threshold" sensitivity level of that location. This procedure is then repeated in different locations, until the entire visual field is tested. The perimeter checks each point several times, so if the patient made a mistake the computer is able to correct this. The computer then automatically maps and calculates the patient's visual field.

Humphrey perimetry is used not only for peripheral visual field testing but for assessing the retinal sensitivity. It is used for rapid screening and follow-up of diseases involving deficits such as scotomas, loss of peripheral vision and more subtle vision loss. Perimetry is important in the diagnosis of glaucoma and as a means of determining progression of glaucomatous damage, and can also be helpful in diagnosing damage from strokes, multiple sclerosis, and other neurological conditions.

Visual field defects may be a sign of lesions of the retina, optic nerve and optic pathways damage, and may occur as the result of many eye diseases or trauma and a stroke or brain injury or tumors. The location of visual field loss indicates the location of the problem (fig. 2.9). Unilateral field loss in the lower nasal field suggests an upper retinal lesion. Central field loss usually indicates macular problems. A homonymous hemianopia indicates problems in the brain rather than the eye. A visual field defect arching over central vision to the blind spot (arcuate scotoma) is a sign of glaucoma.

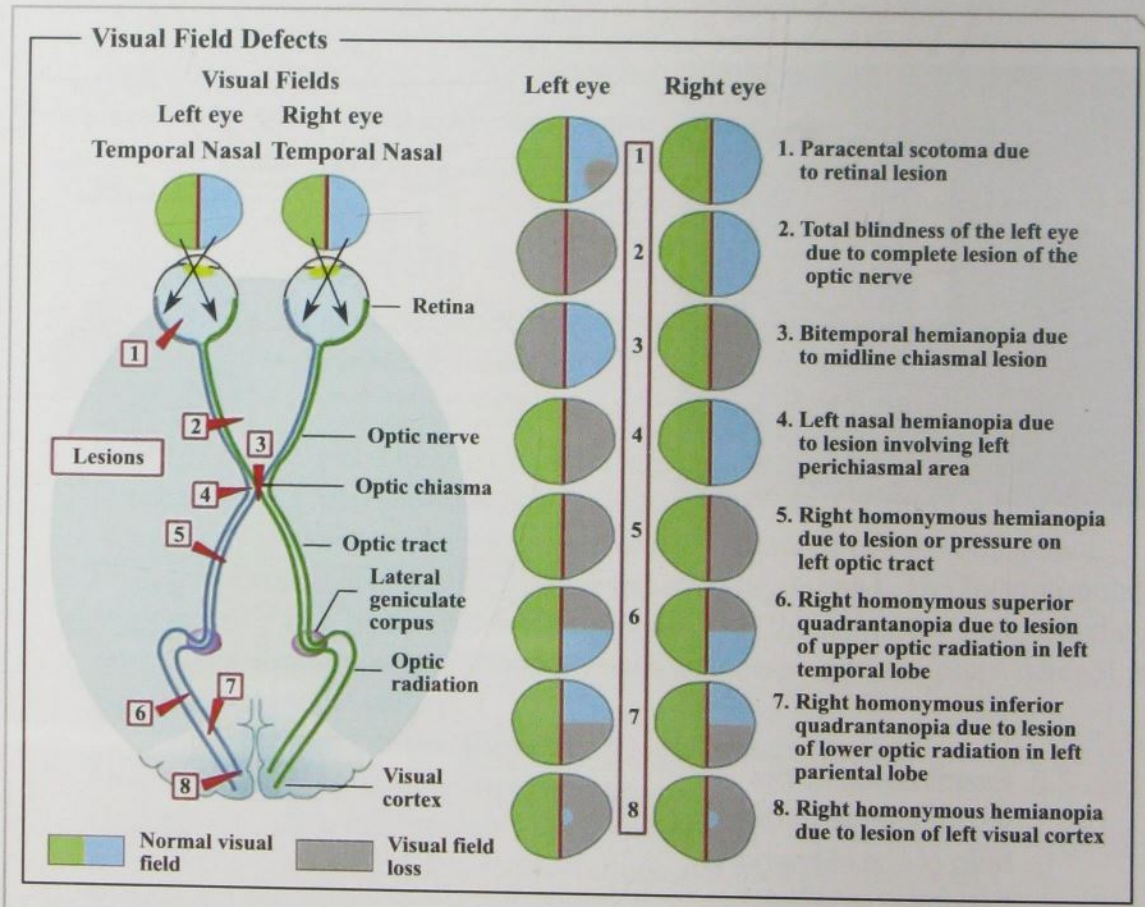


Fig. 2.9. Visual field defects with indicating the lesions location

Common defects of the visual field include scotoma, hemianopia and quadrantanopia. In most cases, field defects are consistent; however, there are times when scotomas can be temporary or even occur from time to time. This is the case that many people experience when they have a migraine headache.

The term *scotoma* is derived from the Greek word for darkness. It is an area of loss of or decreased vision within the visual field.

Every normal mammalian eye has a small scotoma in its field of vision, usually termed as a blind spot. *Blind spot* is a small part of the visual field of each eye that corresponds to the position of the optic disk (also known as the optic nerve head) on the retina. There are no photoreceptors (i.e. rods or cones) in the optic disk, and, therefore, there is no image detection in this area. The optic disk is located on the nasal side of the fovea, is oval in shape, and is approximately 1.5 mm in diameter. With both eyes open, the blind spots are not perceived because the visual fields of the two eyes overlap. Indeed, even with one eye closed, the blind spot can be difficult to detect subjectively because of the ability of the brain to "fill in" or ignore the missing portion of the image.

The main *types of scotomas* include:

— by localization:
  - *central* — visual field loss appears in the central vision and is caused by optic nerve disease or macular disorders;
  - *paracentral* — visual field loss near the central region but definitely not the central region;
  - *peripheral* — visual field loss appears in the peripheral region and is caused by chorioretinal lesions;

— by the degree of sensitivity loss:
  - *absolute* — total loss of vision;
  - *relative* — an area of depressed vision in which large objects are still seen blurred;

— by complaints availability:
  - *positive* — scotoma is perceived by the patient (subjective), may indicate opacity of the lens, retina or optic nerve diseases;
  - *negative* — scotoma is a field of defect of which the patient is not aware, a good example is glaucomatous defects which can only be detected by perimetry.

Hemianopia is blindness or reduction in vision in one half of the visual field due to damage of the optic pathways in the brain. This damage can result from acquired brain injuries caused by stroke, tumor or trauma.

There are several types of hemianopia, each named depending on the region of the visual field that is affected, as well as the size of the affected area:

— *homonymous* — a visual field defect involving either the two right or the two left halves of the visual fields of both eyes;

— *heteronymous* — the loss of half of the visual field on different sides in both eyes:
  - *binasal* — the loss of the nasal halves of the visual field;
  - *bitemporal* — the loss of the temporal halves of the visual field leading to tunnel vision, which is characterized by a loss of peripheral vision.

Homonymous hemianopias indicate a lesion beyond the chiasm. A bitemporal hemianopia is characteristic of a lesion in the area of the chiasm. A binasal hemianopia is very rare and appears at the suppression of the chiasm from both sides.

Quadrantanopia is low vision or blindness in one quarter of the visual field. It is often homonymous; that is, low vision or blindness occurs in the same quadrant of each visual field.

1.4. Eye Exam

Slit Lamp Examination

Slit lamp examination is the most informative technique of anterior eye structures and ocular adnexa inspection carried out with the help of a biomicroscope. A narrow beam of collimated light that can be varied in width, height, incident angle, orientation and color, is passed over the eye. Often, this light beam is narrowed into a vertical "slit" during slit-lamp examination (fig. 2.10). The examiner views the illuminated ocular structures through an optical system that magnifies the image of the eye (can be chosen from eight- to forty-power). This allows inspecting all the ocular media, from the cornea to the vitreous, plus magnified view of the eyelids, and other external ocular related structures. The binocular slit-lamp examination provides stereoscopic magnified view of the eye structures in striking detail, enabling exact anatomical diagnoses to be made for a variety of eye conditions.

The light source and viewing angle can be adjusted relative to each other to allow a view of different parts of the anterior segment that would not otherwise be visible. Various illumination techniques can be used in slit-lamp examination of the eye.

Direct focal illumination or examination with an optical section is the most frequently applied method of slit-lamp examination. It uses a very narrow beam (about 0.1 mm to 0.2 mm) directed from 45 degrees. This angle and slit lamp position send the beam past the pupil margin and through the lens so that there is no reflection from the internal surface. This technique allows a view of the corneal surface. Examination with a slightly wider beam than the direct focal is used to view a cross section of the cornea and endothelium. When the slit lamp is moved peripherally and the focus is shifted posteriorly, the iris is clearly visible. In this case, the slit beam is directed at the iris, not past it, in what is called tangential illumination.

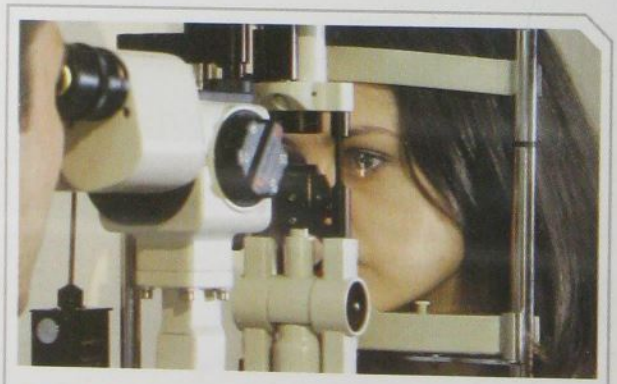


Fig. 2.10. Slit lamp examination if the eye

Diffuse illumination is a method for observing the eye and adnexa in general. For this, the light beam is opened very wide and diffused by inserting a ground glass screen or diffuser in the illuminating path. This method is used when ocular media, especially that of the cornea, are opaque. In these cases direct focal illumination is often impossible depending on severity. Its main purpose is to illuminate as much of the eye and its adnexa as possible at once for general observation.

Indirect illumination means looking at tissue outside the area which is directly illuminated and can be used in conjunction with the above mentioned techniques. Corneal opacities, corneal nerves and limbal vessels are easily seen under indirect illumination as glare is reduced.

Retroillumination is another form of indirect viewing. The light is moved to a position directly in front of the microscope so the beam is aimed through the pupil and lens to the retina. The light reflected back to the viewer from the retina is used to visualize certain structures or abnormalities of the lens, iris, or cornea.

Scleral scatter illumination uses the principle of total internal reflection and is used to visualize abnormalities of the cornea that are not visible with direct illumination. A narrow beam is directed at the temporal or nasal limbus and reflected within the cornea but scattered by the sclera. Corneal opacities, edema or foreign bodies will be made visible by the scattering light, appearing as bright patches against the dark background of the iris and pupil. Even tiny nebular opacities can be seen.

To perform the slit lamp exam the examiner must use a systematic "front-to-back" approach, i.e. beginning with the eyelids and lacrimal system and moving progressively posterior to examine the conjunctiva, sclera, cornea, anterior chamber, iris, lens, and anterior vitreous cavity.

To perform *the slit lamp exam,* follow these steps:

— Ask the patient to place their chin on the chinrest and the forehead against the forehead rest of the slit lamp.
— Adjust table height for your own comfort and that of the patient.
— Ask the patient to look at your right ear when examining the right eye and vice versa.
— Adjust the width of the beam from full (circular) to a thin slit, apply magnification (usually beginning at low power and progressing to higher power if needed).
— During the examination choose the appropriate method of illumination.
— Start examination from the lids to the back of the eye in the following order:

Eyelids

Examine both the upper and lower eyelids. Inspect the lid margins for lacerations, eyelid folding, abnormal lash growth toward the cornea, lash loss, for Meibomian gland dysfunction and signs of blepharitis (eyelid inflammation), for lumps or bumps.

Lacrimal system

Lacrimal system examination begins with inspection of the eyelids as mentioned above. Check for whether entropion and ectropion are present, as these interfere with

drainage through the puncta (minute orifices from which the canaliculi begin). Entropion particularly poses a problem as the eyelid is everted, causing the lower punctum to be turned away from the ocular surface and hence lead to epiphora (overflow of tears). Examine the punctum under the slit lamp to check for any stenosis caused by scarring or papillomas. The medial canthus should be examined in detail for any unusual erythematous swellings and discharge (dacrocystitis).

Conjunctiva

Evaluate the conjunctiva for discharge, follicles, fluid accumulation (chemosis), and hyperemia. Be sure to examine both the upper and lower fornices in both eyes. Assess the color of the conjunctiva. Check for any follicles or papillae. Check if the conjunctival blood vessels aren't injected (red and inflamed). Be also sure to check for any foreign bodies that may be stuck in the folds of the conjunctiva.

Sclera

Check to make sure the sclera is white and non-icteric.

Cornea

The cornea is examined for transparency and any abnormalities of shape or thickness. Check the epithelium for abrasions, edema, ulcers, and foreign bodies. The layers are then examined from epithelium to endothelium with appropriate illumination techniques.

Anterior Chamber

Look for inflammatory cells and flare (light scatter caused by inflammatory proteins in the aqueous humor), which could indicate inflammation or intraocular bleeding. Examine the inferior sector of the anterior chamber for the presence of hyphema (blood) or hypopyon (pus). Also, comment if the anterior chamber is deep and well-formed or shallow and thus a setup for angle-occlusion glaucoma.

Iris

Make sure the iris looks flat and the pupil round. Assess the iris difference in color and any lesions or structural abnormalities. Note how the pupil responds to the light.

Lens

Look for abnormalities in the shape of the lens (e.g. posterior lenticonus), its transparency (cataract) and localization (dislocated or sublaxated). The type and position of any lens opacity are next observed using both direct and retinal retroillumination. Determine the type of cataract present. If the patient has an implant intraocular lens following cataract surgery, determine the clarity of the posterior capsule. Abnormal movements of the lens (phakodonesis) and iris (iridodenesis) should also be recorded.

The lens is best viewed through a dilated pupil.

Vitreous

The anterior vitreous is the last part of the eye that is readily visible without the aid of special lenses. Look for the presence of cells, blood, deposits, calcification or pigment granules and condensations of the vitreous gel.

Tonometry

The pressure within the eye, the intraocular pressure (IOP), is based on the balance between production and drainage of aqueous humor mainly through the trabecular meshwork located in the anterior chamber angle. A balanced intraocular pressure is required to keep the eye in shape, provide nutrients to intraocular structures and maintain normal function of these structures. Increased intraocular pressure occurs when there is a blockage in aqueous humor drainage from the eye. Decreased intraocular pressure is the result of either reduced production, or in case of injury, leakage through a defect in the globe wall.

Tonometry is a measurement of intraocular pressure (IOP) by determining the amount of force needed to slightly temporarily flatten a predetermined area of the corneal surface. It is the fundamental screening test for detecting elevated intraocular pressure and essential for glaucoma screening.

IOP may be measured by several different methods.

Palpation

Palpation (also known as digital tonometry) is the method of IOP estimation by gently pressing the index fingers against the cornea of a closed eye (fig. 2.11).

To perform *the IOP estimation by palpation,* follow these steps:

— Ask the patient to close the eyes and look down.
— Place your index fingers of both hands on the top of the upper eyelid.
— Press the eyeball alternately with one or another finger to feel fluctuation.
— Repeat the test on the contralateral eye for comparison.

This method is not highly accurate as it provides only a gross estimation of IOP, but it allows to differentiate between very low and very high eye pressure and to compare the IOP level in both eyes. However, an experienced ophthalmologist may become quite capable of estimating intraocular pressure to within 2—3 mm Hg.

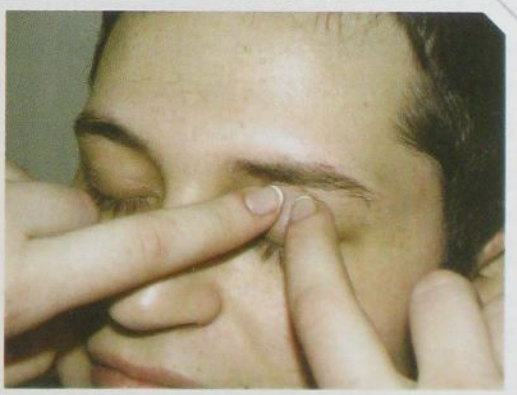


Fig. 2.11. Estimation of IOP by palpation

NOTE!

Palpation of the eye should never be performed in recently operated eyes or in cases of a suspected ruptured globe.

If IOP is increased, it is marked as the letter "T" with the sign "+", if IOP is reduced — with the sign "—". Four levels of eye density degree are distinguished. They are T_{+1} — mildly increased, the eye is firm; T_{+2} — highly increased, the eye is very firm; T_{+3} — the eye is as solid as a rock; T_n — IOP level is normal. For example, the "rock hard" eyeball only occurs in acute angle closure glaucoma.

When IOP is reduced three levels of hypotension are distinguished: T_{-1} — the eye is slightly softer; T_{-2} — the eye is soft; T_{-3} — the eye is very soft, the fingertips do not feel any resistance of the eye.

Applanation Tonometry

Applanation tonometry* is the method of IOP measurement that requires special equipment and refers to mechanical flattening (applanation) of a constant area of the cornea. The tonometer device lightly touches the cornea, ever so slightly flattening it. The resistance to flattening is measured. Several types of tonometers are available for this test, the most common applanation tonometers are Maklakov, Goldmann, Perkins, non-contact and others.

Maklakov Tonometry

Maklakov tonometry was widely used in Europe throughout the last century and is still popular in Ukraine and other Eastern European countries.

The Maklakov tonometer applies a fixed weight on the cornea, determines the flattened area and then calculates the IOP from the size of the area applanated. The tonometer consists of a special wire fork holder into which a weight is inserted. The weight is a four-centimetre hollow metal cylinder, ranging from 5 to 15 g, with both ends being wide and covered with mat glass of one centimetre in diameter. The surface of the weight is painted with a special ink and then a weight is placed onto the anesthetized cornea. The weight is lifted from the cornea, and the area of applanation is taken to be the area of missing ink. The tonometer is then to be "printed" onto a piece of paper (fig. 2.12). The area in the center of the imprint that is devoid of ink is proportional to the IOP**. Intraocular pressure in healthy humans measured by Maklakov tonometry is 18—26 mm Hg.

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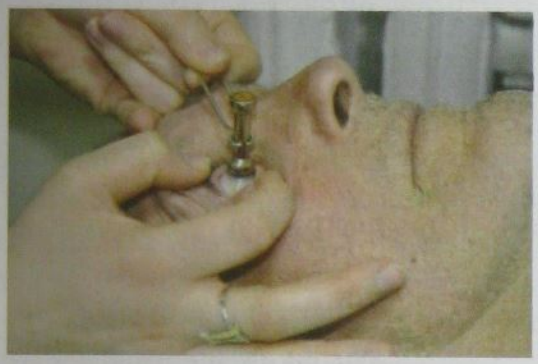
* Applanation tonometry is based on the Imbert—Fick principle, which states that the pressure (P) inside an ideal dry, thin-walled sphere equals the force (F) necessary to flatten its surface divided by the area of the flattening (A): $P = F / A$.

Cornea being aspherical, wet and slightly inflexible fails to follow the law. So according to it, modified Imbert-Fick Law is used that included factors to consider the resistance of the cornea to applanation and the surface tension of the tear meniscus surrounding the tonometer prism during measurement: $P = (F + T - C) / A1$, where P — intraocular pressure, F — tonometer force, T — force of tear film meniscus surface, C — corneal resistance to bending, A1 — inner corneal area of applanation. When flatten area has a diameter of 3.06 mm T and C forces cancel each other. Grams of force applied to flatten 3.06 mm diameter of the cornea multiplied by 10 is directly converted to mmHg.

** IOP is inferred from the weight (W) and the diameter of the area of applanation (d) by using the following formula: $P = W / p (d/2)^2$. IOP is measured in grams per square centimetre and is converted to millimetres of mercury by dividing by 1.36.



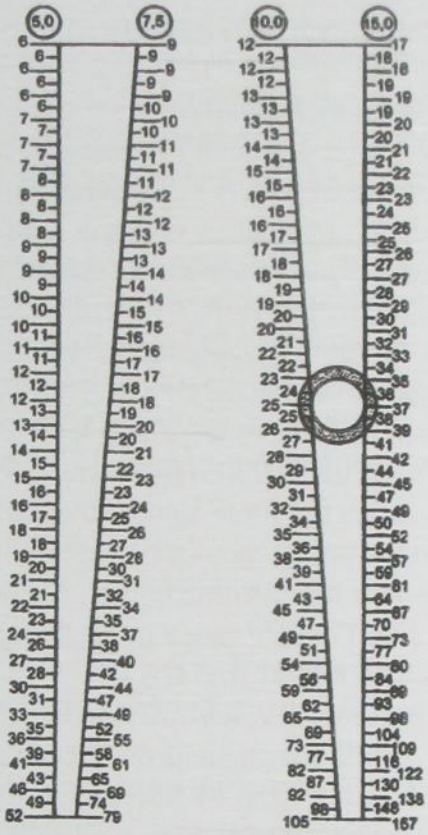


Fig. 2.12. Performing the IOP measuring with Maklakov tonometer and registration of its results with the help of Poliak's ruler

To perform *the measurement of IOP with Maklakov tonometer* follow these steps:

— Sterilize the tonometer surfaces with alcohol and dry with a sterile cotton wool.
— Lubricate tonometer planes with a special ink (usually a mixture of colloidal silver and glycerin).
— Ask the patient to lie down on his or her back and instill the anesthetic solution (0.25 % Dicaine) three times with intervals between instillation of 2—3 minutes.
— Ask the patient to look straight, so that the tonometer is in the center of the cornea once it's applied.
— Use one hand to open the patient's eyelids and another hand to apply a tonometer on the center of the cornea.
— Press the fork handle downwards to over one-third of the tonometer cylinder.
— The tonometer presses the eye, and flattens the cornea. The ink spreads in the corneal area that touches the tonometer plane. A colorless circle appears on the plane of the tonometer.
— Apply the second lubricated end of the tonometer to the cornea of the contralateral eye.
— Instill disinfectant solution to rinse out the patient's eyes from the ink.
— "Print" both tonometer planes on white alcohol moistened paper. (The smaller the bright disc, the greater intraocular pressure.)

— Use Poliak's ruler to measure the diameter of the disc. The result comes in millimetres of mercury (mm Hg).

Goldmann Tonometry

Goldmann tonometry is considered to be the gold standard for IOP measurement and is the most widely accepted method. The Goldmann tonometer measures the force required to applanate a constant area of the cornea (over a circular area of diameter 3.06 mm). This tonometer in which a variable force is used to applanate a constant area of the cornea differs from the Maklakov constant force tonometer.

The Goldmann applanation tonometer is mounted on the slit lamp and consists of a strain gauge connected by a lever to a plastic tip. When the tip face contacts the cornea (after application of anesthetic eye drops and fluorescein), a biprism in the tip splits the view of the tear film meniscus into two semicircles. Rotating a dial attached to the gauge varies the force against the cornea and alters the alignment of the two semicircles. At the point when inner edges of the semicircles meet (it means that a cornea area of 3.06 mm has been flattened), the dial indicates pressure in millimetres of mercury (fig. 2.13). The IOP range in healthy population measured by Goldmann tonometry is between 10 and 21 mm Hg.

To perform the *measurement of IOP by Goldmann tonometry,* follow these steps:

— Instill anesthetic drop in each patient's eye.
— Instill a drop of fluorescein dye in each eye or touch tear layer on the inner surface of each lower lid with a fluorescein strip wetted with sterile saline.
— Clean the tonometer tip with sterilizing solution.
— Set the tension knob at 1 g and the graduation mark of the prism at "0". Set the magnification of the slit lamp at ×10.
— Ensure that the patient is sitting comfortably at the slit lamp: at the right height, with their chin on the rest and their forehead against the headband.
— Move the filters so that the cobalt blue filter is used to produce a blue beam.
— Make sure the beam of light is as wide as possible, and that the light is as bright as possible. This makes visualizing the fluorescein rings easier.

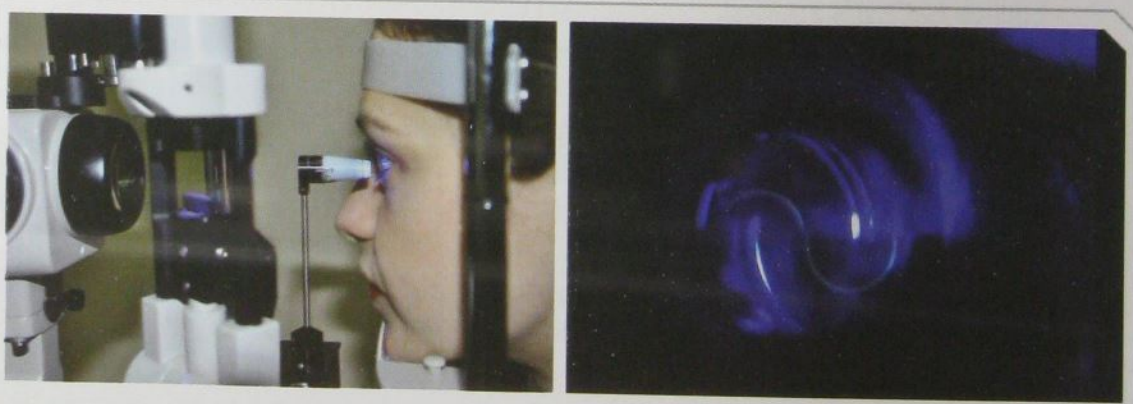


Fig. 2.13. IOP measurement by Goldmann tonometry and its visualisation

— Ask the patient to look straight ahead, open both eyes wide, fix his or her gaze and keep perfectly still.
— Hold gently the patient's eyelids open, taking care not to put any pressure on the globe while holding the lids. Even negligible pressure from the examiner's fingers resting on the globe can cause a significant increase in IOP.
— Direct the blue light from the slit lamp onto the prism head. Make sure that the tonometer head is perpendicular to the eye.
— Move the tonometer forward slowly until the prism rests gently on the center of the patient's cornea. Because of the prismatic effect of the tonometer tip two green semi-circles are seen when the tonometer tip is fully applanated.
— With the other hand, turn the calibrated dial on the tonometer clockwise until the two fluorescein semi-circles in the prism head are seen to meet and form a horizontal 'S' shape. (Note: the correct end point is when the inner edges of the two fluorescein semi-circle images just touch).
— Note the reading on the dial and record it in the notes.
— Withdraw the prism from the corneal surface and wipe its tip.
— Repeat the procedure for the other eye.
— Instill disinfectant solution to rinse out the patient's eyes from the fluorescein.
— Wipe the prism with a clean, dry swab and replace it in the receptacle containing the disinfectant.
— Multiply the reading obtained in grams of force from the rotating drum by 10 to convert to millimetres of mercury (mm Hg).

Perkins Tonometer

The Perkins tonometer works on the same principle as the Goldmann tonometer, except that it is a portable, handheld and counterbalanced device, so it can be used in any position (vertical or horizontal). The light comes from batteries and force comes from a spring, varied manually by the operator. It is useful in a number of situations, including the operating room, at the bedside, with patients who can't be examined at a slit lamp and small children.

Non-contact Tonometry

This type of applanation tonometry (sometimes also called pneumatonometry or air-puff tonometry) is fast, simple, and does not require eye drops or contact with the eye, which makes it easier to use. It uses a rapid air pulse to applanate the cornea. Intraocular pressure is estimated by detecting the force of the air jet at the moment when the central cornea is flattened. There are many types of non-contact tonometers based on this principle that can be table-mounted or handled and portable. The normal intraocular pressure measured this way is between 10 to 21 mm Hg.

When you *test IOP with the non-contact tonometer,* follow these steps:

— Ask the patient to sit and place their chin on the chinrest and the forehead against the forehead rest.

— Align the patient's eyes with the eye level marker by turning the chinrest elevation knob.
— Pressing the safety stopper set the determined space (7—8 mm) between the patient's eye and air nozzle. Be sure that air nozzle doesn't touch and scratch cornea.
— Ask the patient to look straight ahead, open both eyes wide, fix his or her gaze on the fixation light (green LED) and keep perfectly still.
— Use the joystick to get a clear patient's eye image in the center of the screen and adjust focusing on the eye to be measured.
— When the focusing indicator maintains proper alignment air is puffed out automatically and measurement starts. The measurement data appears on the screen.
— A puff of air is directed at the open eye over a very short time (measured in milliseconds) until the cornea is flattened. When the cornea is flattened, a light beam is reflected into a sensor which stops the generation of air and records the force at the moment of applanation. The intraocular pressure is measured through the force of the jet of air.
— Repeat measurement three or more times to receive the average value of the IOP (IOP fluctuates with the patient's pulse and breath).
— Measure the IOP of the other eye in the same way and print out the measured data.

Although not considered to be the most accurate way to measure IOP, it is often used as a simple way to screen for high IOP. It is an easy way to test children. Because non-contact tonometry is accomplished without the instrument contacting the cornea the potential for disease transmission is reduced.

There are many other tonometers designed for IOP measurement which are handheld devices shaped like a pencil, e.g. Tono-Pen, Diaton tonometer, etc.

Ophthalmoscopy

Ophthalmoscopy is the method of examination of the back part of the eye (fundus), which includes the retina, optic disc and blood vessels, through the pupil by means of the ophthalmoscope. It is also called funduscopy or fundoscopy.

Ophthalmoscopy is the only way to directly visualize veins and arteries in their natural state non-invasively (in vivo). Anomalies in the appearance of the fundus may indicate both eye diseases, as well as systemic and neurological conditions, so it is an essential method to avoid missing many serious ocular and general disorders.

The key to successful fundus examination is systematic approach, starting with red reflex assessment and then proceeding to the optic disc and work radially:

— Red reflex — note is it bright or obscured;
— Optic nerve head — inspect its size, shape, color, margins and cup to disc ratio (C / D);
— Blood vessels — evaluate their obstruction, caliber, shape, curvature, color, crossings, light reflex and artery to vein ratio (A / V) after 2nd bifurcation;
— Macula — examine its color, light reflex;
— Peripheral fundus — inspect for color, hemorrhages, exudates, tears or holes.

Ophthalmoscopy is best done when the pupil has been dilated for a more complete view of the entire retina. A limited view can be obtained through an undilated pupil, in which case best results are obtained with the room darkened and the patient looking towards the far corner.

NOTE!

Dilate the patient's pupils only after assessing VA and visual field as it causes blurred vision.

Patients with angle-closure glaucoma should not have their pupil dilated as it can cause an acute glaucoma attack due to blockage of the anterior chamber angle.

Red Reflex and the Lens or Lens Examination by Trans Illumination

The method of trans illumination is used for examination of transparent optical media of the eye and mainly for the lens and vitreous body exam as the cornea is examined by side illumination. The examination is held with the help of a direct ophthalmoscope. The light beam directed to the pupil passing through the transparent optic media reflects from the fundus causing the pupil to shine with red color — the red reflex. This red reflex is best assessed from a distance of about 50 cm from the patient's eye and with the pupils dilated. If the red reflex is either absent or diminished, this indicates opacity between the cornea and retina. Opacities in the media appear as black silhouettes.

NOTE!

White pupillary reflex in an infant may indicate a retinoblastoma.

Opacities in the vitreous body are movable, they move even when the eye is fixed. Opacities in the lens move only with eye movements. To detect the location of the opacities the patient should be asked to look upwards and then downwards. If the opacities are located in the anterior lens layers, under trans illumination they will move in the same direction as the eye, if they are located in the posterior lens layers — in the opposite direction.

To *examine the optical media of the eye for the presence of any opacity* follow these steps:

— Turn the ophthalmoscope on to a low-moderate light intensity.
— Set the ophthalmoscope on the "0" lens. You may need to adjust this to compensate for your own or the patient's refractive error.
— Have the patient comfortably seated.
— Tell the patient to blink and breathe normally.
— Use your right eye to examine the patient's right eye holding the ophthalmoscope in your right hand. Instruct the patient to look to a distant object on their left to prevent constriction of the pupils from accommodation.
— Direct light into the patient's eye and look through the ophthalmoscope into it from a distance ("distant" direct ophthalmoscopy) until the red reflex is elicited.
— Check if any signs of opacities are present.

Direct Ophthalmoscopy

Direct ophthalmoscopy is examination of the ocular fundus with a hand-held ophthalmoscope giving a direct magnified (approximately 15 times) image of the fundus. The basic principle of this type of examination is that the light travels in a straight, direct path from the patient's eye to the examiner. Direct ophthalmoscopy, as the name suggests, allows the practitioner to view the fundus of a patient directly (fig. 2.14).

To *perform direct ophthalmoscopy*, follow these steps:

— Turn the ophthalmoscope on to a low-moderate light intensity.
— Set the ophthalmoscope on the "0" lens. You may need to adjust this to compensate for your own or the patient's refractive error.
— Have the patient comfortably seated.
— Ask the patient to focus gaze on a distant object to prevent constriction of the pupils from accommodation. Tell the patient to blink and breathe normally.
— Use your right eye to examine the patient's right eye holding the ophthalmoscope in your right hand. Instruct the patient to look to a distant object on their left.
— Direct light into the patient's eye and look through the ophthalmoscope into it from a distance ("distant" direct ophthalmoscopy) until the *red reflex* is elicited.

This red reflex is the reflection from the fundus and is best assessed from a distance of about 50 cm. If the red reflex is either absent or diminished, this indicates opacity between the cornea and retina. The most common opacity is a cataract. Opacities in the media appear as black silhouettes.

— Slowly move closer to the patient following the red reflex and at the same time gradually increase the power of the lens to focus on the retina.
— Locate *the optic disc.*

When the patient is looking straight ahead, the optic disc should naturally come into the field of view. If not, try to locate a blood vessel on the retina and then move along it and locate the point at which it branches. Move your field of view in the direction in which the apex of the branch is pointing. By moving along a blood vessel in this manner the optic disc will be located.

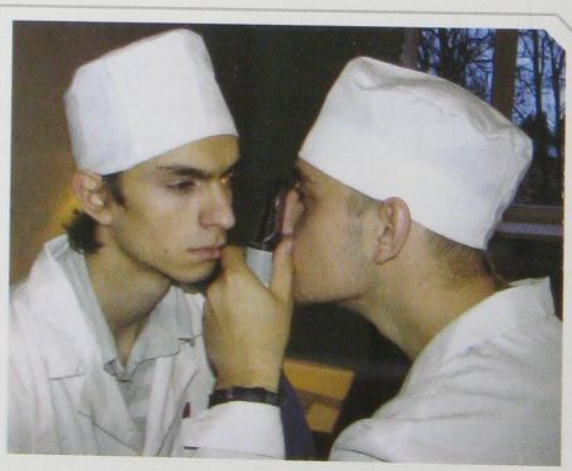


Fig. 2.14. Examination of the fundus by direct ophthalmoscopy

— Examining the optic disc consider its color (normally it is pink), clear definition of its margins, cup (if there is one) and the ratio of the size of the cup to the size of the optic disc (cup disc ratio, denoted as, e.g., 0.3:1, meaning it occupies one-third of the area of the optic disc).

— Note the capillaries on the optic disc and look for the presence of a spontaneous venous pulsation (a spontaneous venous pulse is normal; an arterial pulse is abnormal). Also note the presence of any pigment, choroidal or scleral crescents around the disc.

— Evaluate *the retinal blood vessels*.

The retinal blood vessels should be examined by following the temporal and nasal arcades from the optic disc. Veins are larger and dark red, whereas arteries are relatively thinner and lighter (normal artery : vein ratio is 2:3).

— Focus on and examine *the macula*.

The macula is visualized by asking the patient to look at the light source as this brings the fovea (fixation point) into view. The macula is the area between the superior and inferior temporal arcades and its center is the fovea. Since using an excessively bright light can make the macula difficult to visualize, it may be useful to use a smaller aperture beam and minimal required intensity.

— Examine *the peripheral fundus*.

Ask the patient to look in different directions to allow you to view the peripheral fundus. You will need to adjust the lens in the wheel slightly as the periphery is closer to you than the optic disc, requiring more focusing power.

— Repeat the process to examine the patient's left eye. Hold the ophthalmoscope in your left hand; look through it with your left eye to examine the patient's left eye. Instruct the patient to look to a distant object on their right.

Indirect Ophthalmoscopy

The principle of indirect ophthalmoscopy is to make the eye highly myopic by placing a strong convex lens in front of the patient's eye so that the emergent rays from the fundus are brought to focus as a real inverted image in the air between the lens and the observer' eye. So, indirect ophthalmoscopy requires the observer to view an image of the fundus that has been projected to a point in space.

The image formed in indirect ophthalmoscopy is real, inverted and magnified. Magnification of the image (from two to five times magnification) depends on dioptric power of the convex lens, position of the lens in relation to the patient's eye and its refractive state. The usual powers used are +20 D and +13 D. About 5 times magnification is obtained with a +13 D lens. With a stronger lens, the image will be smaller but brighter and the field of view will be wider.

NOTE!

The indirect ophthalmoscope gives an inverted image of the fundus. The image is upside down and reversed right for left.

Compared to the direct ophthalmoscopy the indirect one gives a wider view of the fundus all the way to the ora serrata, so it is invaluable in diagnosing disorders in the periphery of the retina, but this advantage is at the expense of decreased magnification.

There are several indirect ophthalmoscopy techniques — monocular, modified monocular, binocular, slit-lamp, head mounted and others. Indirect ophthalmoscopy is carried out in a dark room with fully dilated pupils.

To perform *modified monocular indirect ophthalmoscopy* the direct ophthalmoscope can be used in conjunction with a +20 D condensing lens. This combination provides a moderately magnified and wider angle view of the fundus. This avoids the close proximity between the patient and examiner required when using a direct ophthalmoscope alone.

To *perform indirect ophthalmoscopy*, follow these steps:

— Remove any patient spectacle correction (the examiner should keep on any refractive correction).
— Instill mydriatic eye drops in each patient's eye to receive the maximal pupil dilatation (this will usually take 20 to 30 minutes following drug instillation).
— Have the patient comfortably seated.
— Turn on the table lamp (or other light source) located on the left and behind the patient.
— Ask the patient to focus on a distant object straight ahead.
— Direct light beam from the table lamp into the patient's pupil by the ophthalmoscope, held approximately 18 cm from the patient's eye, and looking through it visualize the red reflex.
— Place a +20 D lens 3—5 cm in front of the patient's eye in the path of the ophthalmoscope light beam, and then move slightly toward or away from the patient until a clear image of the retina is observed.

A +20 D lens gives a general view of the fundus, for more detailed and magnified examination use a +13 D lens.

— An inverted, aerial image of the retina is produced, located between the observer and the lens.
— Examine the fundus by the above-stated order — state of the red reflex, the optic disc, blood vessels, and macula.
— Ask the patient to look in extreme gaze and examine the whole peripheral retina up to ora serrata.
— Repeat the process to examine the patient's left eye.

Slit Lamp Ophthalmoscopy

Indirect ophthalmoscopy can be performed with the help of a slit lamp and handheld (non-contact) lens. This is the most common method of examination of the fundus, dilated or undilated, at the ophthalmology clinic. The condensing lenses used in slit lamp indirect ophthalmoscopy are high-plus lenses (60 D, 90 D). When used with a slit lamp they produce a virtual image of the fundus that is laterally reversed and inverted. The slit lamp's optical system also allows additional magnification of the image.

To *perform slit lamp ophthalmoscopy*, follow these steps:

— Ask the patient to sit upright on the exam chair in front of the slit lamp.
— Ask the patient to place their chin on the chinrest and the forehead against the forehead rest of the slit lamp to keep their head steady.
— Adjust table height for your own comfort and that of the patient.
— Instruct the patient to fixate straight ahead, to stare wide and to blink normally.
— Keep the intensity of the light beam to the possible minimum and the magnification preferably set at 10× initially.
— Narrow the beam down to 1 to 2 mm width with a height corresponding to the vertical height of the dilated pupil.
— Hold the lens vertically between the thumb and index finger of the left hand to examine the patient's right eye. Hold the lens in the right hand to examine the patient's left eye.
— Place a magnification lens in front of the patient's eye directly in front of the cornea so that the back surface just clears the lashes (approximately 11 mm for the +60 D, 7 mm for the +78 D, and 6.5 mm for the +90 D from the patient's cornea).
— Direct the light beam form the slit lamp to the patient's pupil through the lens.
— Move the slit lamp gradually forward and back until it comes into focus with the aerial image of the fundus between the condensing lens and the slit lamp.
— As with indirect ophthalmoscopy, the image from a non-contact lens slit lamp biomicroscopic examination is inverted and laterally reversed.
— Once the retinal image is focused, widen the beam to observe a greater area of the fundus. The magnification may also be changed to a higher setting at this time.
— To reduce interfering reflections, tilt the lens or increase the setting of the illumination arm to 10 degrees either nasal or temporal once the fundus has been located and focused.
— To view finer fundus details, temporally increase the lamp intensity and magnification.
— Examine the fundus following the above-mentioned approach starting at the optic disc, then move nasally to the disc and cross the macula.
— Asking the patient to look in different positions of the gaze, examine all parts of the fundus; the lens should be tilted to improve definition. Remember the view is laterally and vertically reversed (when the patient looks up, you can view the superior retina).
— Repeat the process to examine the patient's left eye.

2. Additional Methods of Examination

2.1. Exophthalmometry

Exophthalmometry is a measurement of the amount of forward protrusion of the eyeballs, which is performed by means of an exophthalmometer. It gives the quantitative assessment of the position of the eyeball in the orbit. The normal extent of protrusion of the eyeballs from the orbits is 17—19 mm, the normal difference in protrusion between both eyes is 1.5—2 mm. Displacement of the eyeball can be of two types: excessive forward protrusion — exophthalmos, and other way, retraction of the eye into the orbit or backward displacement — enophthalmos. This measurement is important in the cases of thyroid ophthalmopathy, fracture of the orbit floor, retroocular and orbital tumors.

The most commonly used instrument is the Hertel exophthalmometer, in which the position of the anterior corneal surface is recorded, taking the lateral orbital rim as a reference point. This instrument consists of a horizontal calibrated bar with movable carriers at both sides. Each carrier contains mirrors inclined at 45 degrees to reflect both the scale reading and apex of the cornea profile. The examiner can see the image of the apex of the cornea in this mirror. The apex of the cornea lines up with the scale, and the distance is measured in millimetres.

The protrusion of the corneal apex can also be measured with a plastic ruler placed on the lateral orbital margin but this is not very accurate.

To *perform exophthalmometry*, follow these steps:

— Sit opposite the patient in such away that your eyes bare at the same level as the patient's.
— Hold the exophthalmometer with both hands and place it with the index points at the patient's temporal orbital margins.
— Ask the patient to look straight ahead with the eyelids wide open.
— Keep the patient relaxed and avoiding breath holding.
— Superimpose the apex of the cornea on the millimetre scale reading by the inclined mirrors.
— Record the measurement of each eye, using your right eye to measure the patient's left eye and vice versa.

2.2. Upper Lid Eversion

Upper lid eversion is sometimes required to search for conjunctival foreign bodies or other conjunctival signs.

To perform eyelid eversion, the patient is asked to look down with both eyes open and should be repeatedly told to relax and to avoid tightly shutting the opposite eye. This relaxes the levator palpebrae superioris and orbicularis oculi muscles. The examiner grasps the upper eyelashes between the thumb and the index finger, pulls the upper lid away from the globe, and uses a cottontipped applicator to press gently the region over the superior tarsal plate inferiorly and then evert (lift upward) the upper eyelid using the applicator as a fulcrum. Eversion should be performed with a quick levering motion while applying slight traction. Press the lid margin against the orbit rim while the patient is encouraged to keep looking down. The palpebral conjunctiva can then be inspected and cleaned if necessary. To return the lid to its normal position, the examiner releases the lid margin and the patient is instructed to look up and blink.

2.3. Tear / Lacrimal Function Test

There are numerous and varied techniques, which continue to expand and develop, particularly in clinical research, for assessing the tear film. Tear film evaluation can be divided into two areas — assessing tear volume or quantity, and assessing tear stability or quality.

Tear Volume Test

The tear quantity *Schirmer test* is widely used in clinical practice for assessing tear production. The Schirmer test is performed with the help of a 5 × 35 mm strip of Whatman filter paper which is folded 5 mm from one end and hooked over the margin of the lower lid.

The Schirmer tear test performed with and without anesthesia evaluates tear adequacy and often aids in the diagnosis of dry eye syndrome.

The Schirmer test performed without anesthesia measures basal tear secretion and reflex tear secretion. The patient is asked to look up and not to blink or close the eyes during the test.

The Schirmer test performed with anesthesia measures basal tear secretion only by eliminating the irritation that causes reflex tearing. The patient may continue blinking normally or keep the eyes closed.

After five minutes the examiner removes the strips and measures in millimetres the length of the wetting of the filter paper strip from the bend.

A normal tear film should produce a wetting length of more than 15 mm. Without anesthesia, wetting of less than 15 mm of a Schirmer strip indicates dry eyes. With anesthesia, interpretation is as follows: 0 to 5 mm of wetting, severe dry eyes; 5 to 10 mm of wetting, moderately dry eyes; 10 to 15 mm of wetting, mildly dry eyes; and greater than 15 mm of wetting, normal tear function.

Tear Film Stability Test

Normal tear film is continuously available. Blinking maintains the tear film continuity. However if you keep your eyes open long enough, without blinking, the tear film will start breaking up. Your eye will feel uncomfortable forcing you to blink. In patients with dry eyes the tear film is unstable, and breaks up faster. Therefore the tear break up time in patients who have dry eyes is shorter.

The test for evaluating *tear film break-up time (Norn's test)* is a test to measure the relative stability of the precorneal tear film indicating condition of its mucin and lipid layers. It evaluates the interval between the last patient's blink and appearance of break-up of his or her tear film. It is noted after instilling a drop of fluorescein and examining in a cobalt-blue light of a slit-lamp. Its normal values range from 15 to 35 seconds. Values less than 10 seconds imply an unstable tear film.

2.4. Cornea Examination

Corneal Size Measurement

The size of the cornea is significant examination in cases when a patient is prescribed a contact lens or is to have a refractive operation. The size of a child's cornea is very important in evaluating the type and stage of ocular pathology (congenital glaucoma, myopia, etc.).

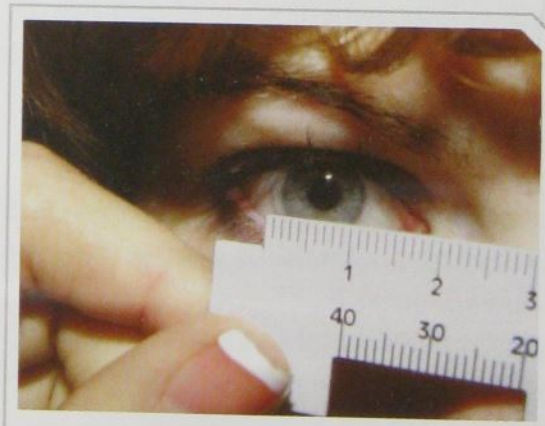


Fig. 2.15. The measurement of the corneal size

The corneal horizontal diameter (white to white) is measured with a millimetre ruler or a calibrated sheet (fig. 2.15).

The average normal horizontal corneal diameter ranges from 9 to 10.5 mm in new-borns, and 12 mm in adults. The cornea reaches the adult size by 2 years of age. Enlargement of the corneal horizontal diameter of 12 mm or more in infants and 13 mm or more in adults is called megalocornea or macrocornea.

Megalocornea in infants may indicate congenital glaucoma. The horizontal diameter less than indicated above is called microcornea.

To *perform the measurement of the corneal size*, follow these steps:

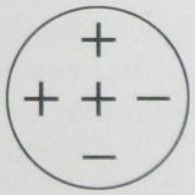
— Ask the patient to sit on a chair.
— Sit in front of the patient and ask him to look straight ahead (at bridge of your nose).
— Place the millimetre ruler on his/her lower eyelid, match the mark "0" with the patient's limbus and measure the length to the limbus of the other side of the cornea.

Corneal Sensitivity

The cornea is the most sensitive part of the eye and its center is highly sensitive as compared to peripheral areas. Assessment of corneal sensitivity is useful in the assessment of dry eyes and other conditions including herpetic keratitis, diabetes and disorders of the trigeminal nerve (CN V). To test corneal sensation there are qualitative and quantitative methods. The most commonly used method in clinical practice, which is qualitative, is the use of a distended cotton wisp. No topical anesthetics should be used before performing the test.

To *perform the evaluation of corneal sensitivity*, follow these steps:

— Ask the patient to look straight ahead.
— Bring a tip of a sterile wisp of cotton from the temporal side to touch the cornea. Different wisps have to be taken for each eye to avoid infection transfer.
— Use your left hand to test the patient's right cornea sensation and vice versa.
— Touch lightly the cornea in the places corresponding to 6, 9, 12, 3 hours and in the center and observe the patient's reaction. Normally there should be a consensual eye blink and rolling up of the eyeballs at any time the cornea is touched.
— The results are represented by writing on the corneal diagram "+" in the places, where corneal sensitivity is normal, and "–", in the places, where it is lost.



Fluorescein Staining of the Cornea

Fluorescein staining of the cornea with fluorescein (a yellow green dye) is one of the important diagnostic methods for detecting epithelial defects and foreign bodies in the surface of the cornea.

Fluorescein is applied in the form of a sterile filter paper strip, which is moistened with a drop of sterile water or saline and then touched to the palpebral conjunctiva. The patient is asked to blink. Blinking spreads the dye and coats the tear film covering the cornea surface. Under the illumination with a cobalt blue

filter any defects on the surface of the cornea will be stained by the dye and appear green. The examiner can determine the location and likely cause of the cornea problem depending on the size, place, and shape of the staining. If the test result is normal, the dye remains in the tear film on the surface of the eye and does not stick to the eye itself.

Pachymetry

Pachymetry is a simple, quick, painless ultrasonic test to measure the thickness of the cornea. Normal corneal thickness ranges from 1 mm at the very periphery to 0.5 mm at the center. Some individuals have thinner or thicker corneas, and some corneal diseases result in changes in corneal thickness. Ultrasonic corneal pachymetry is performed by placing an ultrasonic probe of a pachymeter on the central cornea, after the cornea has been anesthetized with a topical anesthetic.

Measurement of the corneal thickness is important mainly for three reasons:

- for assessing the patient's risk of glaucoma developing (people with thin corneas are at high risk for having glaucoma);
- for evaluating the accuracy of IOP readings (corneal thickness has the potential to influence eye pressure readings — a very thick cornea may cause falsely high IOP readings, a very thin cornea may cause falsely low IOP readings);
- for determining a type of laser refractive surgical procedure.

2.5. Ciliary Body Sensitivity

Diseases of the ciliary body cause pain, so its sensitivity can be detected by palpatory tenderness (palpation) (fig. 2.16).

To *evaluate the ciliary body sensitivity*, follow these steps:

- Ask the patient to have a sit in front of you.
- Ask the patient to look down and tell you if he will have any pain during the procedure.
- Locate your finger at the projection of the ciliary body — on the top of his upper eyelid.
- Lightly press the eye and control the patient's reaction.

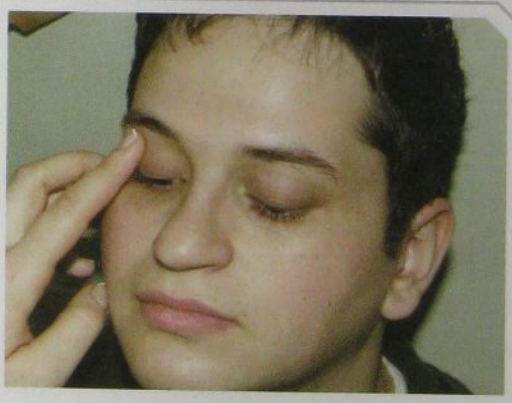


Fig. 2.16. Examination of the ciliary body sensitivity

2.6. Gonioscopy

Gonioscopy is the method of evaluation of the eye anterior chamber angle which is performed with the help of a goniolens (also known as a gonioscope) placed directly on the anesthetized cornea and a slit lamp or operating microscope to gain a view of the iridocorneal angle (anatomical angle formed between the eye's cornea and iris). This type of lenses has a different number of mirrors located at variable angles. The Goldmann three-mirror lens is the best known and the most popular one (fig. 2.17). The importance of gonioscopy is in diagnosis and monitoring of various eye conditions associated with glaucoma.

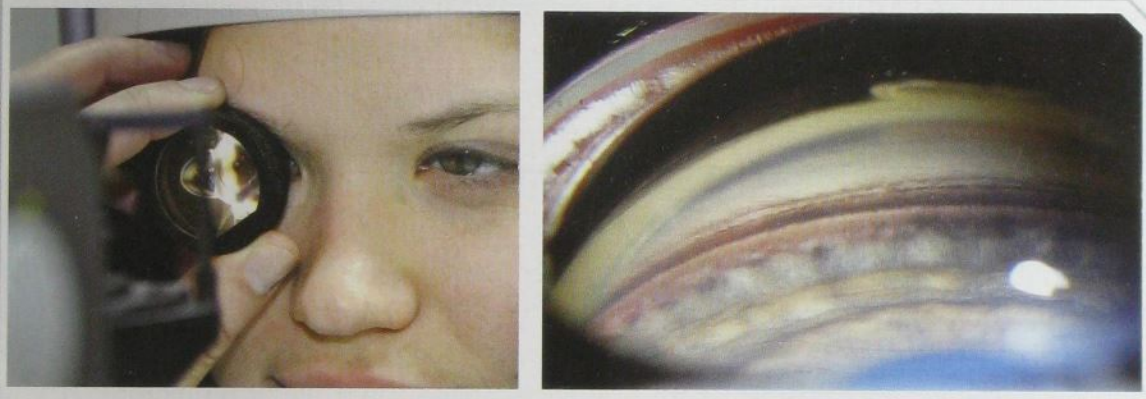


Fig. 2.17. Examination of the anterior chamber angle with the Goldmann three-mirror lens and the view of the wide anterior chamber

2.7. Campimetry

Campimetry is the measurement of the central (30 degrees) part of visual field on a flat surface in contrast to perimetry which is the measurement of peripheral visual field on a curved surface. The tangent screen (Bjerrum) is the standard method to perform campimetry. For tangent screen examination, a black screen, blackboard or other flat surface is used. Vision is tested with the patient seated 1 m to 2 m from the tangent screen by presenting different sized pins attached to a black wand, which may be moved, against a black background. This test stimulus (pins) may be white or colored. The tangent screen is especially valuable for measuring the size of physiological blind spot and for central field defects detection.

2.8. Amsler Grid Test

The Amsler grid is another test for measuring the central (approximately 8 degrees in diameter) visual field which corresponds with the macula.

The test consists of a card with horizontal and vertical black lines intersecting on a white background or reverse (white lines on a black background), and has a dot in the center for fixation. After correction for any near refractive errors, testing each eye separately, the patient should hold the chart at a comfortable reading distance from their uncovered eye, and fix gaze on the central spot of the grid. While focusing on the dot in the center of the grid, areas that are blurry, absent, or distorted are identified by the patient.

This test is an excellent, fairly sensitive method of detecting early macular disorders (macular degeneration) and also helpful in monitoring changes in vision once they have been detected. The Amsler grid test can be performed at home for regular self-evaluation.

2.9. Examination of Contrast Sensitivity

Examination of contrast sensitivity is based on the ability of the visual system to distinguish an object from its background. If you have high contrast sensitivity, you will be able to see objects even when they are outlined poorly and are very similar to their background, for example, a white piece of paper on a white tablecloth. If you have low contrast sensitivity, objects will appear to blend into the background, and you may have difficulty with many common tasks such as seeing spots on clothes or dishes, and reading facial expressions. Contrast sensitivity is a very important visual function, especially in situations of low light, fog or glare, when the contrast between objects and their background is often reduced. Driving at night is an example of an activity that requires good contrast sensitivity for safety.

The current gold standard in the assessment of vision, visual acuity, provides only a limited amount of information, obtained under artificial conditions. Contrast sensitivity testing measures a range of visual performance under real-life conditions. It measures the least amount of contrast needed to detect a visual stimulus and gives a more complete quantitization of patients' visual capabilities.

Contrast sensitivity is tested using sinusoidal grid patterns (which may be dots, bars or letters) of varying contrast and spatial frequency (gradually get more similar to the background). In routine practice, however, this is usually achieved more easily with acuity charts on which contrast is reduced in several stages.

Many instances in which losses in contrast sensitivity were detected when visual acuity was normal have been reported. These include amblyopia, neuroophthalmology, retina, anterior segment disease, and glaucoma. Therefore, contrast sensitivity testing enables the clinician to diagnose selective deficits in visual processing at an earlier stage than is possible with conventional testing methods.

2.10. Color Vision Testing

Color vision is the ability of the eye to discriminate between different colors based on the different ranges of the retinal sensitivity to wavelength of the light coming into the eye. The visible spectrum comprises wavelengths between 780 and 360 nm. Electromagnetic rays outside of the spectrum (infrared and ultraviolet) cannot be detected by the human eye.

Colors are characterized by three parameters:

— Hue — the most obvious characteristic of a color, which is described by the dominant wavelength.

— Saturation — the intensity or purity of a color; high saturation colors look rich and full, low saturation colors look dull and greyish.

— Brightness — lightness of a color; it indicates the quantity of light reflected, from black (no brightness) to white (full brightness).

Color vision, as well as high acuity vision, is a function of cones. There are two major perceptual theories in color vision*.

This theory also helps to explain some types of color vision deficiency. For example, people who are color-blind to red are also color-blind to green, and people who can't see blue also can't see yellow. All these observations led to the conclusion that red and green are paired and that blue and yellow are paired.

These two theories show that color processing occurs in two stages. The trichromatic theory explains color vision phenomena at the photoreceptor level; the opponent-process theory explains color vision phenomena that result from the way in which photoreceptors are interconnected neurally.

The most widely accepted theory (Young—Lomonosov—Helmholtz trichromatic theory) is that there are three types of cones, each containing photopigment that has a different sensitivity to light of different wavelengths — S-cones are sensitive to short-wavelength light ("Blue"); M-cones are sensitive to medium-wavelength light ("Green"); L-cones are sensitive to long-wavelength light ("Red"). Color perception is a result of stimulation of all cone types but in different degree. Equal stimulation of all types of cones results in the perception of white color.

The opponent-process theory of color vision proposed by Edward Hering states that the cone photoreceptors are linked together to form three opposing color pairs: blue/yellow, red/green, and black/white. Activation of one member of the pair inhibits activity in the other. This allows the visual system to record differences between the responses of cones, rather than each type of cone's individual response. Consistent with this theory, no two members of a pair can be seen at the same location, which explains why we don't experience such colors as "bluish yellow" or "reddish green".

Contemporary research has confirmed that, en route to the brain, neurons in the retina and the thalamus code the color-related information from the cones into pairs of opponent colors, as demonstrated by afterimage effect. If we view colored stimuli for an extended period of time, we will see an afterimage in a complementary color. For example, if you stare at something red for a minute and then avert your eyes toward a white surface, then you will see a green afterimage and vice versa when staring at green. The same occurs with blue and yellow, black and white. This vivid color aftereffect can last for a minute or more.

Focus your gaze on the dot in the center of the four-color patch afterimage stimuli below for 30 seconds trying not to blink. Then shift your gaze to a white paper, you should notice that the original colors are all reversed — where you saw red it is now green and vice versa; likewise for blue and yellow.

Normal color vision is called *trichromacy*. It is a state when a person can distinguish all the different colors as well as subtle mixtures of hues due to normal sensitivity of all three types of retinal cone photoreceptors. People with normal color vision are known as trichromats.

Disorders of color vision may be either congenital or acquired. Reasons for an acquired color vision deficiency may be ocular pathology, intracranial injury, or excessive use of therapeutic drugs.

EYE FACTS

According to different scientific references a healthy human eye can distinguish more than 100 000 to about 10 million different colors and differentiate between 500 shades of grey. About 2 % of women have a rare genetic mutation that gives them an extra retinal cone allowing them to see more than 100 million colors.

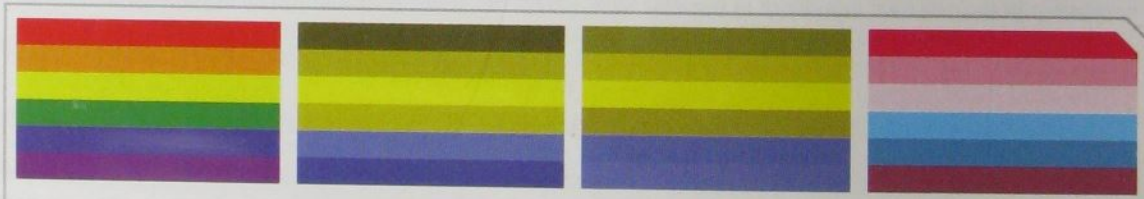
Color vision defects are classified into following groups:

Anomalous trichromacy is an impairment of normal color vision, but not a complete loss. This occurs when all types of the cones are functional, but one of them is anomalous having a shifted spectral sensitivity peak. This results in an impairment, rather than loss of trichromacy. Anomalous conditions are called based on whether the L-cones (Greek: prot-, referring to the red photoreceptors), M-cones (deuter-, the green ones), or S-cones (trit-, the blue ones) are affected:

— *protanomaly* (red-weak) is a reduced sensitivity to red light;
— *deuteranomaly* (green-weak) is a reduced sensitivity to green light and is the most common form;
— *tritanomaly* (blue-weak) is a reduced sensitivity to blue light and is extremely rare.

Dichromacy is a moderately severe color vision defect in which one of the three basic color mechanisms is absent or not functioning (fig. 2.18).

— *protanopy* (red-blind) is a color vision deficiency caused by the complete absence of red retinal photoreceptors;
— *deuteranopy* (green-blind) is a color vision deficiency in which the green retinal photoreceptors are absent;
— *tritanopy* (blue-blind) is a rare color vision deficiency caused by complete absence of blue retinal photoreceptors.



Colors of the rainbow (normal color vision) | Colors of the rainbow (protanopy) | Colors of the rainbow (deuteranopy) | Colors of the rainbow (tritanopy)

Fig. 2.18. How persons with different forms of color blindness perceive the colors of the rainbow (https://commons.wikimedia.org)

Monochromacy is a severe color vision defect in which two cone pigments are missing (can be temporal or constant).

— *erythropsia* (red vision) — defect of vision in which all objects appear reddish;
— *xanthopsia* (yellow vision) — defect of vision in which all objects appear yellowish;
— *chloropsia* (green vision) — defect of vision in which all objects appear greenish;
— *cyanopsia* (blue vision) — defect of vision in which all objects appear bluish.

Achromacy (or achromatopsia) — is also known as "total color blindness". It is the inability to distinguish colors caused by cone defect or absence. People with monochromatic vision can see no color at all and their world consists of different shades of grey ranging from black to white.

EYE FACTS

Color blindness is more common in males.

There are two types of monochromacy:

— *rod monochromacy* — complete lack of cones, it is associated with reduced visual acuity, hypersensitivity to light (photophobia) and other vision impairments;
— *cone monochromacy* — there is only one cone type, it is associated with good visual acuity and none of the other vision impairments are found.

Commonly used tests in clinical practice today include isochromatic plates, anomaloscopes, arrangement tests, and lantern tests. Each category has unique attributes that make it suitable for a particular clinical situation.

The standard test for color sense examination is the *Rabkin polychromatic plates*. These plates are composed of dot colored patterns on colored backgrounds of randomly mixed colors to assess the ability to recognize color differences (fig. 2.19). People with normal color vision are able to identify the number shown by the dot pattern. If color vision is defective, you will be unable to distinguish between the dot pattern and the background. A number of the plates contain also "hidden" figures, which are visible only to the dichromate. Triangles, circles and squares appear on some plates instead of figures, and so can be used for testing children and illiterates. A diagnosis can be made from the tables of dichromacy in general, and of protanopy, deuteranopy, and tritanopy in particular.

Fig. 2.19. The example of Rabkin polychromatic plates

To perform *color vision examination with the help of* the *Rabkin polychromatic plates*, follow these steps:

— Ask the patient to sit comfortably on the chair.
— Provide adequate illumination (day light), so the patient should sit with the back to the window.
— Hold a plate strictly vertically 0.75—1.0 m from the patient so that it is at his or her eye level.
— Show a plate to the patient for 5 sec (10 sec at most for special plates).
— Ask the patient to tell what pattern (number, figure) is seen.
— If plates are read correctly, the color vision is regarded as normal.
— If certain plates are read incorrectly the color vision is regarded as deficient.

It is not necessary in all cases to use the whole series of plates. Some of them may be omitted if the test is held to separate patients with normal color perception from those who have color defects.

There exist other color vision tests — pseudoisochromatic plates of Ishihara, of Yustova—Alekseeva (fig. 2.20), which are similar to the Rabkin test. Pseudoisochromatic tests should be used primarily as screening tests for diagnosis of color defects; their diagnostic value is limited. The results provided by anomaloscopes permit to differentiate the type of color vision deficiency.

Fig. 2.20. Yustova—Alekseeva pseudoisochromatic plates

Color vision defects can be diagnosed by means of spectral *anomaloscopes,* measuring qualitative and quantitative anomalies in color perception. These are optical instruments, based on the principle of color matching — comparison of two colored fields. For example Nagel anomaloscope is used to determine the degree of red or green color vision deficiency. One color field is constantly yellow monochromatic with adjustable brightness (control field) and the other is illuminated by the green and red rays by mixing of which the patient should receive pure yellow, corresponding control field. If the patient is a dichromat, he/she will be able to make a match for all red-green mixture ratios. Anomalous trichromats don't accept the normal match and the distance of their match indicates the severity of their deficiency. On the other side, if the patient suffers from a protan vision deficiency, he/she will use much more red to match the colors compared to people with a deutan defect, which use more green in their mixture.

The arrangement test consists of a certain number of colored discs or plates which have to be arranged in the correct order starting from a pilot plate.

2.11. Examination of Binocular Vision

Binocular vision (or stereoscopic vision) is simultaneous vision with two eyes, which is achieved by the coordinate use of both eyes, so that separate and slightly dissimilar images arising in each eye fovea are received by the brain as a single visual object by the process of fusion. Normal binocular vision also implies depth perception — stereopsis — based on the horizontal separation of the two eyes in the skull. Due to binocular vision we can determine the distance from one object to another, depth, volume and relative position of objects. Humans have about 120° of the binocular field of view.

Normal binocular vision requires:

— same vision acuity in both eyes (not less than 0.3);
— transparency of optical media;
— the same degree of refraction in both eyes that produce equal image size on the retina;
— precise coordination of the movement between the two eyes;
— location of both eyes in one frontal and horizontal plane;
— normal functional ability of the retina, pathways and higher visual centers.

Binocular vision can be tested by different methods.

The Kalfa method or *two pencils test* (test with missing the point) — binocular vision is tested with the help of pencils, ball-point pens, or two knitting needles. The examiner holds a pencil in upright position. The task for the patient is to connect the tip of another pencil vertically with the first one so that both pencils form a straight line. In the presence of binocular vision the task can be done easily. In the absence thereof the point is missed, as can be easily seen repeating the test with one eye closed.

The Sokolov test (test with "a hole in a palm") is that the patient looks with one eye through the tube (e.g., a rolled piece of paper), to the end of which on the side of the other open eye he/she puts the palm. In case of normal binocular vision one gets the impression that the palm has a hole, through which the picture visible through the tube can be seen. This phenomenon can be explained in such a manner: the picture viewed through the tube is overlaid on the image of the palm in the other eye. In case of simultaneous vision, as distinct from binocular, the "hole" does not coincide with the center of the palm, and in case of monocular vision the phenomenon of "a hole" in the palm does not occur.

Reading with a pencil (or a pen) test is as follows: a pencil is placed several centimetres from the patient's nose at the distance of 15 cm above the text so that the pencil covers a part of the text letters. The patient can read the text with such an obstacle without moving the head in case of having normal binocular vision, as the letters, covered by the pencil for one eye, are seen with the other eye and vice versa.

More precise examination of binocular vision can be held by the Belostotskiy—Friedman *4-dot color test*. The apparatus consists of a box containing 4 colored dots — 2 green, 1 red and 1 white (fig. 2.21). The patient looks at the test through the red-green glasses over optimal refractive correction, with a red filter placed in front of the right eye and a green filter — in front of the left eye. The middle white dot, visible through the red and green filters of glasses, will be perceived as green or red, depending on the predominance of the right or left eye. In case of monocular vision of the right eye, the patient sees only two red dots, in case of monocular vision of the left eye — only three green dots. In case of normal binocular vision four dots will be seen — two red and two green or one red and three green dots (depending on the leading eye). In case of simultaneous vision (absence of fusion), the patient sees five dots — two red and three green.

Fig. 2.21. The Belostotskiy—Friedman apparatus for performing the *4-dot color test*

The Bagolini striated glass test consists of two optically clear lenses with narrow fine parallel scratches that barely blur the environment. They don't affect the vision or accommodation of the patient. The lenses are placed (in front of the patient's refractive correction if needed) in the trial frame at 45° for the right eye and 135° for the left eye. The patient fixates a small light viewed at eye level at either 33 cm or 6 m in a well-illuminated room. In the presence of normal binocular vision the patient sees an "X" pattern but if central suppression is present, a break will be reported in one of the lines; when global suppression is present, only one line will be observed.

2.12. Examination of Light and Dark Adaptation

Perception of light is the ability of an eye to perceive the light of different intensities. Vision according to light conditions can be of three types:

Photopic vision (daylight vision) is vision under bright light conditions, normally usual daylight intensity. It is based on cones function and therefore allows color perception as well as higher visual acuity.

Scotopic vision (night or twilight vision) is vision under low light conditions. It is based completely on rods function, so colors cannot be identified (achromatic vision).

Mesopic vision is vision under intermediate levels of illumination. It is basically a combination of photopic vision and scotopic vision, so both cone cells and rod cells are being used.

The visual system is capable to adapt to various degrees of illumination. This adaptation may be to high light intensities (light adaptation) and to the decreased light intensity (dark adaptation).

Light adaptation is adaptation to increased levels of light intensity. It is a protective mechanism — when light intensity is too high: (1) the pupil will constrict in order to decrease the amount of light rays into the eye; (2) sensitivity of the cones and rods are decreased at the same time. With light adaptation, the eye can adapt quickly to the high illumination to be able to distinguish objects in this background. The process for light adaptation occurs by the end of 1st minute.

Dark adaptation is adaptation to reduced light intensity and involves three distinct changes in the visual system:

EYE FACTS

Your eyes are so sensitive that if the Earth was flat you could see a candle flickering at night from up to 30 miles away.

EYE FACTS

Cats have excellent night vision due to several anatomic features of their eyes: six to eight times more rod cells, which are more sensitive to low light, than humans do; elliptical pupils that are oriented vertically that allows it to open much larger than a human's round pupil and so lets more light in; a membrane called the tapetum lucidum, which reflects any light that passes through the retina back into the eye, thereby increasing the eye's sensitivity to dim light. The flash of cat's eyes in the dark, that we see, is light reflecting off of the cat's tapetum.

1. enlargement of the pupil (wider pupil allows more of the available light to enter the eye and stimulate the retina;

2. increased sensitivity of the cones to light (cones become completely dark-adapted within about five to ten minutes, resulting in a loss of color vision);

3. increased sensitivity of the rods (rods become dark-adapted after about 20—30 min and reach the maximum in 50—60 min)*.

Insufficiency of adaptation to different levels of light intensity can be of two types:

— *nyctalopia* (Greek "nyctos" (night) + "alaos" (blind) + "ōpsis" (vision) — night blindness) — decreased dark adaptation, inability to see clearly in dim light;

— *hemeralopia* (Greek "hēmera" (day) + "alaos" (blind) + "ōpsis" (vision) — day blindness) — decreased light adaptation, inability to see clearly in bright light; twilight vision is better, is much rarer.

Hemeralopia can be of three types:

— symptomatic — due to retinal disorders;

— functional — as result of A hypovitaminosis;

— congenital.

To examine the light adaptation a *photo-stress test* may be used. It is timing the recovery of visual acuity after the patient's eyes being illuminated with an intense light source about 30 sec.

To evaluate the dark adaptation a several diagnostic techniques can be used.

Orientation method — observation of a patient. The test is held in dark room. The patient is asked to come to the certain apparatus, sit on the chair, take a certain object or count a number of white papers scattered over the room and so on.

Adaptometry — a vision test performed by means of adaptometer. The test is carried out in the dark room; its duration is 50—60 min. First the patient, looking at the illuminated screen of the adaptometer, adapts to the light during 10 min and then plunges into complete darkness. The patient is presented a dimly light test the brightness of which is gradually increasing. When the light test is seen the patient presses the button. The brightness of the test first changes in 2—3 min and then every 5 min. After 60 min the examination is complete. With the adaptation the patient begins to distinguish the control light test at a lower level of illumination. Results of the study are drawn in a graph.

Control method — a test of Kravkova—Purkinje based on the Purkinje phenomenon (Purkinje shift**).

To perform the test you need a square piece of black cardboard (20 × 20 cm) on angles of which 4 squares of color paper — red, yellow, blue and green (3 × 3 cm) are

---

* Rods at the dark are blind to red light. Thus, to view a map at night without disturbing the dark-adaptation of the rods ("night vision"), illuminate the map in red light. Because the rods do not "see" the red light, they remain fully dark adapted.

** Purkinje phenomenon is the tendency for the peak light sensitivity of the eye to shift under changing illumination levels from the red end of the color spectrum at high illumination levels toward the blue end of the color spectrum at low illumination levels. This phenomenon describes the fact that at high light intensity a rose can be perceived as bright red against the dull green of its leaves or neighbouring blue flowers and then in the dusk the contrast is reversed — red rose appears an almost black, and green leaves and blue flowers appear relatively bright.

glued. In the dark room this test is shown to the patient at a distance of 40—50 cm from the eye. Normally after 30—40 sec the patient distinguish the yellow square then after 50—60 sec — blue square. At the decreased dark adaptation the yellow square appears as a bright spot, the blue square is seeing after more than 60 sec or invisible.

To perform *dark adaptation test by the control method*, follow these steps:

— Ask the patient to sit on the chair.
— Show the patient the test (the black square with for little squares of red, yellow, blue and green color).
— Turn the light off (the test should be performed in the dark room).
— Place the test at a distance of 40—50 cm from the patient's eyes.
— Ask the patient to inform you after seeing the yellow and blue squares.

3. Investigative Techniques

3.1. Keratometry

Keratometry (or Ophthalmometry) is the objective measurement of the form and curvature of the corneal front surface (approx. 2—4 mm radius), and determines the optic power of the cornea. The test is based on the fact that the cornea acts as a convex mirror, so its radius of curvature can be calculated from the size of reflected image formed by anterior corneal surface. In a keratometry procedure, a device is used to measure the degree of curvature in the cornea, looking for the shallowest and steepest curves.

Keratometry is essential in the contact lens assessment, evaluating and following patients with astigmatism and keratoconus, and in calculating of appropriate power of an intraocular lens in cataract surgery.

3.2. Corneal Topography

Corneal Topography (or Videokeratography, or Corneal Mapping) is an imaging technique of corneal curvature examination assisted by a computer analysis that creates a detailed color 3-D map of the shape and power of the cornea (fig. 2.22). It is performed by projecting illuminated rings on to the corneal surface which are reflected back and measured by the instrument. The test results in the generation of a top-

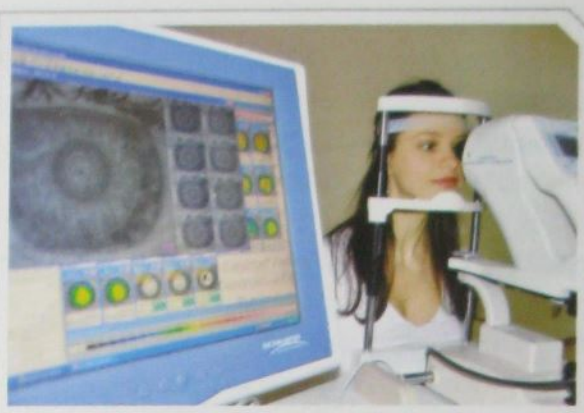


Fig. 2.22. Keratotopography performing

ographical map of the cornea, which color-codes the steepness of curvature according to its dioptric value.

Corneal topography extends the measurement range from the four points a few millimetres apart that is offered by keratometry to a grid of thousands of points covering the entire cornea.

This method is used in estimating the degree of astigmatism, in keratoconus detecting and fitting contact lenses. It is also essential for patients being considered for refractive surgical procedures such as Lasik.

3.3. Pentacam

The Pentacam is a non-invasive and non-contact computer-aided diagnostic system that provides a 3-dimensional image of the anterior segment of the eye. This device combines the use of the slit illumination system together with a rotational camera, called a Scheimpflug, to achieve a complete measurement and analysis of the center of the cornea. The camera is oriented so that the image of the illuminated place appears distinct and sharp from the anterior surface of the cornea right up to the posterior surface of the crystalline lens. The slit-camera device rotates 180 degrees around the eye, generating a series of radially oriented images (at a rate of approximately 25 per second) that are combined to create a three-dimensional model of the anterior eye segment. These images are used to measure the thickness and topography of the cornea, depth of the anterior chamber and density of the lens.

The Pentacam is used to evaluate potential patients for Lasik, cataract, glaucoma surgery and corneal transplants among other procedures. Also, it is a tool used to determine if a patient is a candidate for a refractive eye surgery by screening for keratoconus.

3.4. Aberrometry

The eye is an optical complex system with different refractive surfaces. Ideally, light entering the eye is refracted, mainly by the cornea and the crystalline lens, then is focused on the retina and an image appears clear. In reality, the light is sometimes

distorted by imperfections in the corneal surface or lens pathology leading to lack of focus and resulting in blurry vision. These distortions are called aberrations. In other words, optical aberrations are vision defects that occur when light rays are improperly refracted in the eye that decrease the quality of the retinal image and visual perception.

There are lower order aberrations — myopia, hypermetropia, astigmatism, correctible with glasses, contact lenses or refractive surgery. There are higher order aberrations such as coma, trefoil and spherical aberrations. Patients who complain of glare, halos, starbursts and poor night driving often have increased higher-order aberrations.

Aberrometry is a technique to measure aberrations with the help of an instrument called aberrometers. Aberrometers objectively measure the overall refractive power error of the eye. They do this by mapping how light rays travel through the entire eye optical pathway, comparing to the way light travels through an optically perfect eye. In result aberrometers provide maps using color gradients to represent magnitudes of the refractive errors, which enable to locate even obscure imperfections that cause vision defects.

Aberrometry provides fundamental information used in choosing the design of vision correction such as contact lenses, glasses, intraocular lenses or type of corneal refractive surgery and to evaluate the success of the correction.

3.5. Ultrasonography

Ophthalmic Ultrasonography is the important diagnostic imaging technique to visualize various ophthalmic disorders by reflecting high energy sonic waves off deep ocular structures. It is a safe, non-invasive diagnostic tool that is most useful in the presence of opaque ocular media caused by corneal opacities, anterior chamber opacities, cataract, vitreous hemorrhage, or inflammatory opacities. Ultrasonography allows performing evaluations of the anterior segment of the eye as well as the eye's intraocular and orbital structures.

A-scan (or Biometry) provides data on the axial length of the eye. (Average axial length of normal eye 23.06 mm (majority 22.0 to 24.5 mm)). It is essential measurement in order to calculate the power of IOL for patients undergoing cataract surgery. It is also used for measurement of anterior chamber depth, cornea and lens thickness and detection of lesions.

B-scan (or Brightness scan) produces two-dimensional cross-sectional view of the eye and orbit. This method of imaging is the most important examination technique for determination of various abnormalities in the posterior segment of the eye (lens, choroid, sclera, vitreous, retina), particularly in the presence of anterior segment opacities. Ocular conditions such as cataract, vitreous degeneration, retinal tear or detachment, ocular trauma, choroidal melanoma, and retinoblastoma can be accurately evaluated with this method.

Doppler ultrasonography is an established method for investigation of the ocular and orbital blood flow characteristics. It is especially important for assessment of the circulatory status in much ocular pathology, is more widely used for the investigation of the circulatory status in retinal vascular disorders, including degenerations, dystrophies, tumors, retinal detachment, etc. It has also been reported as a valuable tool for the clinical management of vascular lesions.

3.6. Fundus Photography

Fundus Photography (or Fundography) is a highly specialized form of examination of the retina (also called the fundus) with the help of a fundus camera, or retinal camera, which is a specialized low-power microscope with an attached camera designed to photograph the interior surface of the eye (fig. 2.23). It provides an upright magnified view of fundus and produces a series of detailed color photos of the retina that are helpful for diagnosing, documenting, and monitoring certain eye conditions such as macular degeneration, retinal neoplasms, choroid disturbances and diabetic retinopathy, or for identifying glaucoma, multiple sclerosis, and other abnormalities.

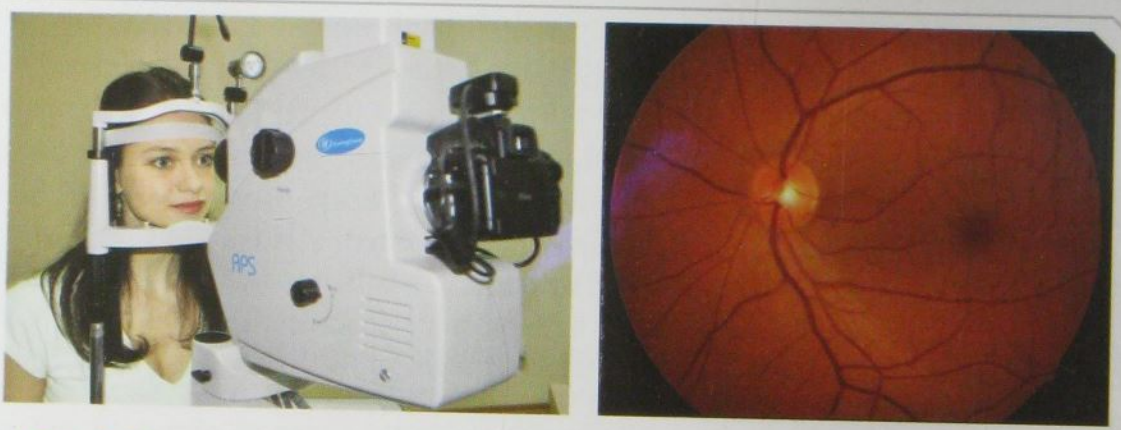


Fig. 2.23. Fundus photography performing and the photo of the normal fundus

3.7. Fluorescein Angiography

Intravenous Fluorescein Angiography (IVFA) or Fluorescein Angiography (FAG) is a technique for detailed examination of the retinal and choroidal circulation using fluorescein dye and a specialized fundus camera. It involves injection of Sodium Fluorescein into the antecubital vein and then an angiogram is obtained by photographing the fluorescence emitted after illumination of the retina with blue light at a wavelength of 490 nanometres. The test uses the dye tracing method.

About ten — twelve seconds after the injection, the dye appears in the arteries of the retina. Over a two to five seconds period, the dye travels through the arterioles and capillaries and fills the veins. Complete filling of the veins occurs over the next ten seconds with maximum vessel fluorescence occurring approximately 30 seconds after injection and recirculation phases follow, this occurs about 2 to 4 minutes after injection. The veins and arteries remain roughly equal in brightness. The intensity of fluorescence diminishes slowly during this phase as much of the fluorescein is removed from the bloodstream. Ten minutes after the injection, the dye has mostly evacuated from the eye, having stained the optic nerve head. The dye is metabolized by the kidneys and is eliminated through the urine within 24 to 36 hours of administration.

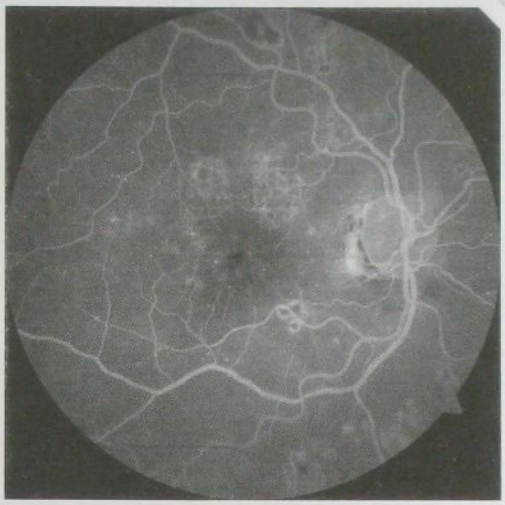


Fig. 2.24. Fluorescein angiography of a patient with diabetic retinopathy after focal laser photocoagulation performing

A series of black-and-white or digital photographs of the retina are taken before and after the fluorescein reaches the retinal circulation (approximately 10 seconds after injection). The early images allow for the recognition of autofluorescence of the retinal tissues. Photos are taken approximately once every second for about 20 seconds, then less often. A delayed image is obtained at 5 and 10 minutes. Some doctors like to see a 15 minute image as well.

Fluorescein angiography diagnoses pathologic changes in the retina by detection of either hyperfluorescence (capillary leakage, aneurysm, neovascularization) or hypofluorescence (blocking defects) (fig. 2.24).

3.8. Optical Coherence Tomography

Optical Coherence Tomography (OCT) is a non-invasive, non-contact imaging technology used to obtain high resolution 3-D cross-sectional images of the retina. The layers within the retina can be differentiated and retinal thickness can be measured to aid in the early detection and diagnosis of retinal diseases and conditions. OCT testing has become a standard of care for the assessment and treatment of most retinal conditions.

In addition to imaging the posterior segment of the eye, OCT can be used to view the cornea, sclera, and anterior chamber angle as a cross-section as well. It can measure the corneal radius and the central corneal thickness, determine the depth and the

volume of the entire anterior chamber, evaluate the anterior chamber angle and visualise any changes in the anterior eye structures (fig. 2.25).

OCT is similar to ultrasound imaging but at a much higher resolution and uses light waves instead of sound ones. OCT can provide cross-sectional images of tissue structure on the micron scale in situ and in real time. OCT can function as a type of optical biopsy and is a powerful imaging technology for medical diagnostics because unlike conventional histopathology which requires removal of a tissue specimen and processing for microscopic examination. OCT can be used where standard excisional

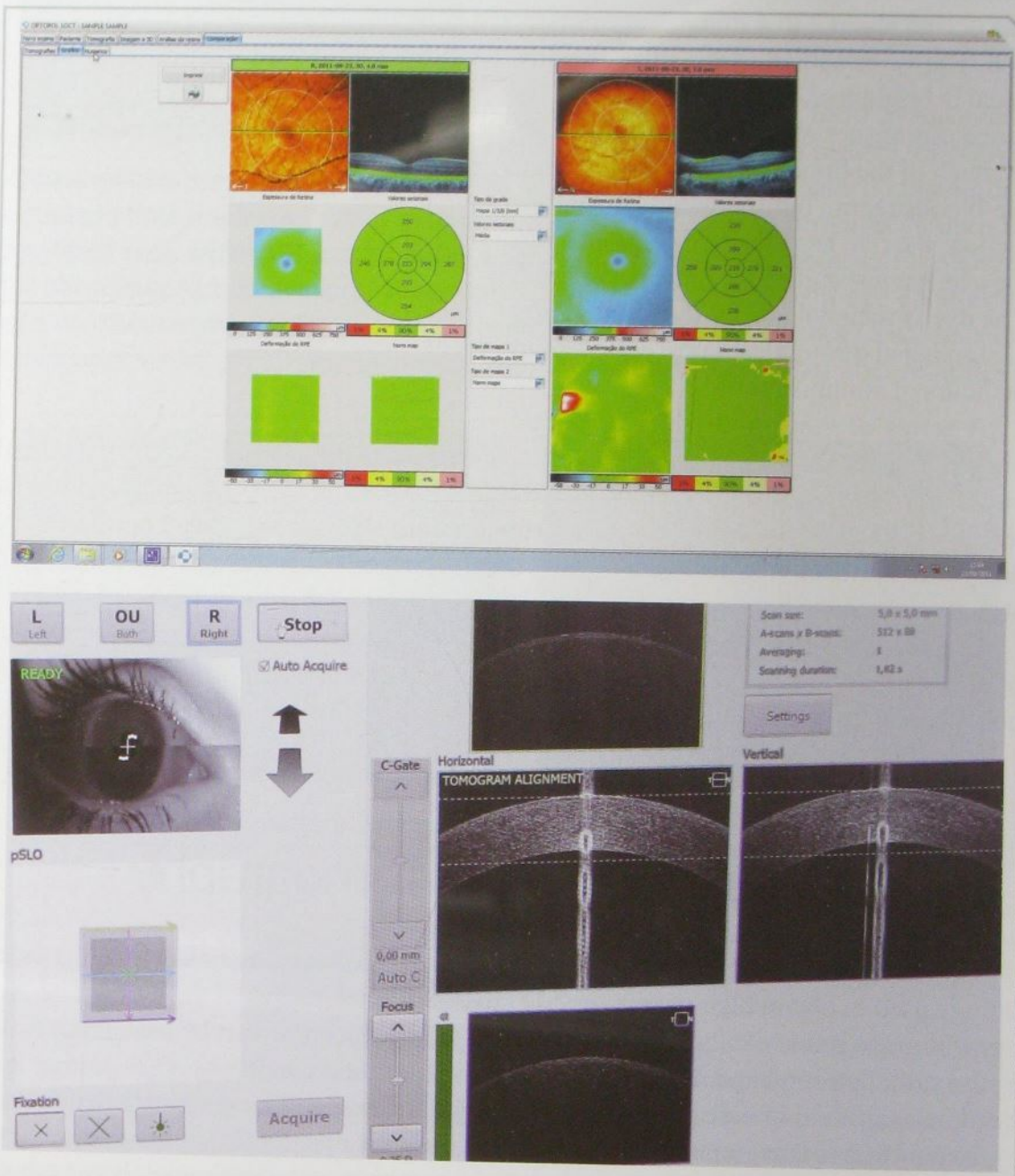


Fig. 2.25. Imaging of the posterior segment and anterior chamber with the OCT

biopsy is hazardous or impossible, to reduce sampling errors associated with excisional biopsy, and to guide interventional procedures.

OCT provides remarkably detailed information about the retina and anterior segment structures and allows seeing abnormalities that are not apparent on regular clinical examination. It is particularly useful in certain diseases like macular holes, diabetic retinopathy, glaucoma, optic nerve damage etc.

3.9. Heidelberg Retinal Tomography

The Heidelberg Retinal Tomography (HRT) is a computerized confocal scanning laser diagnostic technique that produces and analyzes three-dimensional images of the posterior segment of the eye, including the optic nerve head and surrounding retina. The HRT is a system that combines a laser scanning camera and specialized software that starts from photographing the surface of the optic nerve and then focuses on deeper and deeper layers before putting them all together to create the 3-D image. The HRT gives precise measurement of the size, depth and shape of the optic nerve head and is especially useful at detecting changes over time (fig. 2.26).

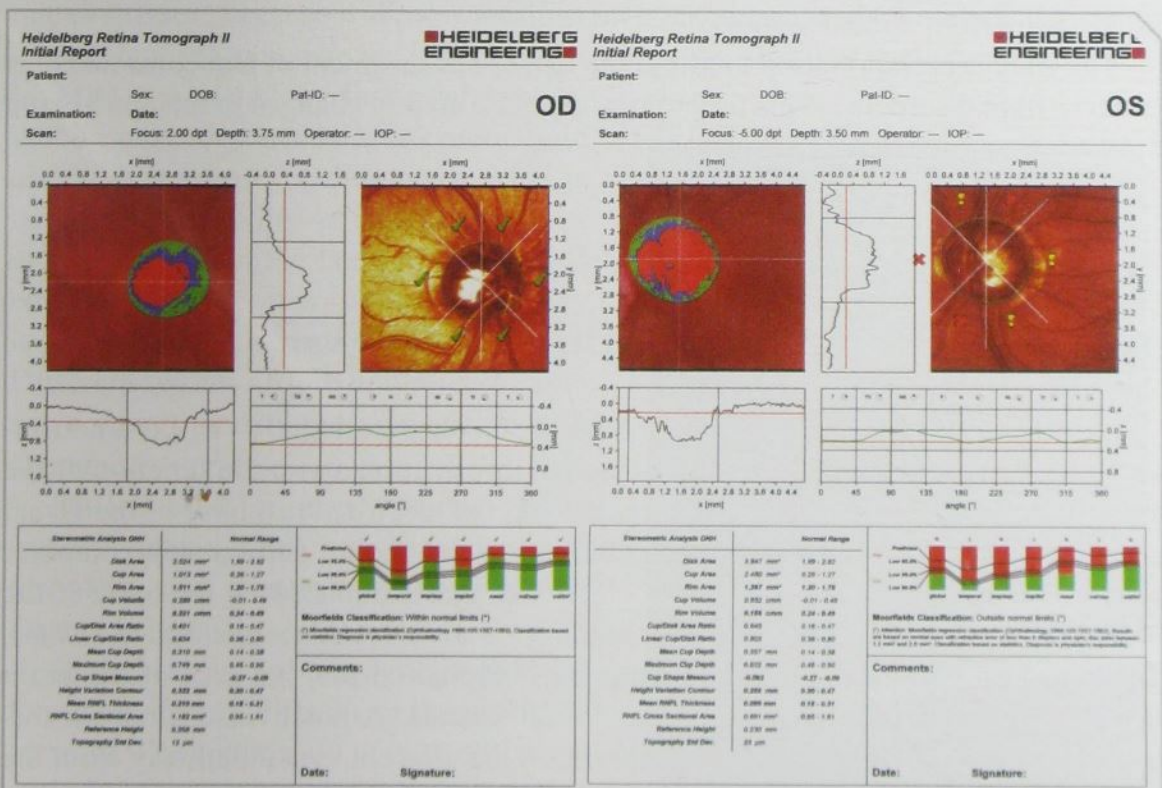


Fig. 2.26. The report of HRT examination of a patient that reveals the normal state of the optic nerve head of the right eye and pathology in the optic nerve head of the left eye

HRT is specifically designed for aiding in glaucoma diagnosing as can detect even small changes in the optic disc before glaucoma symptoms become apparent to a patient and visual field defects are present. This is an extremely important test for patients at risk for glaucoma. Diagnosing glaucoma early means that treatment can begin before damage has been done to the optic nerve in order to save patients' vision.

Additionally, the HRT is a powerful tool for aiding in the diagnosis and management of retinal abnormalities — macular holes and edema, age-related macular degeneration and diabetic retinopathy.

3.10. The GDx Nerve Fibre Analysis

The GDx Nerve Fibre Analysis (or Scanning Laser Polarimetry) is a diagnostic technique that uses a confocal scanning laser ophthalmoscope with an integrated polarimeter. The GDx analyzer measures the thickness of the retinal nerve fibre layer and then analyzes the results and compares them to a database of normative values. A standard GDx scan gives a fundus image, a nerve fibre thickness color map, and a map of deviation from normal values.

The GDx is used primarily to assess the risk or advancement of glaucoma and other nerve fibre disorders as this technology can pick up tiny changes in the optic nerve before vision loss occurs.

3.11. Visually Evoked Response

Visual Evoked Response (VER) or Visually Evoked Potential (VEP) is a non-invasive testing method that provides objective information about the function of the entire vision system. This test measures the time of electrical response of the visual cortex to a visual stimulus such as an alternating chessboard pattern on a computer screen. Responses are recorded from electrodes that are placed on the back of the patient's head and are observed as a reading on an electroencephalogram (EEG). These responses usually originate from the occipital cortex, the area of the brain involved in receiving and interpreting visual signals. A positive result from this test depends upon the adequate functioning of the afferent visual pathway from the retina to the brain.

Visual evoked response (VER) tests the visual function of the optic nerve and brain for the diagnosis of vision loss due to retinal disease, tumors, multiple sclerosis, hydrocephalus and other visual disorders.

3.12. Radiological Imaging Techniques

Computed Tomography (CT) scanning has become an important diagnostic technique as it allows to precisely visualize the orbital structures in various planes.

CT is based on ionizing radiation. As X-ray beams pass through tissues they are absorbed or weakened at different levels depending on the type of tissue they pass through. CT scanners use multiple detectors to measure these X-ray weakening profiles and produce images. The computer converts them into images that resemble 2-dimensional slices (cross-sections) or 3-dimensional images. 3-D reconstructions are useful in trauma cases. The high intrinsic contrast between bone, muscles, orbital fat and air produces excellent visualization of orbital structures and metal foreign bodies.

CT is especially used for orbital fractures, calcification and for determining the size and localization of intraorbital or intraocular metallic foreign bodies. The minimum size of metal piece revealed by computed tomography is 0.2, 0.3 mm. Glass, wood, and plastic are less radiopaque and therefore more difficult to isolate on CT. It doesn't require contact with the ocular surface and so can be used with open globe injuries.

CT is also used for determining some pathologic features, including orbital and ocular tumors, vascular changes or neuro-ophthalmic problems.

Magnetic Resonance Imaging (MRI) is a most useful technique for neuro-ophthalmic evaluation, because it allows visualization of the cranial and intraorbital nerves and detection of space-occupying lesions in the orbit, intracranial or hypophyseal region.

MRI is a technique that produces detailed images by means of magnetic field and high frequency radio waves. MRI is based on the ability of a small number of protons within the body to absorb and emit radio wave energy when the body is placed within a strong magnetic field. Different tissues absorb and release radio wave energy at different, detectable, and characteristic rates.

MRI is used for suspected neurogenic tumors (meningioma, glioma), vascular disorders, and nonorganic, non-metallic foreign bodies (fig. 2.27).

In the presence of magnetic intraocular foreign bodies (IOFB) it can be positively harmful. The application of powerful magnetic field can move the magnetic IOFB and damage the intraocular structures.

Though more advanced imaging modalities (CT, MRI) are available, however,

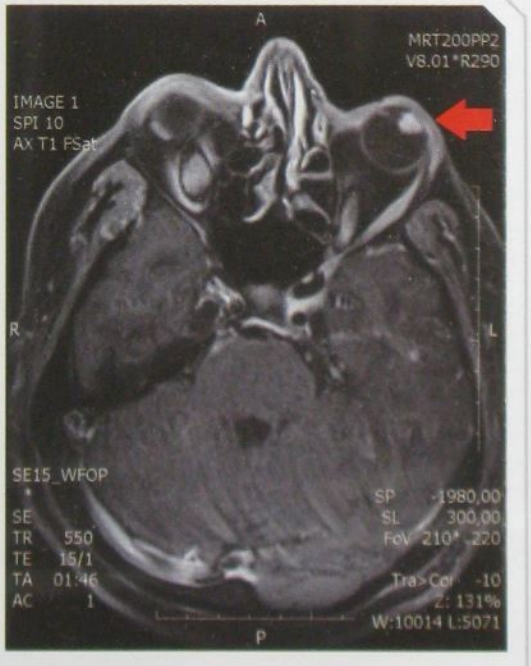


Fig. 2.27. Melanoma of the left eye (shown by the red arrow) revealed by MRI

plain radiographs of the eye and orbit can still be essential for detecting lacrimal drainage pathway pathology, orbital fracture and intraocular/intraorbital foreign body localization.

NOTE!

MRI is contraindicated if metal foreign bodies are suspected.

Dacryocystography is a radiographic visualization of the lacrimal sac and lacrimal drainage system after injection of a contrast medium. To perform dacryocystography a radiographic contrast material is pushed into the lacrimal sac with the help of a lacrimal cannula and X-rays are taken after 5—7 minutes and 15—20 minutes to visualize the entire passage.

Dacryocystography is used to determine the exact site of the mechanical blockage of the lacrimal drainage passageways. It tells also the nature and extent of a block. In addition, it gives information about the mucosa of the sac, presence of any fistulae, diverticulae, stones or suspected tumor in the sac.

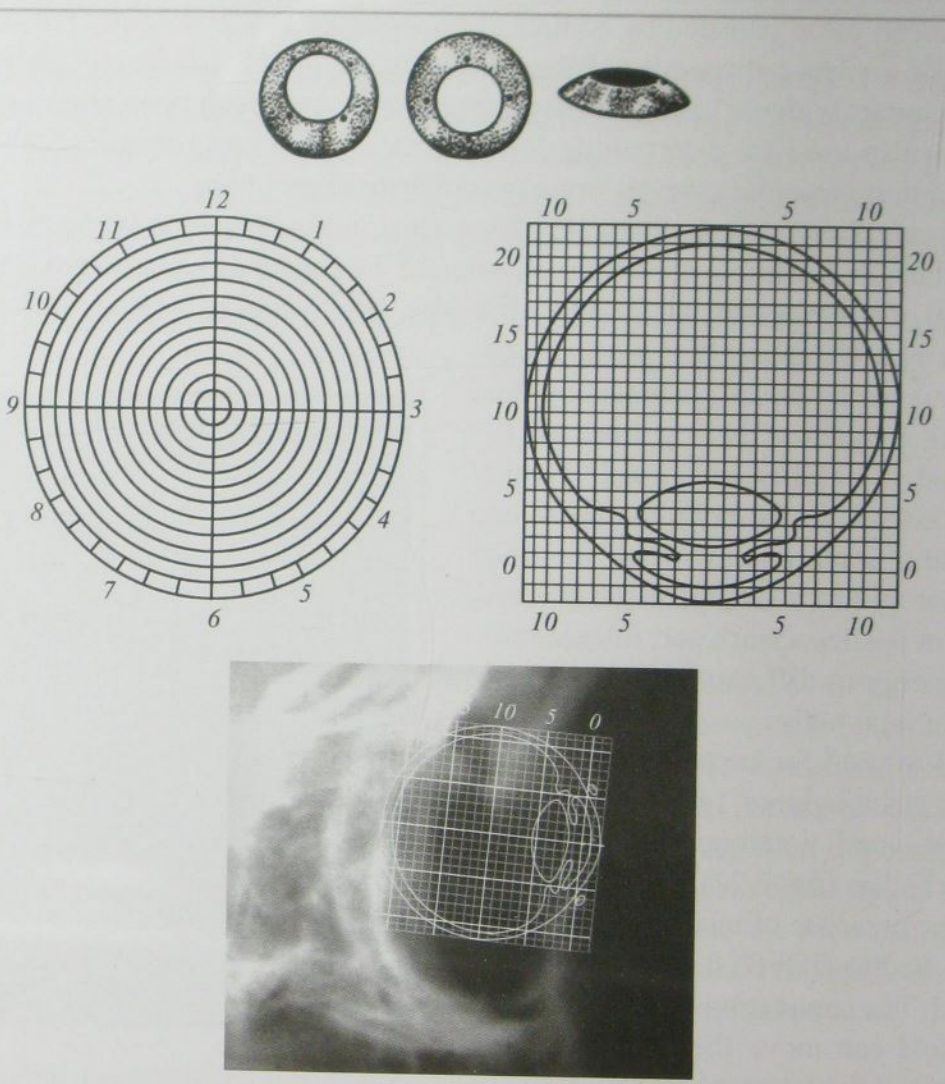


Fig. 2.28. Prosthesis—indicators of Comberg—Baltin with measuring schemes (by E.I. Kovalevskiy) and the example of their use at foreign body localisation

X-Ray Localization of an Intraocular Foreign Body

Patients with suspected penetrating injury of the eye are supposed to be conducted X-ray review of the orbit in order to localize radiopaque (usually metallic) intraocular foreign bodies. For this purpose it is used the *Comberg—Baltin* method of X-ray localization which consists in the use of an aluminium prosthesis — a ring-shaped indicator of 5 mm a with curvature radius corresponding to the sclera curvature with an opening of 11 mm in the center. At a distance of 0.5 mm from the opening edge 4 lead points are pressed into the ring, located on the inter-perpendicular meridians. After instillation of anesthetic drops this prosthesis indicator is put on the eye so that the lead markers correspond to the limbus for 12, 3, 6, and 9 hours. Two X-ray films are made in direct and side projections. The first film determines the meridian where the foreign body is located as well as its distance from the anatomical axis of the eye. The second film helps to establish the distance from the foreign body to the limbus. The exact localization of the foreign body is determined with the help of special measuring schemes and special tables with placing them on the top of X-ray film of a patient (fig. 2.28).

Skeleton-free roentgenography by Fogte is used to diagnose fine foreign bodies in the anterior part of the eye including non-metallic ones (glass, stone).

Review:

1. Key Points

The ophthalmic examination is always preceded by a detailed history, which can help to determine any symptoms the individual is experiencing, and to suggest the disease and its cause.

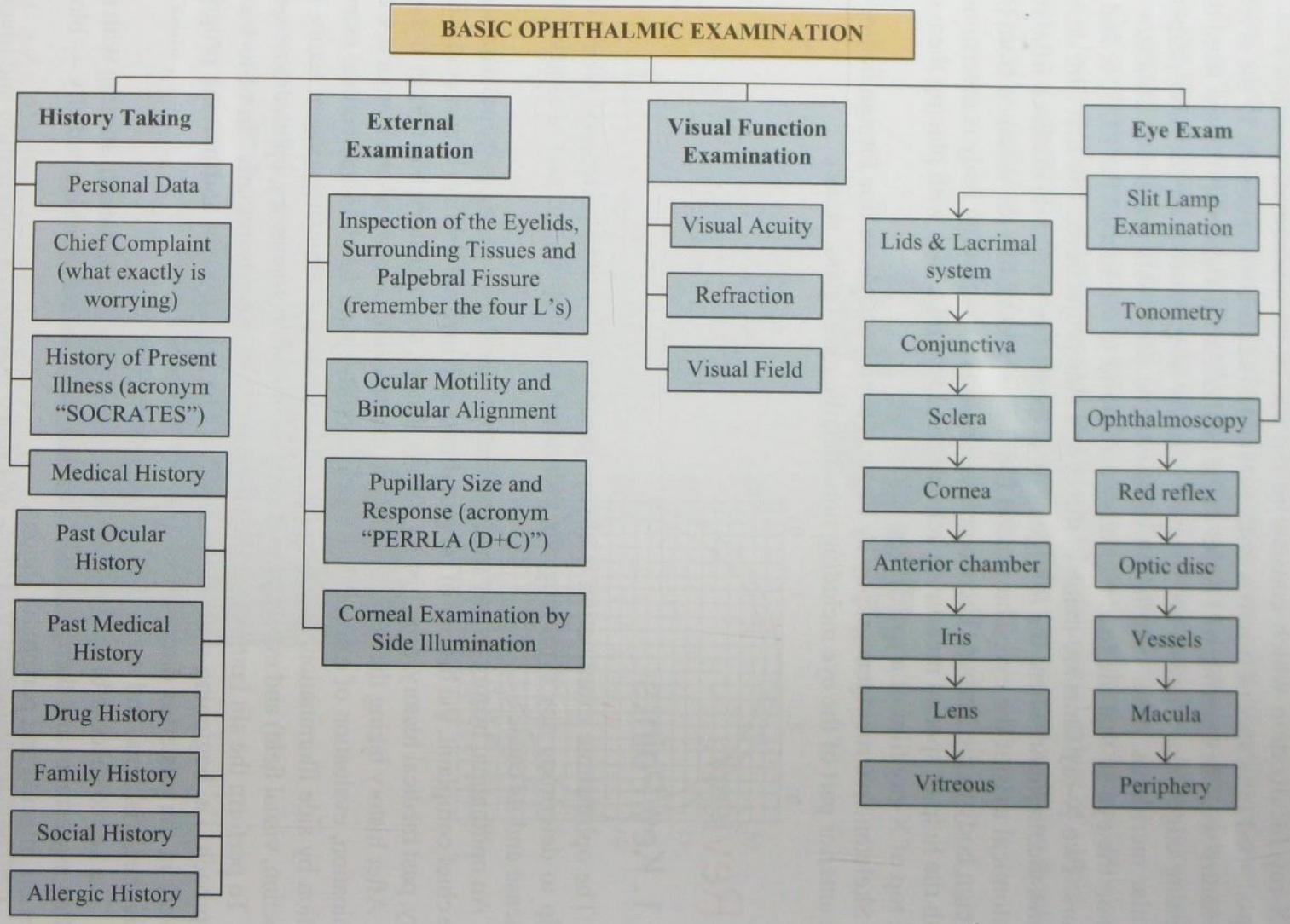
An ophthalmic history as any medical history starts from taking personal data, then the chief complaint, the history of present illness and medical history — past ocular history, past medical history, drug history, family history, social history, and allergy history.

After history taking the basic ophthalmic examination consists of an external eye examination, evaluation of extraocular muscle motility, pupil function and corneal examination by side illumination, followed by visual function examination (visual acuity, refraction, visual field) and eye exam (slit lamp examination, tonometry, ophthalmoscopy).

To perform the slit lamp exam the examiner must use a systematic "front-to-back" approach, i.e. beginning with the eyelids and lacrimal system and moving progressively in the posterior direction to examine the conjunctiva, sclera, cornea, anterior chamber, iris, lens, and anterior vitreous cavity.

The key to successful fundus examination is systematic approach, starting with red reflex assessment and then proceeding to the optic disc and working radially — blood vessels, macula and peripheral fundus.

According to symptoms and signs additional methods of examination and investigative techniques can be performed.

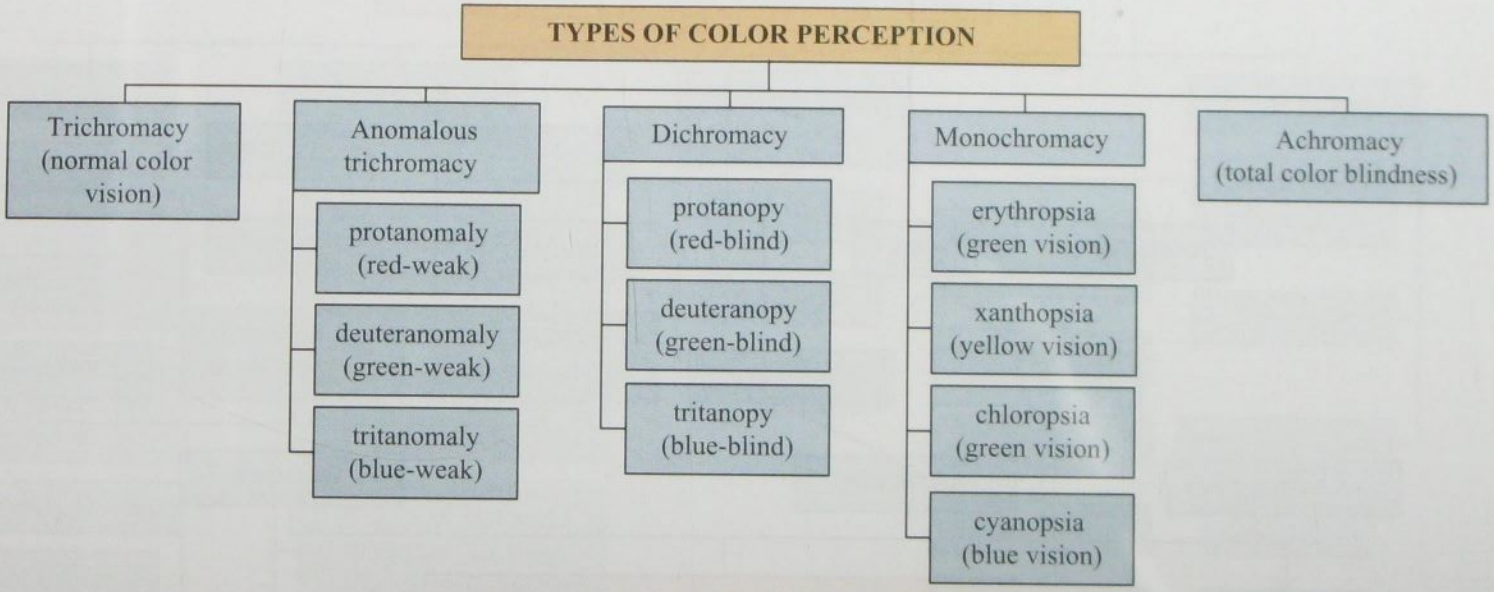

BASIC OPHTHALMIC EXAMINATION
History Taking
Personal Data
Chief Complaint (what exactly is worrying)
History of Present Illness (acronym "SOCRATES")
Medical History
Past Ocular History
Past Medical History
Drug History
Family History
Social History
Allergic History
External Examination
Inspection of the Eyelids, Surrounding Tissues and Palpebral Fissure (remember the four L's)
Ocular Motility and Binocular Alignment
Pupillary Size and Response (acronym "PERRLA (D+C)")
Corneal Examination by Side Illumination
Visual Function Examination
Visual Acuity
Refraction
Visual Field
Eye Exam
Slit Lamp Examination
Lids & Lacrimal system
Conjunctiva
Sclera
Cornea
Anterior chamber
Iris
Lens
Vitreous
Tonometry
Ophthalmoscopy
Red reflex
Optic disc
Vessels
Macula
Periphery

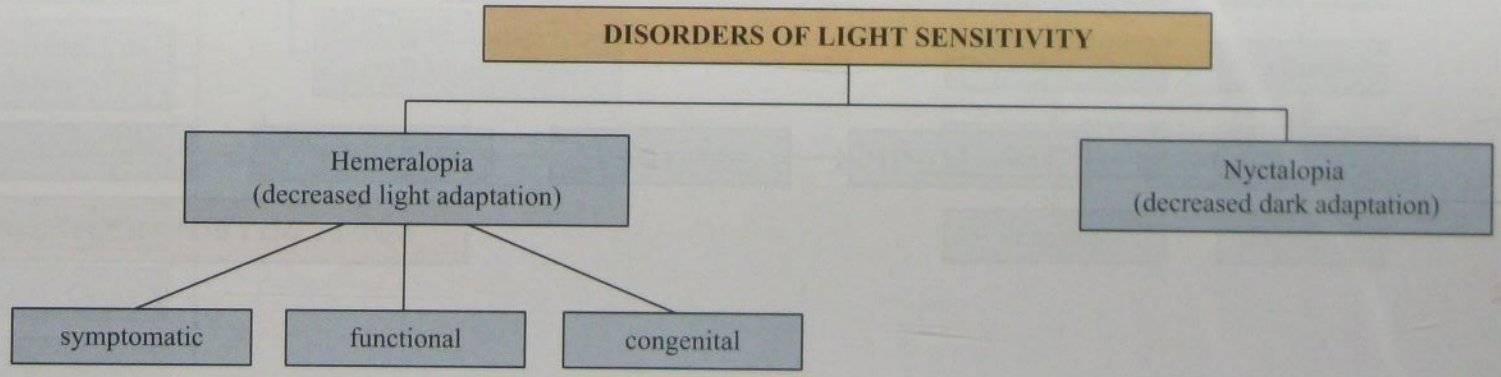
HIERARCHY FOR LOW VISION

VA by a chart → Counting Fingers → Hand Motion → Light Perception → No Light Perception

DEFECTS OF THE VISUAL FIELD

- Scotomas
  - by localization
    - central
    - paracentral
    - peripheral
  - by the degree of sensitivity loss
    - absolute
    - relative
  - by complaints availability
    - positive
    - negative
- Hemianopia
  - homonymous
  - heteronymous
    - binasal
    - bitemporal
- Quadrantanopia


TYPES OF COLOR PERCEPTION
Trichromacy (normal color vision)
Anomalous trichromacy
protanomaly (red-weak)
deuteranomaly (green-weak)
tritanomaly (blue-weak)
Dichromacy
protanopy (red-blind)
deuteranopy (green-blind)
tritanopy (blue-blind)
Monochromacy
erythropsia (green vision)
xanthopsia (yellow vision)
chloropsia (green vision)
cyanopsia (blue vision)
Achromacy (total color blindness)


DISORDERS OF LIGHT SENSITIVITY
Hemeralopia (decreased light adaptation)
symptomatic
functional
congenital
Nyctalopia (decreased dark adaptation)

3. The Review Questions

A. Control Questions

1. What information is essential in history taking?
2. Name steps of basic ophthalmic investigation.
3. What does pupillary function examination include?
4. What is visual acuity and how is it calculated?
5. What are the methods of visual acuity examination?
6. How is to investigate visual acuity in cases of its significant loss?
7. What is visual field and methods of examination?
8. Name the defects of the visual field and their types.
9. What is the main approach of slit lamp eye exam?
10. What is tonometry, what types of tonometry do you know?
11. What is ophthalmoscopy? What types of ophthalmoscopy do you know?
12. What is the main approach of macula examination?
13. What additional methods of corneal examination do you know?
14. What additional methods of central visual field examination do you know?
15. What is color vision? What types of color vision do you know?
16. What methods are used to diagnose color vision defects?
17. What is binocular vision? What are the basic conditions for binocular vision?
18. What methods of binocular vision investigation do you know?
19. What is light sensitivity? What types of eye adaptation to different illumination do you know?
20. What kinds of insufficiency of adaptation to different levels of light intensity do you know?
21. What diagnostic methods of dark adaptation do you know?
22. What techniques of fundus investigation do you know?
23. What investigative techniques can be used in cases of ocular trauma?

B. Tests

1. **Visual acuity is:**
   - A. The ability to see at different distances
   - B. Simultaneous vision with two eyes
   - C. The ability to separate points located at the minimal distance from one another
   - D. The ability to distinguish colors
   - E. The ability of an eye to perceive light of different intensities
   - F. The space seen by an eye while it looks straight ahead

2. **Visual acuity can be examined by means of:**
   - A. Ophthalmoscope
   - B. Perimeter
   - C. Tables or charts
   - D. Phoropter
   - E. Tonometer
   - F. Slit lamp

3. **Visual acuity 1.0 is:**
   A. Reading the 1st line of the Golovin—Sivtsev table
   B. Reading the 10th line of the Golovin—Sivtsev table
   C. Reading the 12th line of the Golovin—Sivtsev table
   D. Distinguishing two points separately, seen at an angle 1 minute
   E. Count fingers from a distance of 5 m
   F. Inability to see any light

4. **A patient reads the 1st line of the Golovin—Sivtsev table from a distance of 3 m. His visual acuity is:**
   A. 0.1
   B. 0.3
   C. 0.03
   D. 0.06
   E. 0.6
   F. 0.8

5. **A patient reads the 8th line of the Golovin—Sivtsev table making 3 mistakes. His visual acuity is:**
   A. 1.0
   B. 0.8
   C. 0.7
   D. 0.08
   E. 0.07
   F. 0.3

6. **Peripheral vision or visual field is:**
   A. The ability to see at different distances
   B. Simultaneous vision with two eyes
   C. The ability to separate points located at the minimal distance from one another
   D. The ability to distinguish colors
   E. The ability of an eye to perceive light of different intensities
   F. The space seen by an eye while it looks straight ahead

7. **The methods of visual field examination are:**
   A. Ophthalmoscopy
   B. Perimetry
   C. Biometry
   D. Confrontation test
   E. Adaptometry
   F. Campimetry

8. **The pathological changes of visual field are:**
   A. Hemeralopia
   B. Hemianopia
   C. Dichromasia
   D. Quadrantanopia
   E. Scotoma
   F. Nyctalopia

9. **The normal values of visual field borders are:**
   A. Nasally — 60°
   B. Nasally — 35°
   C. Up — 60°
   D. Down — 75°
   E. Temporally — 180°
   F. Temporally — 90°

10. **The main approach to slit lamp examination:**
   A. Anterior vitreous cavity → sclera → lens → anterior chamber → cornea → conjunctiva → iris → lacrimal system and eyelids
   B. Eyelids and lacrimal system → conjunctiva → sclera → cornea → anterior chamber → iris → lens → anterior vitreous cavity
   C. Anterior vitreous cavity → lens → iris → anterior chamber →

cornea → sclera → conjunctiva → lacrimal system and eyelids

D. Cornea → sclera → anterior vitreous cavity →eyelids and lacrimal system → conjunctiva → anterior chamber → iris → lens

E. Eyelids and lacrimal system → lens → sclera → cornea → anterior vitreous cavity → conjunctiva → anterior chamber → iris

F. Sclera → cornea → eyelids and lacrimal system → conjunctiva → anterior chamber → iris → lens → anterior vitreous cavity

11. The methods of corneal investigation are:

A. Corneal staining
B. Keratometry
C. Corneal topography
D. Aberrometry
E. Pachymetry
F. Campimetry

12. The anterior chamber angle is studied by:

A. Slit lamp
B. Indirect ophthalmoscopy
C. Direct ophthalmoscopy
D. Gonioscopy
E. Ultrasonography
F. Pentacam

13. The methods of fundus examination are:

A. Campimetry
B. Direct ophthalmoscopy.
C. Intravenous fluorescein angiography
D. Funduscopy
E. Indirect ophthalmoscopy
F. Adaptometry

14. The methods of intraocular pressure measurement are:

A. Control method
B. Tonometry
C. Ophthalmoscopy
D. Topography
E. Palpation
F. Gonioscopy

15. The normal values of intraocular pressure are:

A. Measured by Maklakov tonometry — 18—26 mm Hg
B. Measured by Maklakov tonometry — 10—21 mm Hg
C. Measured by Goldmann tonometry — 20—25 mm Hg
D. Measured by Goldmann tonometry — 10—21 mm Hg
E. Measured by pneumato-nometry — 18—26 mm Hg
F. Measured by pneumato-nometry — 10—21 mm Hg

16. Color vision is:

A. The ability to see at different distances
B. Simultaneous vision with two eyes
C. The ability to separate points located at the minimal distance from one another
D. The ability to distinguish colors
E. The ability of an eye to perceive light of different intensities
F. The space seen by an eye while it looks straight ahead

17. Color vision is a function of:

A. Rods only
B. Rods and cones
C. Cones only
D. Optic disc

E. Macula
F. Retinal periphery

18. Color vision can be examined by means of:
A. Optotypes
B. Anomaloscope
C. Adaptometer
D. Polychromatic tables
E. Slit lamp
F. Arrangement test

19. Color vision defects are called:
A. Trichromacy
B. Anomalous trichromacy
C. Dichromacy
D. Monochromacy
E. Achromacy
F. Nyctalopia

20. Binocular vision is:
A. The ability to see at different distances
B. Simultaneous vision with two eyes
C. The ability to separate points located in the minimal distance from one another
D. The ability to distinguish colors
E. The ability of an eye to perceive the light of different intensities
F. The space seen by an eye while it looks straight ahead

21. The methods of binocular vision examination are:
A. Orientation method
B. Two pencils test
C. Test with "a hole in a palm"
D. Adaptometry
E. 4-dot color test
F. Bagolini striated glass test

22. Light perception is:
A. The ability to see at different distances
B. Simultaneous vision with two eyes
C. The ability to separate points located at the minimal distance from one another
D. The ability to distinguish colors
E. The ability of an eye to perceive light of different intensities
F. The space seen by an eye while it looks straight ahead

23. The methods of dark adaptation examination are:
A. Orientation method
B. Two pencils test
C. Control method of Kravkova—Purkinje
D. Adaptometry
E. Sokolov test
F. Bagolini striated glass test

24. Insufficiency of adaptation to different levels of light intensity can be:
A. Hemeralopia
B. Anomalous trichromacy
C. Dichromacy
D. Monochromacy
E. Achromacy
F. Nyctalopia

Appendix

Example of Ophthalmic History and Examination Form

Ophthalmic History Form

Date: _____

Name: _____
(Last) (First) (Middle)

Date of Birth: _____ Age: _____ Sex: ☐ Male / ☐ Female

Address_____ Phone (_____)_____

CHIEF COMPLAINT

What is the main reason for the visit: ☐ Routine exam; ☐ Follow-up visit
or any of the following symptoms:

☐ Loss of vision;	☐ Burning, itching;	☐ Redness;
☐ Blurred or fuzzy vision;	☐ Foreign body sensation;	☐ Flashing lights;
☐ Double vision;	☐ Tearing;	☐ Cobwebs, dark spots;
☐ Eye pain/soreness;	☐ Discharge;	☐ Problems with glasses;

☐ Other: _____

HISTORY OF PRESENT ILLNESS

Site/location	*Which eye has a problem?*	☐ Right ☐ Left ☐ Both
Onset	*How did it start?*	☐ Gradual ☐ Sudden
Character	*How is affecting you?*	☐ Bothersome ☐ Aware ☐ Painful
Radiation	*Does the pain radiate?*	☐ Forehead ☐ Temple ☐ Other _____
Associations	*Is there associated symptoms?*	☐ Headache ☐ Vomiting ☐ Other _____
	Do you associate the problem with...	☐ Infection ☐ Injury ☐ Other _____
Time course	*How long does the problem last?*	_____
	Is it new, ongoing, returning?	☐ New ☐ Ongoing ☐ Returning
Exacerbating	*Does anything make the problem better or worse?*	☐ Drops ☐ Medication ☐ Other _____
Severity	*How severe is the problem?*	☐ Mild ☐ Moderate ☐ Severe

PAST OCULAR HISTORY

Have you ever been diagnosed with any of the following and which eye?

☐ Cataract	☐ Glaucoma	☐ Myopia	☐ Retinal disorders
☐ Corneal disease	☐ Lazy eye	☐ Macular degeneration	☐ Other _____

Do you wear glasses? ☐ No ☐ Yes
If Yes, how long and dioptric power _____
Do you wear contacts? ☐ No ☐ Yes
If Yes, how long, what type and dioptric power _____
Have you had ocular surgery? ☐ No ☐ Yes

Cataract	Date _____	Type _____	Which eye _____
Glaucoma	Date _____	Type _____	Which eye _____
Laser surgery	Date _____	Type _____	Which eye _____

Other _____
Did you have any ocular injury? ☐ No ☐ Yes

Date _____	Type _____	Which eye _____
Date _____	Type _____	Which eye _____

PAST MEDICAL HISTORY

Have you had any serious medical problems?

☐ Autoimmune diseases	☐ Diabetes	☐ Heart disease	☐ Hypertension
☐ Kidney disease	☐ Lung disease	☐ Stroke	☐ Thyroid disease

☐ Other _____
Please, list all major surgeries, injuries and/or hospitalizations and dates:
_____
_____
_____

DRUG HISTORY

*Please, list **all** medications, that you are currently taking, including systemic medicattions, eyedrops, vitamins, herbs, minerals and birth control pills:*

Medication	Dose	Frequency	Medication	Dose	Frequency

FAMILY HISTORY

Does anyone in your family (blood relative) have any of the medical or ocular conditions listed below? If yes, please, indicate who (parent, sibling or child).

Blindness ☐ No ☐ Yes, Who _____; Lazy eye ☐ No ☐ Yes, Who _____;
Cataract ☐ No ☐ Yes, Who _____; Glaucoma ☐ No ☐ Yes, Who _____;
Macular degeneration ☐ No ☐ Yes, Who _____;
Retinal detachment ☐ No ☐ Yes, Who _____;
Other eye problems _____
Cancer ☐ No ☐ Yes, Who _____; Heart disease ☐ No ☐ Yes, Who _____;
Diabetes ☐ No ☐ Yes, Who _____; Hypertension ☐ No ☐ Yes, Who _____;
Kidney disease ☐ No ☐ Yes, Who _____; Stroke ☐ No ☐ Yes, Who _____;
Thyroid disease ☐ No ☐ Yes, Who _____; Other diseases _____

SOCIAL HISTORY

Occupation _____ Retired? _____ Other _____
Marital status: ☐ Single ☐ Married ☐ Divorced ☐ Widowed Number of children? ____
☐ Any dietary restrictions ☐ Pregnancy
☐ Smoking, how often _____; ☐ Alcohol use, how often _____; ☐ Drug use/abuse
☐ Driving ☐ Hobbies/sports _____

ALLERGIC HISTORY

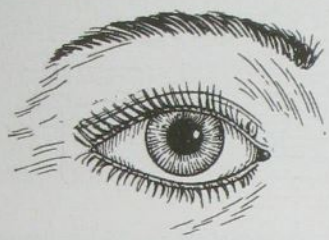
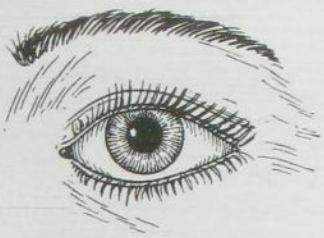
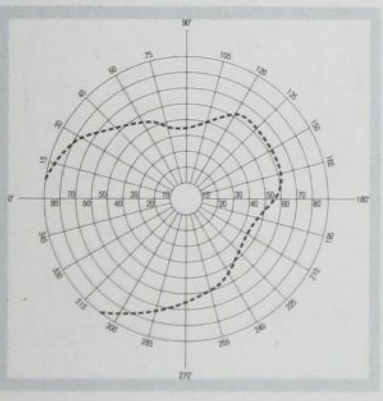
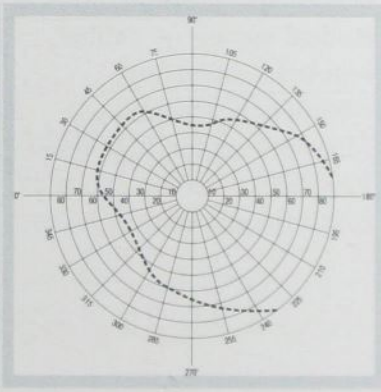
☐ Drug allergies, if yes, please, list: _____
_____
_____
☐ Seasonal allergies _____ ☐ Food allergies _____
☐ Allergies to dust/mold/ animal hair _____ ☐ Other _____

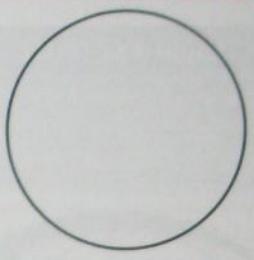
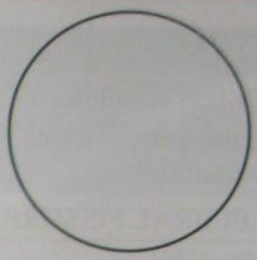
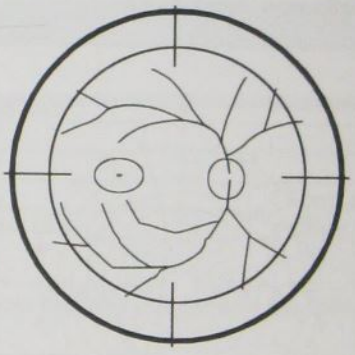
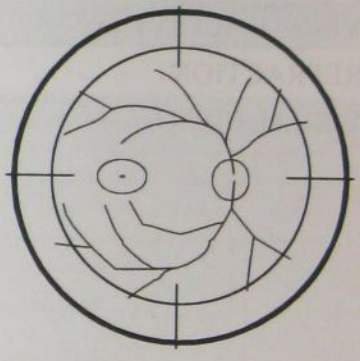
REWIEV OF SYSTEMS

Do you currently, or have you ever had any problems in the following areas? Please, underline and specify.

Cardiovascular (hest pain, heart failure, arrhythmia, hypertension, blood clots) _____
Constitutional (fever, weight loss, night sweats or chills, excess thirst) _____
Dermatological (eczema, psoriasis, rosacea) _____
Endocrine (hormonal dysfunction, diabetes, thyroid dysfunction) _____
Gastrointestinal (colitis, diarrhea, constipation, ulcer, hepatitis) _____
Genitourinary (kidney or bladder disease, prostate, infections) _____
Hematologic/Lymphatic (anemia, bleeding disorder, transfusions) _____
Immunologic (Sjogren's syndrome, herpes zoster, HIV or AIDS) _____
Musculoskeletal (muscular pains or distrophy, arthritis, joint pain, osteoporosis) _____
Neurologic (headaches, migraines, dizziness. seizures) _____
Otorhinolaryngological (hearing loss, sinusitis, laryngitis) _____
Psychiatric (anxiety, depression, insomnia) _____
Respiratory (shortness of breath, bronchitis, pneumonia, asthma, emphysema, tuberculosis)
_____
_____

Ophthalmic Examination Form

	Right Eye (OD)	Left Eye (OS)
EYELIDS (condition of skin, margin, position, growth of lashes)		
PALPEBRAL FISSURE (shape, size)		
LACRIMAL APPARATUS (state, function)		
LYMPH NODES		
EYEBALL (position in the orbit, motility, convergence)		
PUPILLARY RESPONSE (size, equality, form, reaction to light)		
VISUAL ACUITY		
REFRACTION		
VISUAL FIELD		
COLOR VISION		
BINOCULAR VISION		
LIGHNT/DARK ADAPTATION		
CONJUNCTIVA (transparency, color, vessels)		
SCLERA (color, vessels)		
LIMBUS (width, changes)		

	Right Eye (OD)	Left Eye (OS)
CORNEA (transparency, size, form, sensitivity)		
ANTERIOR CHAMBER (transparency of intraocular fluid, depth)		
IRIS (color, picture, form)		
LENS (transparency, position)		
VITREOUS (transparency)		
CILLIARY BODY (pain, sensitivity)		
IOP		
RED REFLEX (color, brightness, regularity)		
FUNDUS Optic nerve disk — color, size, shape, position. Retinal vessels — caliber, regularity, curvature, obstruction, a/v ratio. Macula — color, light reflex. Periphery — color, exudates, hemorrhages, tears or holes.	 Optic disk _____ Retinal vessels _____ Macula _____ Periphery _____	 Optic disk _____ Retinal vessels _____ Macula _____ Periphery _____

CHAPTER 3

Optical System of the Eye. Refractive Errors, Accommodation and Amblyopia

OBJECTIVES

As a primary care physician, you should be able to estimate visual acuity function and recognize the signs and symptoms of ametropia, amblyopia and strabismus; be able to perform the necessary tests to screen for these conditions; and, if the patient is a child, be aware of the need to arrange for prompt ophthalmologic consultation, particularly when intraocular disease is suspected.

To achieve these objectives, you should learn how to:

- measure or estimate visual acuity in children and adults;
- detect ametropias such as myopia, hyperopia and astigmatism;
- understand phenomena of accommodation and presbyopia;
- perform ophthalmoscopy in a child to rule out any organic causes of impaired vision when amblyopia is suspected;
- explain to parents the need for prompt treatment of amblyopia.
- perform vision distance and near acuity testing.

Upon completion of the chapter the student should be able to:

- know the main structures of the optical system of the eye and the principles of its functioning;
- know the types of clinical refraction;
- know the main refractive errors and methods of their correction;
- know the basic diagnostic methods for refractive errors estimation;
- describe the clinical sings and complications of myopia;
- evaluate and manage patients with refractive errors;
- know the principles of refractive errors treatment.

Plan:

1. Optical System of the Eye and Its Structural Elements

The human eye is a complex biological device that has structural elements and is functioning like a photo-camera. The eye refracts and focuses incoming light rays from external objects and forms an image on the retina, thus enabling the human ability to see. There are four transparent media in the eye:the cornea, the anterior chamber, the lens, and the vitreous (fig. 3.1).

The cornea is a transparent "window" in the front part of the eyeball that looks like a glass covering an old-style watch. The cornea has an optic power of nearly 42.00—45.00 D. The cornea has two main mportant functions: it protects the internal eye structures and refracts incoming light rays. Then part of refracted light is entering through the pupil — a central circular opening in the center of the iris that acts like the diaphragm in the camera, automatically adjusting the amount of light needed for optimal retinal image quality. Under bright light conditions the pupil is constricted, and under dim light — dilated.

After passing through the pupil, light is coming to the *crystalline lens* that is attached by zonules to the ciliary muscles. The lens of the human eye acts like the lens of a camera, providing further focusing of the incoming rays of light. In contrast to the camera lens, the crystalline lens is changing the focusing distance by changing its power due to changes of the size and curvature, thus providing "fine-tuning" of image sharpness on the retina surface. Changes of the lens curvature are possible due to contraction of the ciliary muscles and corresponding changes of traction of the zonules on the surface of the lens. The average optical power of the human crystalline lens is 21.50 D.

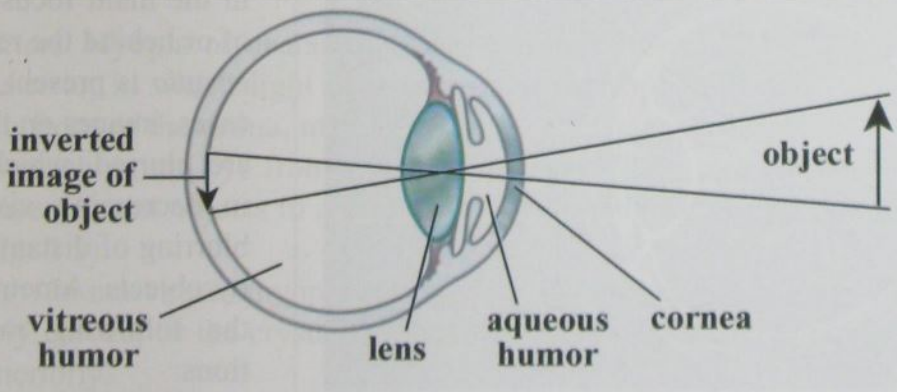


Fig. 3.1. Optical system of the eye*

* Figures 3.1—3.7 are taken from *Анатомія людини*: у 3 т. Т. 2 / А.С. Головацький, В.Г. Черкасов, М.Р. Сапін та ін. — Вид. 4. — Вінниця: Нова Книга, 2009. — С. 371—397.

The aqueous and *vitreous* do not have any refractive power of consequence.

Every mentioned part of the eye has a specific role in the focusing of the incoming light rays on the retina, thus enabling humans to see surrounding objects clearly.

2. Refraction and Classification of Its Types

Refraction is bending of light rays when passing from one medium into another of different density and is measured in diopters (D) which describes the power that a structure has to focus parallel rays of light. The higher this value, the stronger its focusing ability.The refractive power of an eye is determined by the refractive power of transparent media and its normal average value in newborns is about 80.0 D, in adults — 60—65 D but can vary within 52.0—68.0 D. It is *physical refraction* of the eye. In clinical presentation the position of the main focus relative to the retina has a great significance for refractive power of the eye. This determines the concept of *clinical refraction* — correlation between physical refraction and the axial length of the eye (which is normally about 24 mm).

According to the position of the focus relative to the retina two types of clinical refraction are possible. The first is *emmetropia* — the condition when the parallel light rays, having refracted, focus directly on the retina; it is a proportional refraction that gives a clear view of the object we are looking at (fig. 3.2).

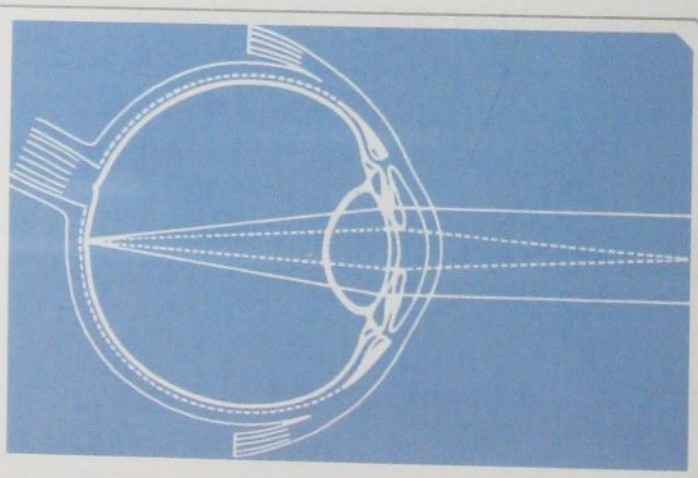


Fig. 3.2. Emmetropia (focal point is directly on the retina, image is clear)

If the parallel rays, having refracted, concentrate in the main focus in front of or behind the retina *ametropia* is present. In such cases, images on the retina are blurred, which results in decreasing vision and blurring of distant or nearby objects. Ametropia has the following manifestations:

— myopia;
— hyperopia;
— astigmatism.

3. Examination Methods

Visual acuity test:
— Distance VA Testing
— Near VA Testing
Refraction Test:
— Subjective Refraction Testing — by the kind of corrective lenses
— Objective Refraction Testing — skiascopy, retinoscopy, refractometry

4. Refractive Errors

Refractive errors or ametropia may be caused by the following factors:
— the axial length of the eye (shortening or lengthening of the eye);
— refractive index of the eye media due to changes in the shape of the cornea, or aging of the lens;
— integrity and transparency of the eye structures.

4.1. Myopia

Definition. Myopia or nearsightedness (strong clinical refraction) is a type of refractive error, in which light rays from the distant objects entering the eye are focused in front of the retina, making distant objects appear out of focus. Only the light rays reaching the eye from a short distance are refracted correctly. This condition causes distant objects to appear blurred, while close objects can still be seen clearly (fig. 3.3).

Etiology. In most cases myopia occurs when the eyeball is too long, the cornea is excessively curved, or the crystalline lens has exsessive refractive power, or is displaced anteriorly.

Clinical Picture. Myopic refractive changes in most cases happen during childhood due to growing of the eyeball. Uncorrected distance vision acuity decreases slowly during adolescence and normally stabilizes during the third decade of life.

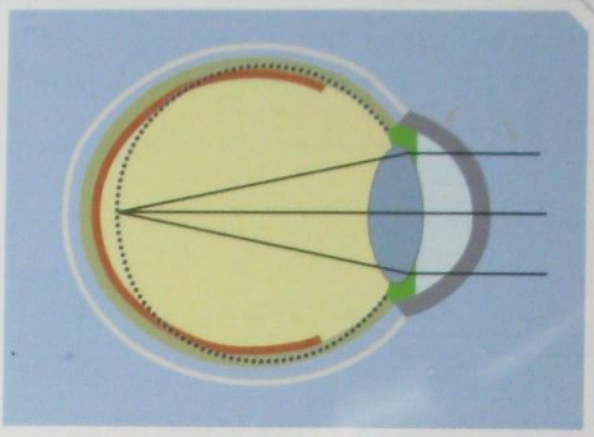


Fig. 3.3. Myopia (focal point is in front of the retina, image is blurry)

Myopia can be of three grades according to the focusing power of the corrective lens, which is measured in diopters (D):

— mild: up to –3.0 D;
— moderate ranges from –3.25 D to –6.0 D;
— severe/high: –6.25 D and over.

Complaints. Patients complain of blurred vision, difficulties in seeing distant objects, recognizing faces and driving. Other persistent symptoms: the patient needs to squint or close the eyelids to see clearly, has headaches, eyestrain. Closer objects are seen clearly.

Signs. Steep cornea, deep anterior chamber, long eye.

Treatment. No established effective therapeutic treatment is available. Correction of vision acuity by glasses, contact lenses, or refractive surgery. Optical correction is provided by divergent (negative) glasses that make distant objects seen clearly. Young myope with normal accommodation will also see clearly close objects at 30 cm through the same corrective glasses.

Prognosis. Usually good, except degenerative myopia (axial length more than 26.0 mm), because vision-threatening conditions are more common.

Complications. Myopic patients are at higher risk of such vision-threatening conditions as maculopathy, retinoschisis, retinal detachment or open-angle glaucoma.

Prophylaxis. Unknown.

4.2. Hyperopia

Definition. Hyperopia or far-sightedness, hypermetropia (weak clinical refraction) is a type of refractive error, in which light rays entering the eye focus behind the retina. This condition causes people with hyperopia see distant objects very well, but have difficulty focusing on objects that are up close (fig. 3.4).

Etiology. In most cases hyperopia occurs when the eyeball is too short, or the cornea is too flat, the lens sits farther back in the eye than normal.

Clinical Picture. Hyperopia mostly develops during childhood. In most mild cases, hyperopia recedes and disappears before adulthood due to elongation of the eye during teen age.

Hyperopia can be of three grades according to the focusing power of the corrective lens, which is measured in diopters (D):

— low: up to +2.0 D;
— moderate ranges from +2.25 D to +5.0 D;
— high: +5.25 D and over.

Complaints. A patient with hyperopia is not able to see close objects clearly and it is why this condition is probably more annoying to a person than myopia. Other symptoms can include eyestrain, aching eyes, squinting, fatigue and headaches.

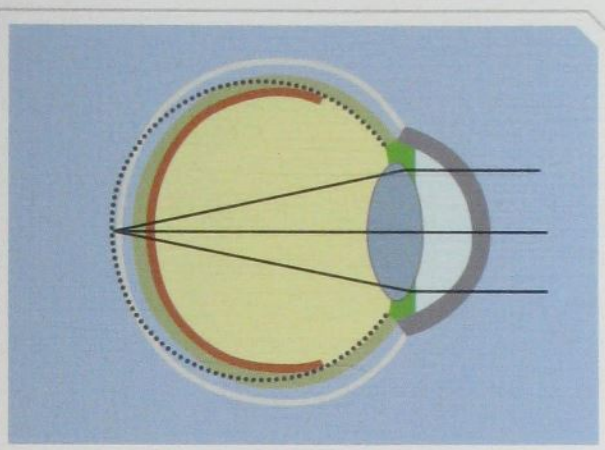


Fig. 3.4. Hyperopia (focal point is behind the retina, image is blurry)

Signs. Flat cornea, shallow anterior chamber, short eye.

Treatment. No established effective therapeutic treatment is available. Correction of vision acuity with glasses, contact lenses, or refractive surgery. Optical correction is provided by converging (positive) lenses that make distant objects seen clearly. Young hyperope with normal accommodation will also see near objects clearly at 30 cm through the same corrective glasses.

Prognosis. Usually good, except severe hyperopia (axial length less than 21.0 mm), because vision-threatening conditions are more common.

Complications. Children with severe hyperopia can develop strabismus or amblyopia, which leads to a slow decrease of vision due to insufficient communication between the retina and the cortical centers of the brain. Hyperopic patients after 35 years of age are at higher risk of such vision-threatening condition, as angle-closure glaucoma.

Prophylaxis. Unknown.

4.3. Astigmatism

Definition. Astigmatism is a type of refractive error of the eye when light rays entering the eye from different directions are focused on different focal points, so multiple focus points occur, either in front of the retina or behind it (or both), causing images both near and far to appear distorted (fig. 3.5).

Etiology. In the vast majority of cases, astigmatism is a result of the shape of the cornea (corneal astigmatism) that is present from birth. Normally, human cornea has a sphere-like, regular

EYE FACTS

Most people have some degree of astigmatism, but it is often not severe enough to affect vision noticeably.

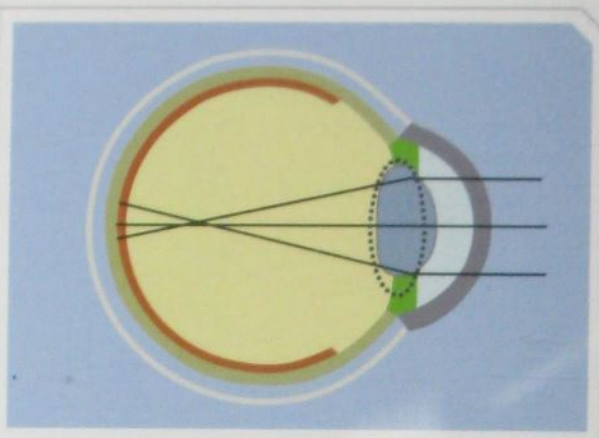


Fig. 3.5. Astigmatism (multiple focal points are on or before the retina, image is distorted)

and smooth shape in all directions. Astigmatism usually is caused by an irregularly shaped cornea. Instead of the cornea having a symmetrically round shape (like a baseball), it is shaped more like a football, with one meridian being significantly more curved than the meridian perpendicular to it. Certain types of surgery or eye injuries that cause scarring of the cornea may result in astigmatism. Keratoconus, a degenerative disorder of the eye where the cornea gradually thins and changes to a more conical shape, can also cause astigmatism.

Sometimes, the crystalline lens may have irregular curvature — lenticular astigmatism.

There are three primary types of astigmatism:

— myopic astigmatism — one or both principal meridians of the eye are nearsighted;
— hyperopic astigmatism — one or both principal meridians are farsighted;
— mixed astigmatism — one prinicipal meridian is nearsighted, and the other is farsighted.

Astigmatism is also classified as regular or irregular. In regular astigmatism, the principal meridians are 90 degrees apart (perpendicular to each other). In irregular astigmatism, the principal meridians are not perpendicular. Most astigmatism is regular corneal astigmatism. Irregular astigmatism can result from an eye injury that has caused scarring on the cornea, from certain types of eye surgery or from keratoconus, a disease that causes gradual thinning of the cornea.

Clinical Picture. If a person suffers from astigmatism, his/her vision is reduced in quality and blurry at any distance. However, small amounts of astigmatism usually are well tolerated as blinking helps maintain the cornea's symmetrical curvature. In most cases astigmatism is a non-progressive state of the eye.

Complaints. A person with astigmatism is not able to see distant and closer objects clearly, and they may look more blurred than in case of myopia or hyperopia. Significant astigmatism may cause eye strain, headaches, aching eye, squinting, eye fatigue, eye irritation.

Signs. Astigmatic cornea, signs of myopia (myopic astigmatism), or hyperopia (hyperopic astigmatism).

Treatment. No established effective therapeutic treatment is available. Correction of vision acuity with glasses, contact lenses, or refractive surgery. Optical correction is provided by toric lenses for astigmatism correction plus converging (positive) or divergent (negative) lenses for residual sphere that make distant objects clearly seen.

A young astigmatic patient with normal accommodation will also see near objects clearly at 30 cm through the same corrective glasses.

Prognosis. Good, except progressive corneal ectasia, because vision-threatening condition is possible.

Complications. Children with severe astigmatism can develop strabismus or amblyopia, which leads to a slow decrease of vision due to insufficient communication between the retina and the cortical centers of the brain. Sometimes astigmatism can progress very rapidly, and then corneal ectasia, called keratoconus, has to be suspected. This disease is characterised by progressive thinning and conic-like deformation of the central cornea. As a result, vision is significantly reduced, distorted, and if left untreated, can lead to severe vision impairment.

Prophylaxis. Unknown.

5. Accommodation

Accommodation is the ability of the eye to adjust its refractive power by altering the shape of the crystalline lens in order to maintain a clear vision or focus objects located at different distances. This phenomen is realised through changes in the curvature of the crystalline lens achieved by the action of the ciliary muscles. While looking at nearby objects the ciliary muscles contract allowing the zonular fibres to relax, which increases lens convexity (roundness) and correspondingly eye refractive power. When looking at distant objects the ciliary muscles relax and the lens returns to a flatter shape (fig. 3.6). Increasing lens curvature corresponds to a shorter focal length and decreasing — to a longer focal length. Our ability to accommodate is automatic and instantaneous. In a young emetropic person there is no delay in focusing while turning gaze from distant to close objects.

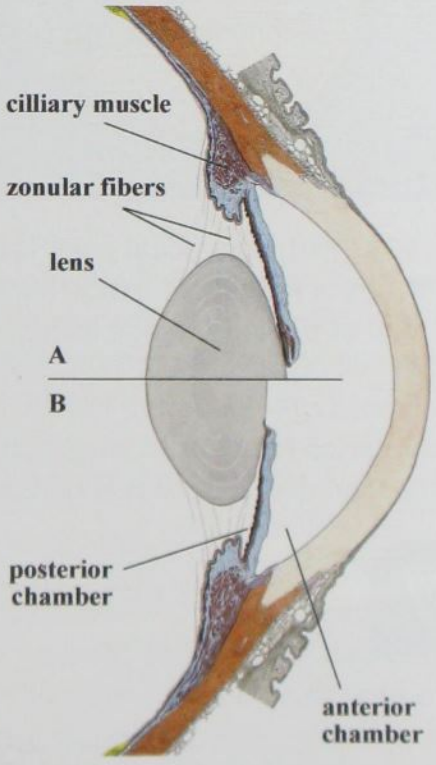


Fig. 3.6. Accommodation; (A) shape of the lens when focusing on the nearby objects and (B) distant objects

6. Presbyopia

Presbyopia is loss of the lens ability to change its shape to focus on near objects due to aging.

Because the crystalline lens hardens with age, changes of the lens curvature become less prominent leading to minimal shifting of the focal length of the eye, thus reducing the eye's ability to focus on close objects (fig. 3.7). It is a natural part of aging. Normally it starts at the age of 40—45 years and continues to worsen until around age 65. Only in rare cases this condition is related to other factors. Trauma and certain diseases, especially a contagious disease called diphtheria, may weaken the muscles in the eye.

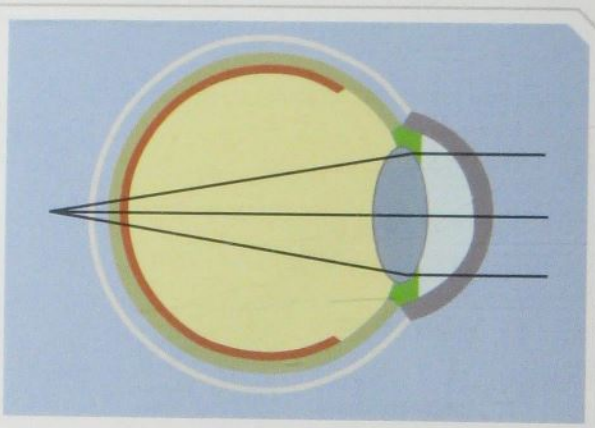


Fig. 3.7. Presbiopia (focal point is behind the retina due to age hardening of the lens, image is blurry)

An emmetrope is facing gradual loss of the ability to focus on nearby objects. Patients may become aware of presbyopia when they start holding books and newspapers at arm's length to be able to read.

Capacity of accommodation at the age of 1 year is 17 D, up to 10 years of age — 14 D, at 40 years of age — 3.5 D, and after 65 years of age less than 1 D. Presbyopia is compensated with reading glasses.

7. Treatment of Refractive Errors

7.1. Optical Correction

For optical correction of ametropia, different types of glasses or contact lenses are used. Dioptric power for optical correction is precisely calculated by an ophthalmologist after complex examination of the refractive state of the eye.

Correction with glasses:

Myopia, hyperopia, and astigmatism can be corrected with special lenses. Myopia is corrected with concave and hyperopia — with convex lenses. These lenses are focusing rays of light on the retina, correcting eye refractive power and correspondingly the existing problems of poor far or near vision.

In case of astigmatism, special cylinder lenses that correct abnormalities of corneal shape are used.

Presbyopia can be corrected with different types of lenses depending on visual tasks of a patient. The simplest solution is convex lenses that are used for near visual task, but need to be removed for distance vision.

Bifocal lenses use two sections — the lower convex part for near vision and the upper part for distance vision.

Progressive lenses make it possible to have good quality vision at all distances using a smooth complex optical surface that employs correction from distance to near without any lens separation.

Correction with contact lenses:

Most cases of ametropia can be easily corrected with contact lenses. Moreover, contact lenses provide better quality of vision than glasses, especially in case of high myopia. There are two types of contact lenses — soft and hard, which use the same principles as glasses. A patient, who wears contact lenses, has to strictly follow the handling and hygiene instructions, otherwise inflammation or infections of the cornea could take place.

7.2. Refractive Surgery

Different types of refractive surgery are used in order to make patients less dependent on glasses and/or contact lenses. Most popular are different techniques of laser vision correction, which work in a way of changing the corneal shape to the amount, which is calculated from patient's refraction. When laser correction of vision is contraindicated (for example, a thin cornea, or a high degree of myopia) an intraocular phakic lens is implanted, or even a clear crystalline lens is extracted with implantation of an intercapsular intraocular lens.

Laser Vision Correction

Photorefractive keratectomy (PRK) consists of removal of the superficial corneal epithelium mechanically, by ethylene oxide or laser, followed by reshaping of the cornea with a computer-controlled laser under surgeon control. At the end of the procedure a contact lens is placed over the cornea in order to aid epithelisation of the central cornea that normally lasts for 3—5 days, after which the contact lens is removed. PRK flattens the cornea in a myopic eye and makes it steeper in a hyperopic eye leading to convergence of light rays on the retina. The LASEK (Laser subepithelial keratectomy), epi-LASIK, transepithelial PRK and ASA (advanced surface abblation) techniques are modern modifications of the PRK technique.

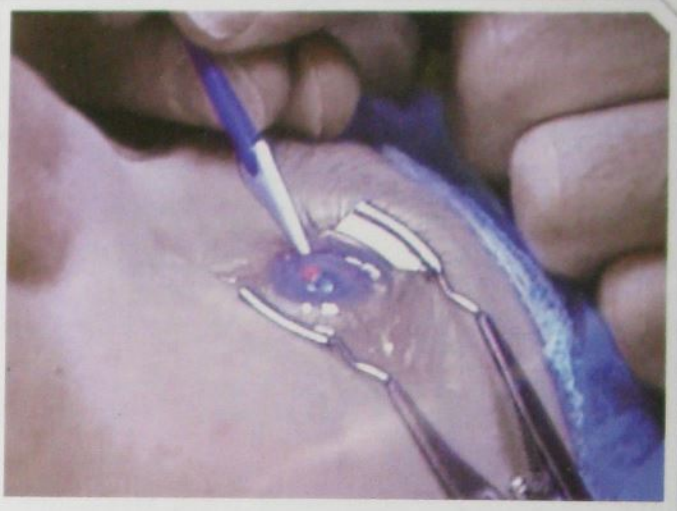


Fig. 3.8. Eximer laser surgery by LASIK technique

Laser-in-situ keratomeliusis (LASIK) utilises creation of a super ficial corneal flap of 90—120 micron thickness by mechanical keratome or femtosecond laser (fig. 3.8). Then the flap is raised and the cornea is reshaped by laser in the same way as in PRK. Afterwards the flap is repositioned, smoothed, and no suture is applied. After the surgery, visual recovery is very rapid, and there is only a minor, only 2—3-hour (3—5-day with PRK) period of surgery related irritation. With modern techniques of laser vision correction 95 % of patients receive 20/40 uncorrected visual acuity and will no longer need glasses for most of their activities.

Intraocular Lens Implantation

Phakic intraocular lens (IOL) implantation (without the natural crystalline lens extraction) is an alternative surgical procedure to PRK and LASIK for correcting moderate to severe myopia, in cases when the cornea is too thin or too flat and can produce sometimes better and more predictable outcomes than laser correction. Phakic IOLs are clear implantable lenses (like contact lenses) that are placed between the cornea and the iris (Verisyse) or between the iris and the lens (Visian ICL) during the surgery through a small incision in the peripheral cornea (2.0—3.0 mm), without removing the natural crystalline lens. In contrast to contact lenses, phakic IOLs work from within the eye, providing a permanent correction of myopia and do not require any maintenance.

Clear lens extraction (CLE), also called refractive lens exchange (RLE), is the name of surgical procedure when a noncataractous natural lens is removed through a small incision in the peripheral cornea (2.0—2.2 mm) with IOL implantation in the lens capsular bag as a refractive procedure. Such procedure is usually reserved for highly ammetropic patients that are not suitable for LASIK, PRK, or phacik IOL implantation. The main drawback of CLE is surgically indiced presbyopia that can be managed to some extent by implantation of multifocal IOL during surgery.

8. Amblyopia

Definition. Amblyopia is defined as unilateral or bilateral decrease of visual acuity for which no organic cause can be detected. It is the medical term used when the vision in one of the eyes is reduced because the eye and the brain are not working together properly. The eye itself looks normal, but it is not being used normally because the brain is favoring the other eye. This condition is also called *lazy eye*. Amblyopia is the most common cause of impaired vision in childhood. It is not correctable by glasses or contact lenses and is not due to any eye disease.

Etiology. Amblyopia develops in infancy or early childhood and usually can be detected in very young patients, principally by measuring or estimating visual acuity. Amblyopia results from a disruption of the normal development of vision, which distinguishes it from vision loss resulting directly from organic ocular defects, such as cataract, retinoblastoma (a life-threatening tumor of early childhood), and other inflammatory and congenital ocular disorders. It is usually unilateral, but it can (rarely) affect both eyes. Amblyopia does not cause learning disorders. Amblyopia may develop in young children who receive visual information from one eye that is blurred or conflicts with information from the other eye. To understand how amblyopia may develop in this way, consider that the brain is receiving two stimuli for each visual event: one from a visually aligned (fixating) eye and one from an 'abnormal' eye (vision blurred or eye misaligned on another target). The child's brain selects the better image and suppresses the blurred or conflicting image, which results in the faulty development of vision in the amblyopic eye. In other words, the brain continually 'favors' the eye with better vision, to the eventual detriment of visual development in the other eye.

A number of predisposing factors can lead to the development of amblyopia. These are summarized below.

Strabismic Amblyopia. A child can develop amblyopia in the context of strabismus (misaligned eyes). An adult onset of strabismus generally will cause diplopia (double vision) because the two eyes are not aligned on the same object. The brain of a child, on the other hand, is more adaptive. In a similar strabismic situation, the child's brain ignores (suppresses) the image from one of the eyes — usually the one that provides the more blurry image. Although such an adaptation overcomes the troublesome symptom of diplopia, this cortical suppression of sensory input from one eye may interrupt the normal development of vision in the higher centres of the brain; this interruption may result in reduced vision, which is amblyopia. Sometimes the degree of misalignment between the two eyes is very slight, making detection of strabismus and suspicion of strabismic amblyopia difficult. Even with a small angle of strabismus, amblyopia may be quite dense.

Refractive or Anisometropic Amblyopia. Amblyopia can result from a difference in refractive error between the two eyes — anisometropia. The eye with the lesser refractive error provides the clearer image and usually is favored over the other eye; suppression occurs and amblyopia develops. Children with asymmetric hyperopia are susceptible, because unequal accommodation is impossible; the child can bring only one eye at a time into focus. Refractive amblyopia may be as severe as that found in strabismic amblyopia. However, the pediatrician or family physician may overlook the possibility of amblyopia because there is no obvious strabismus. Detection of amblyopia must be based on an abnormality found in visual acuity testing.

Form-Deprivation and Occlusion Amblyopia. Form-deprivation amblyopia (amblyopia ex anopsia) can result when opacities of the ocular media, such as cataracts or corneal scarring, prevent adequate sensory input and, thus, disrupt visual development. The amblyopia can persist even when the cause of media opacity is removed. Rarely, occlusion amblyopia can result from patching of the normal eye.

Clinical Picture. The pediatrician or family physician will most likely be the first to see a young patient with amblyopia or strabismus and, therefore, will have the principal responsibility for screening. The child's physician must be familiar with the different kinds of amblyopia and strabismus, the close relationship of these two conditions, and how best to detect them. Vision is a developmental sensory function. Vision at birth is relatively poor, but through proper visual stimulation in the early months and years of life, a normal acuity is achieved at approximately 3 years of age. If this developmental process — the stimulation of the vision-receptive cells in the brain— is prevented because of strabismus, abnormal refractive error, congenital cataract, or some other condition, vision will not develop properly. This is a failure of the developmental process, not primarily an organic abnormality of the eye.

Complaints. Most of the time, there are no symptoms of amblyopia. Since only one eye is affected, the other eye usually has reasonably good vision and tends to take over all visual tasks. Unless the good eye is covered, the person will rarely notice the poor vision in the amblyopic eye. Other symptoms are decreased and blurred vision; strabismus; crossed or wandering eyes; poor depth perception; closing one eye while reading or watching television, tilting the head when looking at an object; headaches.

Signs. Decreased visual acuity and binocular vision, strabismus.

Examination. Pediatric vision screening is important for detecting not only amblyopia and strabismus but also congenital cataract, glaucoma, retinoblastoma, and other vision- or life-threatening conditions. Regular screening by the pediatrician or family physician helps ensures that the child's vision is developing normally or, if it is not, that early treatment is instituted. At a minimum, all children should undergo an evaluation to detect eye and vision abnormalities during the first few months of life and again at about 3 years.

Examination methods — visual acuity testing, refractometry, corneal light reflex test, cover test. Additional tests are pupillary reactions, direct ophthalmoscopy.

Amblyopia Testing. Amblyopia can be detected by testing visual acuity in each eye separately. Although there is no specific Mendelian pattern of inheritance, strabis-

mus and amblyopia cluster in families. Restoration of normal visual acuity can be successful only if treatment is instituted during the first decade of life, when the visual system is still in the formative stage. Techniques for measuring or estimating visual acuity (or visual function) and detecting amblyopia vary with the child's age, as described below.

NOTE!

The importance of visual acuity testing in detecting amblyopia cannot be overemphasized. Amblyopia may be present in the eyes without strabismus, so vision in each eye may not be normal even if the eyes appear normally aligned.

Newborns. True visual acuity cannot be measured in newborns. However, infants' general ocular status should be assessed through corneal light reflex testing, evaluation of the red reflex, pupillary testing, and, if possible, fundus examination.

Infants to 2-year-olds. With infants, it is possible only to assess visual function, not visual acuity. To test for amblyopia in infants (from a few months to about 2 years old), cover each eye in turn with a hand or, preferably, an adhesive patch and note how the child reacts. The infant should be able to maintain central fixation with each eye. If amblyopia is present, the child will likely protest — vocally or by evasive movements — the covering of the 'good' eye. Visual function, including ocular motility, may be further assessed by passing an interesting object, such as a ring of keys, before the baby and noting how the infant watches and follows the moving object. Moving the child's head can be used to demonstrate full ocular motility if not otherwise documented by following movements.

Age 2 to 4 or 5. A picture card may be used to test visual acuity in children between 2 and 3 years old. At age 3 (or before, if the child can follow directions and communicate adequately), visual acuity should be tested with the tumbling E chart. Use of an adhesive patch is the best way to ensure full monocular occlusion and accurate acuity measurement in children at these ages. Vision should be rechecked annually once visual acuity has been determined to be normal in each eye. Young children may not quite reach 20/20 acuity; this is no cause for concern as long as vision is at least 20/40 and both eyes are equal.

Age 4 or 5 and Up. The Snellen chart may be used to test visual acuity in children aged 4 or 5 and up who know the letters of the alphabet. A recent advance in early detection of amblyogenic factors is photoscreening. A computerized camera takes a photograph of the child's undilated eyes. Refractive errors, strabismus, anisometropia, and media opacities are visible in the photos. This technique permits screening of preverbal children and those unable to cooperate with other types of testing.

Treatment. In children younger than 5, strabismic amblyopia can usually be treated effectively by the ophthalmologist through *patching* of the unaffected eye. The child wears an adhesive patch over the good eye, forcing the brain to utilize the previously suppressed eye. In general, the success

NOTE!

Amblyopia must be detected early to be treated successfully. Left untreated, vision loss may range from mild to severe.

of occlusion treatment for amblyopia patients between the ages of 5 and 9 will depend on the age of the patient, the degree of amblyopia, and the persistence of patient compliance with treatment. Treatment is better tolerated by younger children but can be successful in children as old as 10. A treatment program started early in life often must be continued throughout the patient's first decade. Amblyopia treatment by patch occlusion of the unaffected eye must be monitored carefully, especially during the younger years, to avoid causing amblyopia through sensory deprivation of the occluded eye.

Treatment of refractive amblyopia consists first of wearing *glasses*, followed by patching of the better eye if the visual acuity difference persists after 4 to 8 weeks of wear. Equal vision in both eyes is readily achievable with parental cooperation. In general, the earlier the individual with amblyopia is diagnosed and treated, the better the chance of achieving equal vision. The most effective way to support fusion (binocular vision) is to treat amblyopia and equalize vision. Glasses can treat some or all of esotropia in a farsighted, or hyperopic, individual and may decrease the frequency of deviation in a myopic individual with exotropia.

However, *surgical correction* of the misalignment may still be necessary, particularly in those children who develop esotropia before the age of 6 months (congenital esotropia). Even when binocular vision may not be achievable, the impact of a disfiguring strabismus on a patient's self-image is a valid indication for surgery. It must be stressed that surgery is not an alternative to glasses and patching when amblyopia is present. Optometric 'vision training' is rarely indicated for the treatment of amblyopia or strabismus.

Prognosis. Good, when adequate diagnosis and treatment are provided. If detected and treated early, amblyopia can be cured. For best results, treatment should begin before age 5; treatment for amblyopia is rarely successful if initiated past age 10. At least half of all patients with amblyopia also have strabismus, a misalignment of the two eyes.

Complications. The early detection of amblyopia is an important responsibility for those involved in infant and child health care. Delayed diagnosis may have serious consequences for visual acuity, eye disease, or systemic disease. If an abnormality is suspected, the patient should be referred promptly to an ophthalmologist.

Prophylaxis. Early detection and treatment of underlying cause.

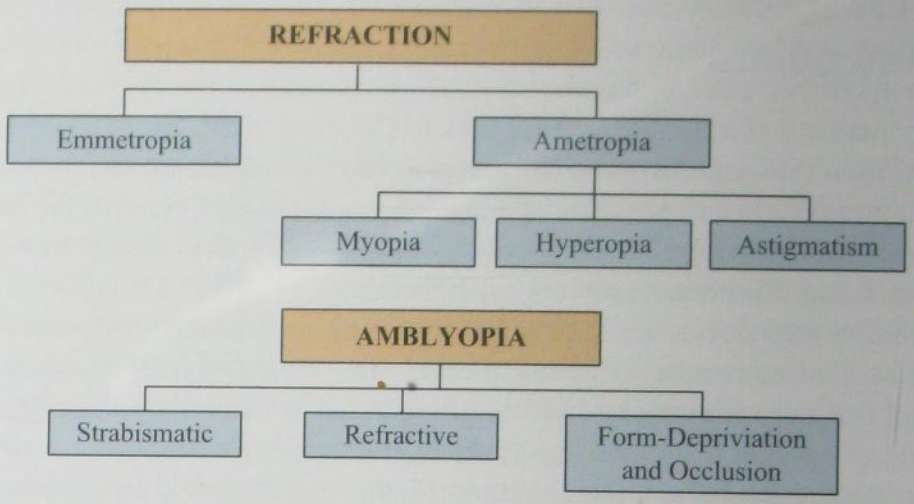
Review:

1. Key points

The human eye is a complex device that acts like a camera. The eye is able to focus incoming light rays and form a clear image on the retina, giving the man the ability to see. Light rays have to pass through the following parts of the eye: the cornea, the aqueous humor, the crystalline lens, and the vitreous body, deviating at a certain angle on each part and this phenomenon is called refraction.

Total combined deviations (refractions) of light rays lead to convergence on the retina (so-called emmetropia) giving a clear view of the object we are looking at. When light rays are converging in front (myopia) of or behind (hyperopia) the retina, or at different points (astigmatism) (both in front, or both behind, or some in front and other behind the retina), the eye has refractive errors called ametropia. In such a case images on the retina are blurred. Ametropia results in degradation of vision and blurring of distant or nearby objects. In order to receive a clear image different types of optical corrections (glasses or contact lenses) or refractive surgery (LASIK, PRK, phakic IOL, CLE) are employed. The ability of the eye to focus on objects located at different distances is called accomodation. As the crystal line lens hardens with age, accomodation gradually reduces the eye's ability to focus on close objects. Normally it happens at the age of 40—45 years and an emmetrope is facing gradual blurring of near vision — a phenomenon called presbyopia. Amblyopia is a form of treatable visual loss found in the young and adult population. It can be defined as a loss of visual acuity not correctable by glasses in an otherwise healthy eye. Amblyopia develops in infancy or early childhood and usually can be detected in very young patients, principally by measuring or estimating visual acuity. Amblyopia results from a disruption of the normal development of vision, which distinguishes it from vision loss resulting directly from organic ocular defects, such as cataract, retinoblastoma (a life-threatening tumor of early childhood), and other inflammatory and congenital ocular disorders. It is usually unilateral, but can (rarely) affect both eyes. Amblyopia does not cause learning disorders, and if detected and treated early, can be cured. For best results, treatment should begin before age 5; treatment for amblyopia is rarely successful if initiated past age 10. At least half of all patients with amblyopia also have strabismus, a misalignment of two eyes.

2. Diagrams



3. The Review Questions

A. Control Questions

1. What is refraction and its types?
2. What is myopia?
3. What are the main reasons for myopic refraction?
4. What is hyperopia?
5. How are light rays converged in an eye of a hyperopic patient?
6. What phenomen makes it possible to see distant and near objects clearly?
7. How does accommodation work?
8. What is presbyopia?
9. What is amblyopia?
10. What types of amblyopia do you know?

B. Tests

1. **If light rays converge in front of the retina, this condition is called:**
   A. Emmetropia
   B. Hyperopia
   C. Myopia
   D. Astigmatism
   E. Amblyopia

2. **If light rays converge behind the retina, this condition is called:**
   A. Emmetropia
   B. Hyperopia
   C. Myopia
   D. Astigmatism
   E. Amblyopia

3. **If light rays converge on different points in front, behind or on different sides of the retina, this condition is called:**
   A. Emmetropia
   B. Hyperopia
   C. Myopia
   D. Astigmatism
   E. Amblyopia

4. **The main anatomical causes of myopia are:**
   A. Long eye and/or steep cornea
   B. Long eye and/or flat cornea
   C. Short eye and/or steep cornea
   D. Short eye and/or flat cornea
   E. Long eye and/or irregular cornea

5. **The main anatomical causes of hyperopia are:**
   A. Long eye and/or steep cornea
   B. Long eyeand/or flat cornea
   C. Short eye and/or steep cornea
   D. Short eye and/or flat cornea
   E. Long eye and/or irregular cornea

6. **Presbyopia is a condition induced by:**
   A. Aging
   B. Cataract
   C. Trauma
   D. Keratitis
   E. Glaucoma

7. **Amblyopia results from:**
   A. Vision loss due to cataract
   B. Disruption of the normal development of vision
   C. Vision loss due to retinoblastoma
   D. Vision loss due to congenital macular disorder
   E. Headache

8. **Accomodation is a process of:**
   A. Adjustment of the retinal image to the pupil size
   B. Adjustment of the eyeball length during focusing
   C. Adjustment of the retinal photoreceptors during focusing
   D. Adjustment of focal length of the eye
   E. Adjustment of the pupil diameter

9. **Contact lenses:**
   A. Are used only in high-degree astigmatism
   B. Cannot be used in hyperopia
   C. Can have higher risk of inflammation of the cornea than glasses
   D. Can be fitted without any special examination
   E. Are used prior to refractive surgery

10. The goal of refractive surgery is:

A. To change refraction to the opposite type
B. To receive 20/20 vision
C. To increase spectacle independence
D. To increase the refractive power of the crystalline lens
E. To increase the refractive power of the cornea

C. Clinical Cases

Case 1

A 14-year-old boy is seen for a physical examination at school. He admits a difficulty reading from the blackboard but not reading textbooks. He does not wear glasses. You record VA as OD 20/100, pinhole 20/25; and OS 20/100, pinhole 20/25. What is your diagnosis? Would you manage or refer this patient?

Case 2

A 78-year-old woman is seen for an annual physical examination and complains of mild difficulty reading and seeing street signs. You record OD 20/70, no improvement with pinhole; and OS 20/50, no improvement with pinhole. Upon direct ophthalmoscopy, you note a dullness of the red reflex and you have difficulty seeing fundus details in both eyes. What is your diagnosis? Would you manage or refer this patient?

Case 3

A 40-year-old man is seen for an annual executive physical. He has no complaints and does not wear glasses. You record VA as OD 20/15; and OS 20/100, no improvement with pinhole. During examination, the patient revealed that he has been aware since childhood that his left eye was a so-called lazy eye — in other words, that he suffered from amblyopia. Would you refer this patient?

Case 4

A 50-year-old man visits your office because he noted decreased visual acuity in the right eye the preceding day while accidentally occluding his left eye. When his present glasses were prescribed 2 years ago, his vision was equal in both eyes. You record VA as OD 20/50, no improvement with pinhole; and OS 20/20. Upon ophthalmoscopy, no abnormalities are detected. What, if any, is your diagnosis? Would you manage or refer this patient?

Case 5

A 3-year-old boy is brought to you by his mother, who tells you that she suspects his right eye is not straight. What steps would you determine if a significant problem is present?

Case 6

A family has just moved into your area and the mother brings her 6-month-old baby to your family practice office for a routine checkup. She mentions that the child's grandfather has noted that in several photographs the baby's left eye appears crossed. He is adamant in his observation and feels that "something should be done". The mother has felt that, at times, the eye has appeared crossed, but the baby's father has not observed this phenomenon. How should you proceed?

Case 7

A 2-year-old boy is brought to your office because his mother has noticed that over the past 2 weeks his right eye has deviated inward during periods of fatigue. On the previous evening, the boy's father claimed to have noted a white reflex in the child's right eye. How should you proceed?

Case 8

A 54-year-old man has early cataracts in both eyes. With glasses, the right eye cannot be corrected to better than 20/200, whereas with the left eye he can read the 20/40 line with best correction. The amount of cataract is exactly the same in each eye. Examination of the optic disc and macula, pupillary reaction, color vision, and retinal blood vessels proved entirely normal in each eye. However, the right eye appears to be turned slightly inward when you evaluate the corneal light reflex, and the patient has not experienced diplopia. Additional questioning reveals that the patient wore a patch over one eye as a child. Why would information concerning his childhood ocular condition be relevant in this situation?

Case 9

A 4-year-old boy with attention-deficit disorder comes to your office for his routine preschool physical examination. Your nurse tests his visual acuity with apicture card and obtains 20/30 vision on the right. During testing of the left eye the patient loses attention and refuses to cooperate further with testing. What course of action should you take?

Case 10

A mother reports that her 1-year-old child is sensitive to light and his right eye looks larger than the left one. On examination you note that although the child's right eye does look larger, the pupillary reactions are equal in both eyes, the corneas are clear, and there is a good red reflex in each eye. What should you tell the mother?

a. Do not worry, the child will "grow into" his eyes.
b. Return in 1 month for a reexamination.
c. Take the child to an ophthalmologist on immediate referral.
d. This is probably a cancer of the right eye, and you should take the child to an oncologist.

WHO IS WHO IN EYE CARE

Ophthalmologist

An ophthalmologist is a physician (doctor of medicine) who specializes in the medical and surgical care of the eyes and visual system and in the prevention of eye disease and injury. The ophthalmologist has completed four or more years of college premedical education, four or more years of medical school, one year of internship, and three or more years of specialized medical, surgical, and refractive training and experience in eye care. The ophthalmologist is a specialist who is qualified by lengthy medical education, training, and experience to diagnose, treat, and manage all eye and visual system problems and is licensed by a state regulatory board to practice medicine and surgery. The ophthalmologist is the medically trained specialist who can deliver total eye care: primary, secondary, and tertiary care services (i.e., vision services, spectacles, contact lenses, eye examinations, medical eye care, and surgical eye care), diagnose general diseases of the body, and treat ocular manifestations of systemic diseases.

Optometrist

An optometrist is a health service provider who is involved exclusively with vision problems. Optometrists are specifically educated and trained by an accredited optometry college in a four-year course, but they do not attend medical school. They are state-licensed to examine the eyes and to determine the presence of vision problems. Optometrists determine visual acuity and prescribe spectacles, contact lenses, and eye exercises. Optometrists may perform all the services listed under the definition of opticians. Most states have passed legislation that permits optometrists to treat some eye conditions.

Optician

An optician is a professional who makes, verifies, delivers, and fits lenses, frames, and other specially fabricated optical devices and/or contact lenses upon prescription to the intended wearer. The optician's functions include prescription analysis and interpretation; determination of the lens forms best suited to the wearer's needs; preparation and delivery of work orders for lens grinding and eye wear fabrication; verification of finished ophthalmic products; and adjustment, replacement, repair, and reproduction of previously prepared ophthalmic lenses, frames, and other specially fabricated ophthalmic devices.

CHAPTER 4

Diseases of the Eyelids

OBJECTIVES

To know:

— classification of eyelid diseases;
— basic diagnostic methods;
— the principles of emergency care of eyelid diseases;
— treatment methods of inflammatory diseases of the eyelids;
— the types of developmental anomalies of the eyelids;
— specifics of the clinical course of eyelid disease complications.

Plan:

1. CLASSIFICATION OF EYELID DISEASES
2. SYMPTOMS OF EYELID DISORDERS
3. EXAMINATION METHODS
4. EYELID DISEASES
   - **4.1. Developmental anomalies**
     - Coloboma
     - Epicanthus
     - Blepharophimosis
     - Ankyloblepharon
     - Distichiasis
   - **4.2. Eyelid Malposition**
     - Entropion
     - Ectropion
     - Ptosis
     - Lagophthalmos
     - Trichiasis
   - **4.3. Inflammatory Diseases**
     - Blepharitis
     - Chalazion
     - Hordeolum (stye)
     - Eyelid Edema
     - Eyelid Abscess
     - Eyelid Cellulitis (phlegmon)
   - **4.4. Tumors**
     - Papilloma
     - Xanthelasma
     - Molluscum contagiosum
     - Cutaneous Horn
     - Basal Cell Carcinoma
     - Eyelid Adenocarcinoma
     - Melanoma

1. Classification of Eyelid Diseases

— By origin — congenital or acquired.
— By the type of the inflammatory process — acute or chronic.
— By pathogenesis — inflammatory diseases, eyelid malposition, tumors.

1.1. Developmental anomalies — coloboma, epicanthus, ankyloblepharon, blepharophimosis, distichiasis, etc.
1.2. Eyelid malposition
1.2.1. Due to myogenic and neurological factors — entropion, ectropion, ptosis, lagophthalmos.
1.2.2. Structural abnormalities — trichiasis.
1.3. Inflammatory diseases
1.3.1. Inflammation of the eyelid margin — blepharitis.
1.3.2. Disorders of the eyelid glands — chalazion, hordeolum (stye).
1.3.3. Inflammation of the eyelid skin — edema, abscess, cellulitis (phlegmon).
1.4. Tumors
1.4.1. Benign — papilloma, hemangioma, nevus, seborrheic keratosis, hydrocystoma, xanthelasma, molluscum contagiosum, cutaneous horn.
1.4.2. Malignant — basal cell carcinoma, squamous cell carcinoma, melanoma, adenocarcinoma.

2. Symptoms of Eyelid Disorders

1. Swelling.
2. Redness.
3. Foreign body sensation.
4. Burning, itching.
5. Pain.

3. Examination Methods

— External examination or inspection of the eyelids and lid margins.
— Asesssment of the eyelid position and the width of the palpebral fissure.
— Palpation.
— Slit-lamp examination (biomicroscopy).
— Upper and lower lid eversion.

4. Eyelid Diseases

4.1. Developmental Anomalies

Coloboma

Definition. A coloboma is a full-thickness defect involving absence of a portion of the eyelid tissues. The defect may be unilateral or bilateral, and may vary from a small notch of the lid margin to a complete absence of the entire lid. Although an eyelid coloboma can occur in many locations, the most common position is the nazal side of the upper eyelid.

Etiology. Incomplete closure of the embryonic facial cleft and pressure of the amniotic bands are implicated in the etiology of coloboma. It is caused by genetic (inherited) or/and environmental factors (fetal alcohol syndrome, vitamin A deficiency, teratogenic drugs, infections — toxoplasmosis, cytomegalovirus). It may be isolated or associated with other ocular or systemic anomalies. Coloboma may also occur as a result of trauma.

Clinical Picture. Depending on the extent of coloboma, desiccation symptoms on the conjunctiva and cornea with incipient ulceration may arise from the lack of regular and uniform moistening of the conjunctiva and cornea.

Symptoms. Aesthetic problems with the absence of a part of the eyelid, photophobia, tearing.

Signs. Absence of the upper eyelid part, cornea exposure.

Methods of Examination. External examination, assessment of the eyelid position.

Treatment. Corneal protection is the primary goal in the medical treatment of eyelid colobomas (artificial tears and ointment) followed by surgical reconstruction of all layers of the eyelid by means of different surgical methods, which depend on the size of the defect.

Prognosis is excellent to good, depending on the size of the lesion and the rate of therapy.

Complications of upper eyelid coloboma depend on the size of the defect, presence of eye anomalies, and reconstruction methods. The lid defect leaves part of the cornea uncovered, which can lead to its excessive dryness, keratitis, corneal leukoma and amblyopia.

Prophylaxis. No specific steps are identified for coloboma prevention. Parents with genes related to this genetic anomaly can be given genetic counseling and warning before child birth. Another way to prevent any of developmental anomalies is healthy maternal nutritional status and environmental factors.

Epicanthus

Definition. Epicanthus is the fold of the skin of the upper eyelid that covers the inner canthus of the eye. It is normal in persons of certain races (Asians), but anomalous in others. Epicanthal folds may also be seen in young children of any race before the bridge of the nose begins to rise.

Etiology. Epicanthal folds may be normal for people of Asian descent and some non-Asian infants. However, it may also be due to certain medical conditions, including: Down syndrome, fetal alcohol syndrome, Turner syndrome, phenylketonuria, Rubinstein—Taybi syndrome, blepharophimosis.

Clinical Picture. A vertical fold of skin over the angle of the inner canthus of the eye. It may be slight or marked, covering the canthus and the caruncle. The condition is usually bilateral. Thirty percent of newborns have epicanthal fold until the age of six months. The nasal bridge becomes more pronounced as the child grows, and most epicanthal folds disappear by the age of four.

Symptoms. A skin fold that covers the inner corner of the eye. It runs from the nose to the inner side of the eyebrow.

Signs. A crescentic skin fold extending bilaterally between the upper and lower eyelids and covering the medial angle of the eye.

Methods of Examination. External examination, asessment of the width of the palpebral fissure.

Differential Diagnosis. Epiblepharon, blepharophimosis, entropion.

Treatment. It can be corrected by plastic surgery.

Prognosis is excellent to good.

Complications. Epicanthus is usually only of cosmetic importance, but in some cases it may obscure the medial visual field and can cause amblyopia or esotropia.

Prophylaxis. In most cases, epicanthus cannot be prevented. A way to prevent any of developmental anomalies is healthy maternal nutritional status and environmental factors.

Blepharophimosis

Definition. Blepharophimosis is a decrease in the size of the palpebral fissure without eyelid margin fusion. The palpebral fissure, normally 28—30 mm wide,

may be reduced to half that width. The nasal bridge is flat and there is a hypoplastic orbital rim.

Etiology. This condition is usually genetically inherited by an autosomal dominant pattern, in which one set of the modified gene in each cell is responsible for causing the disorder. In some cases, the patient receives the mutation from one affected parent. Other cases result from new mutations in the gene and take place in people with no significant family history of the disorder. It can also be acquired due to scar contracture or aging.

Clinical Picture. Abnormal narrowness of the palpebral fissure horizontally and vertically, the nasal bridge is flat and there is a hypoplastic orbital rim. Aside from small palpebral fissures, the clinical picture includes epicanthus inversus (folds curving in the mediolateral direction, inferior to the inner canthus) and bilateral ptosis of the eyelids.

Symptoms. Both the vertical and horizontal sizes of the palpebral fissures are reduced in length, the eyelids shorten from side to side, ptosis, epicanthus, the nose and orbital rim are flattened, other facial abnormalities may occur.

Signs. Decreased size of the palpebral fissure, ptosis, epicanthus, entropion, flat nose, hypoplastic orbital rim.

Methods of Examination. External examination, asessment of the width of the palpebral fissure.

Differential Diagnosis. Epicanthus, ankyloblepharon, ptosis.

Treatment. Surgical enlargement of the palpebral fissure by canthotomy or plastic surgery is required.

Prognosis is good, more than one surgery may be required.

Complications. Patients with blepharophimosis have a high rate of amblyopia, strabismus or facial paralysis.

Prophylaxis. The disease has no effective preventive methods. Parents with genes related to this genetic anomaly can be given genetic counseling and warning before child birth. Another way to prevent any of developmental anomalies is healthy maternal nutritional status and environmental factors. Early treatment is the key to complication prevention.

Ankyloblepharon

Definition. Ankyloblepharon is partial or complete adhesion between the upper and lower eyelid margins (stitched eye). Ankyloblepharon is frequently associated with other skull deformities.

Etiology. Congenital failure of the eyelids to separate during embryonic development. It also can be acquired due to chemical burn.

Clinical Picture. Partial fusion of the upper and lower lids with single or multiple bands of fine connective tissue that prevent opening of the eye.

Symptoms. Inability to open the eye due to fusion of the upper and lower eyelids with single or multiple bands.

Signs. The margins of the upper and lower eyelids are fused, full eyelid opening is impaired.

Methods of Examination. External examination, asessment of the width of the palpebral fissure.

Differential Diagnosis. Microphthalmos, cryptophthalmos, symblepharon.

Treatment consists of simple surgical resection of the fibrous bands.

Prognosis is good.

Complications. Treatment should be performed to minimize the risk of amblyopia.

Prophylaxis. In most cases, ankyloblepharon cannot be prevented. A way to prevent any of developmental anomalies is healthy maternal nutritional status and environmental factors.

Distichiasis

Definition. Distichiasis is the presence of a double row of eyelashes in the lid margin that may be turned inward towards the eye. It can affect either upper or lower eyelid. Additional lashes are usually bilateral.

Etiology. It is hereditary, but the mode of inheritance has not been determined. The congenital form of distichiasis is autosomal dominant with complete penetrance and metaplasia of the meibomian glands.

Clinical Picture. When abnormal lashes come in contact with the cornea it causes severe irritation, epiphora, corneal abrasion, or even corneal ulcers.

Symptoms. It can cause irritation, tearing, foreign body sensation, and blepharospasm.

Signs. An extra row of eyelashes that grow out of the meibomian gland opening at the lid margin, tearing, conjunctival injection.

Methods of Examination. Biomicroscopy (slit-lamp examination). Examination shows eyelashes turned to the eyeball. After compression the degree of entropion increases.

Differential Diagnosis. Trichiasis, entropion.

Treatment. To relieve symptoms and protect the cornea lubricants can be used. Definitive treatment is removal of the abnormal eyelashes, usualy by cryotherapy, direct surgical excision or electrolysis.

Prognosis is mostly excellent.

Complications. Untreated distichiasis can cause corneal ulcer and corneal vascularization.

Prophylaxis. In most cases, distichiasis cannot be prevented.

EYE FACTS

Elizabeth Taylor, the famous actress, was considered the world's most beautiful woman in her heyday. She was born with an extra row of eyelashes caused by a rare genetic mutation that highlighted her eyes. Although she avoided any of the complications associated with distichiasis.

4.2. Eyelid Malposition

Entropion

Definition. Entropion is a condition in which the lid margin is turned inwards (toward the eyeball) (fig. 4.1).

Etiology. There are four etiological forms of entropion: congenital, senile, spastic, and cicatricial.

- Congenital entropion is associated with microphthalmos.
- Involutional (senile) entropion is caused by horizontal laxity of the eyelid, muscle atony, instability of the tarsal plate.
- Spastic entropion results from orbicularis oculi spasm, particularly when the eyeball is deeply set, small (microphthalmos) or absent.
- Cicatricial entropion is caused by contraction or shortening of the eyelid conjunctiva due to injuries, burns, surgeries, inflammatory processes (e.g., trachoma).

Clinical Picture. The eyelids (mostly lower) are turned toward the eye, which leads to the eyelashes being rubbed against the cornea.

Symptoms. Patients complain of discomfort, pain, irritation, excessive tearing, foreign body sensation, and photophobia.

Signs. Injection of the eyeball, lacrimation, blepharospasm, the eyelid margin is turned towards the eyeball.

Methods of Examination. Slit-lamp examination.

Differential Diagnosis. Trichiasis, distichiasis, trachoma.

Treatment. Ocular lubrication and tear preparations help protect the ocular surface and also may break the cycle in patients with spastic entropion due to dry eye syndrome. Eyelid hygiene, antibiotics, and corticosteroids are useful for the treatment of blepharitis, which may cause spastic entropion. For surgical management of entropion multiple surgical procedures can be used according to the class of entropion being treated.

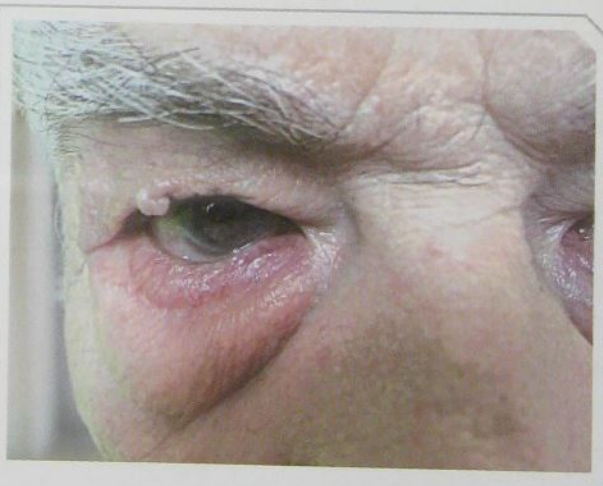


Fig. 4.1. Entropion

Prognosis. Congenital entropion is usually asymptomatic and often resolves wihtin the first month of life. The prognosis of spastic entropion and cicatricial entropion is favorable with prompt surgical intervention but before any corneal changes occur.

Complications. If entropion is left untreated, this may lead to cornea damage or formation of a corneal ulcer that casues vision reduction.

Prophylaxis. Adequate medical and surgical treatment of ocular disorders.

Ectropion

Definition. Ectropion is abnormal eversion of the eyelid margin (usually lower) away from the eyeball with exposure of the palpebral and bulbar conjunctiva.

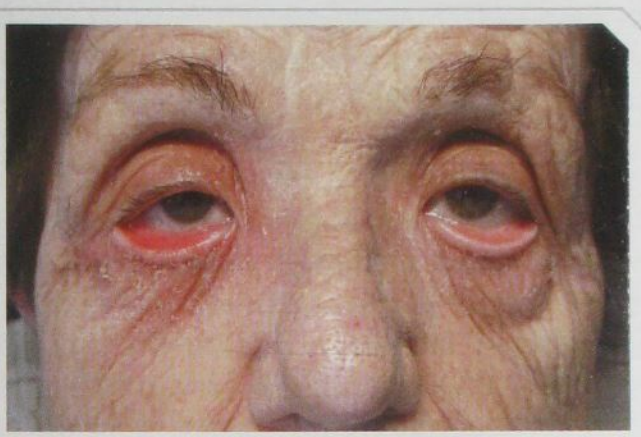


Fig. 4.2. Ectropion

Etiology. Ectropion occurs in five forms: spastic, cicatricial, senile, paralytic, and mechanical.

— Cicatricial ectropion results from destruction of the lid skin by injury, burns, ulcers, chronic conjunctivitis or blepharitis.

— Senile (involutional) ectropion occurs in old people due to laxity of the tissue of the lower lid or loss of tone of the orbicularis oculi (fig. 4.2).

— Paralytic ectropion occurs when there is the damage of the facial nerve.

— Mechanical ectropion is caused by the weight of a tumor or granuloma of the lower eyelid.

Clinical Picture. The lower eyelid is eversed out of the eyeball that becomes very dry and sore. The eyes become red, watery, and tears would roll down the drooping lower eyelid onto the cheek. As a result, the cornea can also be damaged since the patients cannot close their eyes properly. The cornea is left exposed and sooner or later, corneal ulcer can develop.

Symptoms. Patients complain of tearing, dryness, and redness of the eye. They may constantly wipe their eyes, thereby exacerbating lid laxity and ectropion.

Signs. Examination shows that the lower eyelid is not ajacent to the eyeball, so the lacrimal point is not immersed into the lacrimal lake, chronic conjunctivitis may be present as well as keratitis.

Methods of Examination. Slit-lamp examination.

Differential Diagnosis. Eyelid malignancy.

Treatment. Artificial tears and eye lubricant ointments can be used to keep the eye moist and soothe irritation prior to ectropion repair. The methods of surgical treatment depend on the etiology; the main purpose of them is to tighten the muscles that hold the eyelid in place.

Prognosis is favorable when the disorder is treated promptly.

Complications. Because the eyelid does not cover the surface of the eyeball chronic blepharitis and conjunctivitis are gradually developing, keratitis and corneal ulcers may occur.

Prophylaxis. Patients with tearing and incipient ectropion or early punctal ectropion should be instructed to wipe the eyelids in the upward and inward direction (toward

the nose) to avoid worsening of medial ectropion. Adequate medical and surgical treatment of ocular disorders.

Ptosis

Definition. Ptosis is drooping of the upper eyelid (fig. 4.3). It can be unilateral or bilateral, partial or complete, acquired or congenital.

Etiology. Ptosis may be classified according to the underlying etiology:

— Myogenic ptosis is associated with improper development of the levator palpebrae superioris (LPS) or muscular weakness.

— Neurogenic ptosis is caused by aplasia of the nucleus of the oculomotor nerve or violation of sympathetic innervation to the eye muscles.

— Aponeurotic ptosis is the most common form of acquired ptosis caused by stretching and disinsertion of the levator aponeurosis owing to frequent rubbing of the eyes, wearing rigid gas permeable contact lenses, and traction during ocular surgery.

— Mechanical ptosis is caused by excessive weight of the upper lid due to multiple chalazia, neurofibromatosis, excessive cicatrization of the tarsal plate in trachoma, and benign and malignant tumors of the upper lid.

— Traumatic ptosis is caused by trauma to the LPS muscle or its aponeurosis.

Clinical Picture. There are three forms of ptosis according to the degree of its severity: partial ptosis (the eyelid is lowered compared to the norm by 1—3 mm), incomplete (the upper eyelid reaches the middle of the pupil), and complete ptosis (the upper eyelid completely covers the eyeball and the lower eyelid hangs over). Examination of patients with ptosis finds the edge of the upper eyelid covering the cornea by more than 2 mm. Patients raise the upper eyelid straining the muscles of the forehead or throwing their head (astrologer's pose). The superior palpebral groove is weak or absent.

Symptom of ptosis is drooping of the upper eyelid/eyelids. Except for this patients complain of difficulty seeing. Sometimes they tilt their heads back in order to see better or raise their eyebrows repeatedly trying to lift the eyelids.

Signs. Drooping eyelids are the most obvious sign of ptosis.

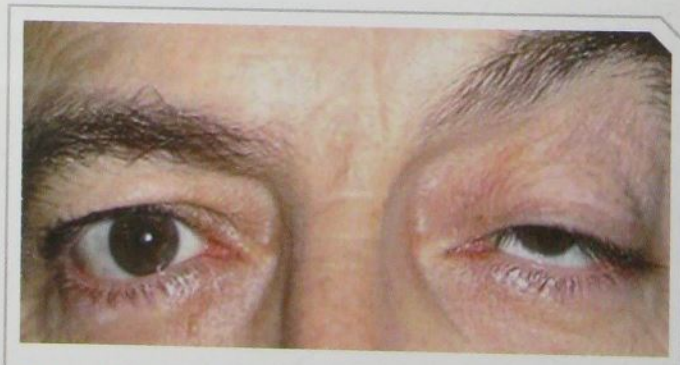


Fig. 4.3. Ptosis

Methods of Examination. Assessment of the eyelid position and the width of the palpebral fissure, CT scan or MRI may be required in cases of diagnostic difficulty.

Differential Diagnosis. Lagophthalmos, exophthalmos, pseudoptosis, malignancy.

Treatment. First, underlying disease should be treated. Acquired neurogenic ptosis

requires early conserveative treatment. In children, to prevent the development of amblyopia, doctor should carry out timely surgical treatment. Surgical options include frontalis slings, levator resection, and correction of any abnormality of the levator muscle. Sometimes lubricants may be required to prevent or treat exposure keratitis.

Prognosis is mostly positive if surgery is timely performed by experienced surgeons. However, ovecorrection is possible in incomplete closure of the eyelids.

Complications. In preschool children ptosis leads to a sustainable reduction due to the development of amblyopia or strabismus. In adults, reduced field of vision and headaches due to tension of the frontalis muscle can occur. Anxiety about appearance may result in social isolation and cause significant psychological complications.

Prophylaxis. In most cases, ptosis cannot be prevented.

Lagophthalmos

Definition. Lagophthalmos is a condition in which patients are unable to close their eyes completely. In other words, it is incomplete closure of the eyelids.

Etiology. The facial nerve (the seventh cranial nerve) innervates both the frontalis muscle, which raises the eyebrow, and the orbicularis oculi muscle, which closes the eyelids. Loss of function of the facial nerve inhibits eyelid closure as well as the blink reflex and the lacrimal pump mechanism. Facial nerve weakness may result from a host of causes:

- Trauma. Fractures to the skull base (the petrous portion of the temporal bone) or mandible can damage the nerve or one of its branches.
- Cerebrovascular accidents. The facial nerve receives its blood supply from the anteroinferior cerebellar artery.
- Bell's palsy. This is an idiopathic facial nerve palsy that is thought to be associated with an acute viral infection or reactivation of herpes simplex virus.
- Tumors. Acoustic neuromas in the cerebellopontine angle and metastatic lesions are most commonly associated with lagophthalmos.
- Infectious, immune-mediated causes. Less common causes of lagophthalmos include Lyme disease, chickenpox, mumps, polio, Guillain—Barre syndrome, leprosy, diphtheria, and botulism.
- Möbius' syndrome. This rare congenital condition is characterized by cranial nerve palsies (especially sixth and seventh cranial nerve palsies), motility disturbances, limb anomalies, and orofacial defects.

Clinical Picture. The inability to blink and effectively close the eyes leads to corneal exposure and excessive evaporation of the tear film that cause corneal inflammation and ulcers. In case of facial nerve damage on the affected sidethe nasolabial folds are smoothed, the corner of the mouth droops, wrinkling of the forehead is impossible.

Symptoms. Patients complain of discomfort, tearing, dryness and redness of the eye.

Signs. Incomplete closure of the eye, epiphora.

Methods of Examination. External examination of the eyelids, asessment of the eyelid position and the width of the palpebral fissure, slit-lamp examination.

Differential Diagnosis. Ptosis, exophthalmos.

Treatment of lagopthalmos can include both supportive care and surgical methods. If unable to receive surgery, artificial tears should be administered at least four times a day to the cornea to preserve the tear film. Leading up to surgery, a patient can undergo tarsorrhaphy which partially sews the eye shut temporarily to further protect the cornea as the patient is waiting for care. In case of facial nerve damage treatment is carried out in cooperation with a neurologist. In case of scarring and symblepharon of the eyelids one should resort to plastic surgery of the conjunctiva and eyelids. To eliminate the cause of lagophthalmos, the eyelid is temporarily fixated with plaster; one performs lateral and medial eyelid stitching or blepharorrhaphy. Symptomatic treatment is aimed at moisturizing of the cornea (with artificial tears and eye gels) and prevention of infections.

Prognosis. Considerable improvement is possible.

Complications. Left untreated, lagophthalmos can lead to permanent loss of vision due to corneal damage.

Prophylaxis. In general, lagophthalmos cannot be prevented, unless it is a result of eyelid retraction after blepharoplasty surgery through the skin or other surgical scars.

Trichiasis

Definition. Trichiasis is an abnormality in which the eyelashes are directed towards the eyeball.

Etiology. Trichiasis can be either congenital or acquired as a result of aging changes, inflammatory conditions or traumas.

Clinical Picture. Misdirected eyelashes rub on the cornea resulting in ocular pain, tearing, and redness.

Symptoms. The patient complains of discomfort in the eye, itching, irritation, foreign body sensation, and tearing.

Signs. The eyelashes are misdirected toward the eyeball, tearing, conjunctival injection.

Methods of Examination. Slit-lamp examination.

Differential Diagnosis. Chronic blepharitis, distichiasis.

Treatment. Lubricants, such as artificial tears and ointments, may decrease the irritant effect of lash rubbing. Several types of surgical methods can be used for trichiasis treatment: epilation, cryosurgery, electrolysis, radiofrequency ablation.

Prognosis is generally good.

Complications. Recurrence of eyelashe growth, corneal erosion or ulcer.

Prophylaxis. Timely and adequate treatment of inflammatory and traumatic ocular disorders.

4.3. Inflammatory Diseases

Blepharitis

Definition. Blepharitis is inflammation of the eyelid margins.

Etiology. Blepharitis occurs with chronic bacterial lid infection, meibomian gland dysfunction, seborrhea, and acne rosacea that affect the eye. The common causes of blepharitis are uncorrected or miscorrected refraction anomalies; diseases of the gastrointestinal tract (gastritis, colitis, intestinal parasites, etc.), inflammation of the sinuses, endocrine system disorders (diabetes), immunodeficiency; chronic dermatologic conditions (seborrheic dermatitis, herpes zoster dermatitis, staphylococcal dermatitis, rosacea); bacterial (Staphylococcus aureus, S. epidermidis, Streptococcus pneumonia, etc.), viral (Herpes simplex et zoster), fungal (Pityrosporum ovale et orbicularis), and parasitic (the ticks Demodex folliculorum humanis et brevis and the louse Phthirus pubis) infections; malfunctioning of the meibomian glands. Chronic blepharitis can be associated with exposure to chemical fumes, smoke, smog, conditioned air, and other irritants.

Blepharitis is classified into two forms: anterior and posterior. Anterior blepharitis affects the eyelid margins near the eyelashes. This can be caused by bacterial (or sometimes viral) infection. Posterior blepharitis is a condition that results from meibomian gland dysfunction (meibomitis). It has immunological origin.

Clinical Picture. Clinically blepharitis falls into the following groups:

— Simple — a mild form of blepharitis that is characterized by redness and thickening of the lid margin due to inflammatory changes.

— Scaly or squamous blepharitis — a mild form of blepharitis that causes reddening of the lid margins with white scales among the lashes. It is essentially an outpost of dandruff from the scalp in seborrheic subjects.

— Ulcerative — more severe blepharitis that involves a staphylococcal infection on top of the seborrheic inflammation and the lid margins become significantly indurated and even ulcerated. It is characterized by yellow crusts and scales on the inflamed thickened lid margins.

Symptoms. Patients complain of itching, irritation, burning; foreign body sensation in the eye; sensitivity to light (photophobia) or smoky atmosphere; blurred and decreased vision; excessive tearing or dry eyes (blepharitis alters the lipid layer of the tear film and accelerates its evaporation) and crusting around the eyes upon awakening. For some people, blepharitis causes only minor irritation and itching.

Signs of blepharitis may vary depending on its form.

Simple blepharitis is characterized by edema and hyperemia of the eyelid margins.

Scaly (squamosus) blepharitis — except hyperemia and edema of the eyelid margins there appear dandruff-like scales around the eyelashes especially upon awakening; scanty, broken eyelashes (fig. 4.4).

Ulcerative blepharitis — the eyelid margins are erythematous and thickened with yellow purulent crusts and debris within the lashes. The crusts are difficult to remove,

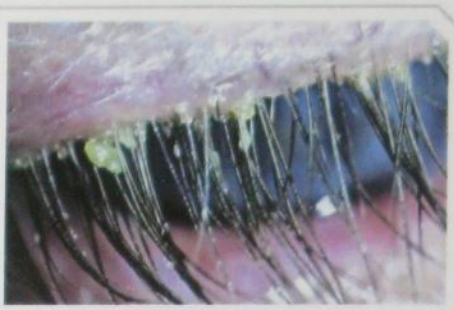


Fig. 4.4. Squamosus blepharitis

and then bleeding ulcers remain. Conjunctival injection or mild mucus discharge may be present.

Methods of Examination. Slit-lamp examination.

Differential Diagnosis. Chronic conjunctivitis, dry eye syndrome, atopic dermatitis, tumors of the eyelid margins.

Treatment. The management of blepharitis is systematic, prolonged, and depends on the cause of the disorder. It is very important to find out and eliminate this cause. It can be correction of refraction anomaly, management of gastrointestinal disorders or other systemic diseases, diet or fish and flaxseed oil consumption. When scalp dandruff is present, a dandruff shampoo for the hair is recommended as well.

The management of blepharitis depends on its type but the treatment approach is the same. The mainstay of blepharitis therapy is improved eyelid hygiene. This alone may enable control of the symptoms and prevention of further complications. Warm compresses with calendula or chamomile decoction for 5 minutes once to twice per day. Application of hot packs helps to soften the scales and crusts. The heat causes the blood vessels to dilate, which improves blood circulation, opens and cleans out the infected gland pores in the eyelid margin. Then the eyelid margins, particularly around the roots of the lashes, must be cleaned up from scales and debris. In addition to warm compresses, patients with posterior blepharitis will need massage of the eyelids after topical anesthetics instillation (Proxymetacain or Oxybuprocain). This helps unblock the meibomian glands and expresses the secretion.

If a bacterial infection is the cause of blepharitis, various topical and systemic antibiotics may be prescribed along with eyelid hygiene. They include Gentamicin, Tobramycin, Tetracycline, Erythromycin, and Ciprofloxacin in either ointment or drop form (the ointment form is more preferable).

Under certain conditions, such as acute excessive nonulcerative inflammation, treatment with topical steroids or an antibiotic-steroid combination may be indicated in either ointment or eye drop form — Tobramycin + Dexamethasone, Dexamethasone + Neomycin sulphate + Polymyxin B, etc.

Prognosis is good although the clinical course of the disorder is often quite protracted.

Complications. In case of ulcerative blepharitis it can be the growth of potentially misdirected new eyelashes (trichiasis), lash loss (madarosis) and scar deformity of the lid margins or tylosis (thickening of the lid margin). In severe cases, it may also cause styes, chalazion, irritation and inflammation of the cornea (keratitis) and conjunctiva (conjunctivitis) or dry eye syndrom.

Prophylaxis is permanent hygiene of the eyelids, right correction of refractive errors, timely and adequate treatment of systemic disorders.

Chalazion

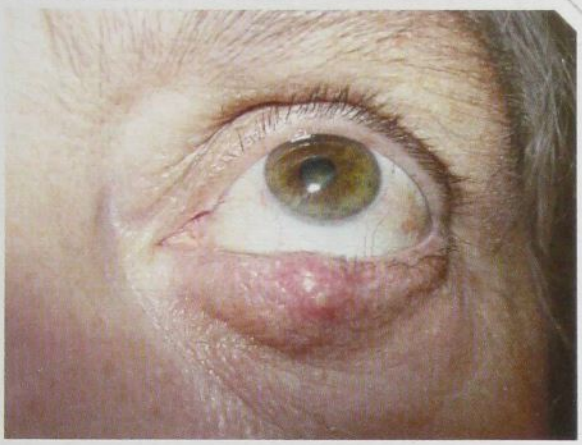


Fig. 4.5. Chalazion

Definition. Chalazion is a chronic non-infectious granulomatous inflammation of the cartilage around the meybomian gland.

Etiology. Chalazion may develop due to inflammation, obstruction, and retained secretions of one of the meibomian glands. There are other medical conditions that may possibly cause chalazion. It is usually secondary to some local inflammation or local infection such as a sty. Cases of recurrent chalazion are frequently caused by chronic blepharitis or poor lid hygiene.

Clinical Picture. Chalazion is a painless movable nodule under the skin (fig. 4.5). Aside from being a cosmetic flaw, it is usually asymptomatic.

Symptoms. Tenderness in the area of swelling, sometimes it causes increased tearing and sensitivity to light. If chalazion is large, it may cause blurred vision, astigmatism by distorting the shape of the eye.

Signs. A roundish firm mobile nodule; signs of infection are absent.

Methods of Examination. Slit-lamp examination, palpation, eyelid eversion.

Differential diagnosis with hordeolum and adenocarcinoma (see table 4.1). Chalazion differes from hordeolum by the absence of hyperemia, and from adenocarcinoma by mobility under the skin.

Treatment. Chalazion is a granulomatous non-infectious condition, thus antibiotics are not indicated. Frequently, a small chalazion will resolve on its own. Steroid ointment and dry heat are used for treatment. If a lesion is recalcitrant and persistent, direct corticosteroid injection can be used for further treatment. If chalazion is large and becomes infected, it usually requires surgical incision and curettage. It is usually removed under local anesthesia. This is usually done from the inside of the eyelid to avoid a scar on the skin.

Prognosis. Chalazion usually responds well to treatment, although some people are prone to recurrences and may require continuing medication. If a chalazion recurs in the same place, you may suggest a biopsy to rule out a more serious problem.

Complications are rare. Large chalazia can induce astigmatism by pressing on the cornea or cause a mechanical ptosis. Complications of operative removal are rare but can include hemorrhage.

Prophylaxis. In predisposed individuals (with blepharitis, acne rosacea, seborrheic dermatitis) regular lid hygiene may have some sort of prophylactic role but the evidence of this is little.

NOTE!

Do not prescribe antibiotics for chalazion as it is a non-infectious inflammation!

Hordeolum (Stye)

Definition. Hordeolum (or stye) is an acute inflammatory infection of an eyelash follicle or sebaceous gland on the eyelid margin.

Etiology. Styes are generally caused by a Staphylococcus aureus bacteria infection, which is responsible for 90—95 % of stye cases. They can be triggered by stress or poor nutrition. Poor hygiene and excessive use of cosmetics may be contributing causes. Stye is often associated with diabetes, gastrointestinal disorders, or acne.

Clinical Picture. A hordeolum is a red, painful lump with a central core of pus (fig. 4.6) near the edge of the lid margin. It can be external or internal. An external stye appears on the margin of the eyelid where the sweat and sebaceous glands, hair follicles are located. An internal stye is a similar infection of the meibomian glands (meibomitis) and it is usually revealed only by everting the eyelid.

A hordeolum begins as a small red bump with a yellow spot at its center as pus expands. It looks like a pimple along the edge of the eyelid. It continues to swell (~3 days) until it breaks open and drains.

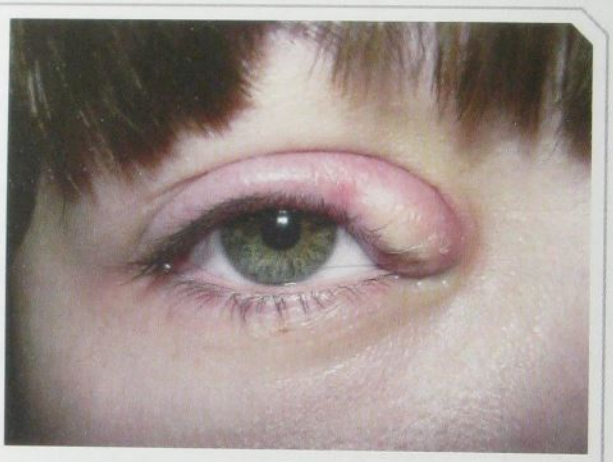


Fig. 4.6. Hordeolum with pus

Fig. 4.7. Hordeolum at the infiltration stage

Symptoms. The patient complains of tenderness and pain at the site of a lump, itching, tearing, sensitivity to light, blurred vision.

Signs. A lump on the eyelid, localized swelling, redness, crusting of the eyelid margins, mucous discharge (fig. 4.7).

Methods of Examination. External, slit-lamp examination, palpation.

Differential diagnosis includes chalazia with their acute onset, fast growth, exquisite tenderness, and location at the lid margin (see table 4.1).

Treatment. Most hordeola will heal spontaneously within 1—2 weeks after swelling, breaking open, and draining on their own. Warm dry compresses (15 minutes, four times per day) and applying of 1 % brilliant green on the stye with a cotton swab can assist with resolution of these lesions at the early stage. A topical ophthalmic antibiotic will hasten

healing (Levofloxacin, Ofloxacin, Tobramycin). When hordeola are multiple and recurrent conduct tests for flora and sensitivity to antibiotics. If the disorder recurs frequently, systemic antibiotic therapy is required as well.

In case of a proclivity for abscess formation the hordeolum has to be dissected. If an internal stye rarely ruptures by itself, it has to be drained surgically.

Prognosis is good. After pus eruption and drainage, the symptoms will rapidly disappear, but hordeolum often recurs.

Complications. Eyelid abcess.

Prophylaxis is good hygiene of the eyelids, avoiding sharing personal towels or facecloths with anyone and making diet changes.

NOTE!

DO NOT squeeze styes as the infection may spread!

NOTE!

DO NOT impose a bandage if you have a hordeolum!

Eyelid Edema

Definition. Eyelid edema is a condition in which the eyelids are swollen due to excess fluid in the connective tissues surrounding the eye. Swollen eyes can be painful and non-painful, and affect both the upper and lower eyelids.

Etiology. Normal eyelid skin is pale pink, delicate, gentle. Edema of the eyelids can have inflammatory and noninflammatory nature, but also occur in allergic processes and injuries (fig. 4.8). Swelling of the eyelids may be caused by local conditions such as insect bites or allergy or by systemic conditions such as cardiovascular disease, renal disease, certain collagen vascular diseases, or thyroid eye disease.

Clinical Picture. Eyelid edema may have few forms:

— Inflammatory edema is usually one-sided and accompanied by other signs of local inflammation, hyperemia, pain, increased skin temperature.

— Swelling in systemic diseases is always bilateral and often combined with swelling of the legs and dropsy. The eyelid skin is pale, induration and pain are absent, local skin temperature is not changed.

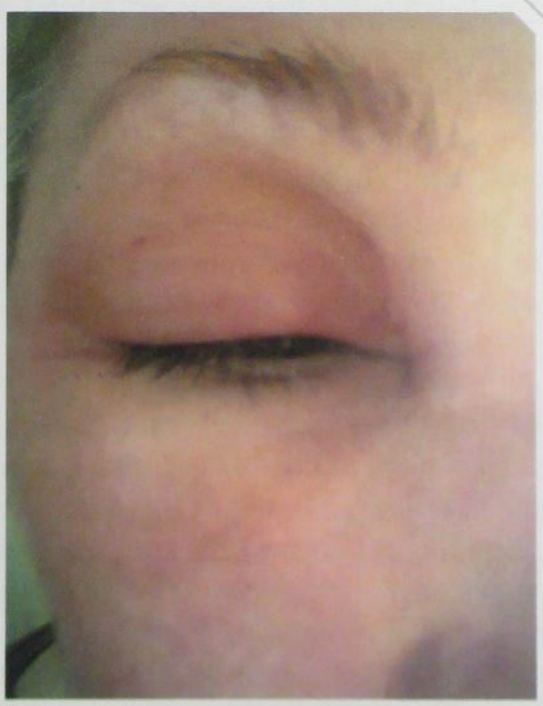


Fig. 4.8. Eyelid edema

— Allergic edema develops rapidly after contact with the allergen and is usually one-sided, can be accompanied by severe itching, skin paleness, watery eyes, not compacted, painless.
— Traumatic edema results from subcutaneous hemorrhage and may be cyanotic or bluish-purple in color.

Symptoms of eyelid edema differ depending on the cause, but one common symptom is swollen eyelids that can be accompanied by irritation, itching, epiphora, redness.

Signs. Inflammatory edema signs are swelling, reddening, sensation of heat; it is painless, usually unilateral. Non-inflammatory edema signs are swelling, pale and cool skin; it is also painless, usually bilateral.

Methods of Examination. External examination, palpation.

Differential Diagnosis. Eyelid cellulitis, orbital cellulitis.

Treatment. Treatment is directed at the underlying disorder. There is no specific treatment for swelling. Generally, if the eyes are swollen due to allergies, antihistamine eye drops or oral allergy medication, as well as lubricating artificial tears will help relieve symptoms. Applying a cool compress sometimes can reduce eyelid swelling. Eyelid swelling caused by viruses and infections is treated with antibiotics, antiviral eye drops, and anti-inflammatory ointments. If the swelling is caused by systemic diseases, their treatment are required.

Prognosis is good, but eyelid edema can be a sign of more serious, potentially vision- or health-threatening problems.

Prophylaxis. Maintaining proper hygiene, timely and adequate treatment of systemic and ocular diseases that can cause eyelid swelling.

Eyelid Abscess

Definition. Eyelid abscess is a limited suppurative inflammation of the eyelid with swelling and subsequent fluctuation.

Etiology. An abscess of the upper or lower eyelid can form as a sequela of infected lid trauma, local inflammatory process (chalazion, furuncle, ulcerative blepharitis), insect sting, or spread of inflammation from the paranasal sinuses.

Clinical Picture. The entire lid with abscess is reddened, has a fluctuating swelling, and is painful to touch. Severe inflammation and swelling often make it impossible to open the eye. The preauricular or submandibular lymph nodes are swollen and tender. The contents of the abscess can fluctuate during the clinical course of the disorder. Spontaneous perforation with pus drainage may occur. Sometimes common systemic symptoms can occur: headache, rise in temperature.

Symptoms. Swelling of the eyelid, redness, pain. Patients also complain of impossibility to open the eye, headache, fever.

Signs. Inflammatory edema, hyperemia of the skin, rise in temperature of the inflamed area, tenderness and pain on palpation.

Methods of Examination. External examination, palpation.

Differential Diagnosis. Hordeolum.

Treatment. For the management of lid abscess dry heat and intensive systemic antibiotic therapy (oral or intravenous) are indicated. Instillation of disinfectant solutions is also the treatment of choice. A stab incision with drainage can relieve tension at the onset of fluctuation. To avoid damaging of the eyelid muscles the opening is made parallel to the lid margin.

Prognosis is generally good.

Complications. Eyelid abscess sequelae include eyelid or orbital cellulitis, sinus thrombosis, which are life-threatening complications.

Prophylaxis. Good hygiene, adequate and timely treatment of other ocular diseases prevent infection spread.

Eyelid Cellulitis (phlegmon)

Definition. Eyelid cellulitis (or palpebral phlegmon) is a diffuse inflammation of the eyelid tissues. It is usually unilateral.

Etiology. The cause of inflammation is infiltration of pyogenic microorganisms into the thickness of the eyelids due to local trauma, insect or animal bites or from the adjacent areas (paranasal sinuses or dacryocystitis) or other inflammatory lesions of the body through the blood flow.

Clinical Picture. Inflammation develops acutely with infiltration in the region of the eyelids and hyperemia of the skin. The eyelid swells, becomes tense, sharply painful and hot to the touch. Swelling spreads to the adjacent areas of the face and the other eyelid. Enlargement of the regional lymph nodes. Increasing body temperature, headache, and malaise. Leukocytosis. After 3—4 days the site of the eyelid cellulitis begins to soften, and soon opens with the release of pus out.

Symptoms. Redness, tenderness, swelling of the entire eyelids, pain. Eyelid burning, watery eyes, blurry vision, discharge of pus, and often mild fever.

Signs. Skin redness, hyperemia, swelling of the eyelids, increased local temperature.

Methods of Examination. External examination, palpation, CT or MRI imaging may be helpful to determine infection severity.

Differential Diagnosis. Eyelid edema, eyelid abscess, orbital cellulitis.

Treatment. General antibiotic and sulfanilamide therapy and detoxification treatment. Antibiotic drops are frequently administered into the conjunctival bag. In the process of infiltration dry heat is prescribed. As fluctuations appear, surgical treatment is performed — cellulitis incision under local anesthesia.

Prognosis. The prognosis is typically very good with treatment. The condition almost always improves quickly with antibiotics.

Complications. Eyelid phlegmon may be complicated by orbital phlegmon, thrombophlebitis of the orbital and facial veins, thrombosis of the cavernous sinus, and purulent meningitis.

Prophylaxis. Good hygiene, adequate and timely treatment of other ocular and systemic diseases prevent the spread of infection.

4.4. Tumors

The eyelids are a highly specialized region of the ocular adnexa consisting of multiple tissue types, all having the potential to give rise to a spectrum of benign and malignant lesions. Accurate identification and classification of these lesions is important for proper care and management. Tumors of the eyelids can be classified based on their origin such as tumors of the epidermis/dermis, tumors of melanocytic origin, and those of glandular, neural, vascular, metastatic, xanthomatous, histiocytic, and inflammatory origin.

In most cases the exact cause of a tumor is not known, but factors which are thought to play a role are:

— the environment — the best example is exposure to sunlight which increases the risk of eyelid skin tumors, particularly in fair-skinned individuals,
— the immune system,
— inheritance — most tumors have a genetic component.

A very wide variety of benign, premalignant, and malignant tumors can occur around the eyelids and the structures adjacent to the eye. The symptoms vary according to the location and nature of the tumor. Tumors of the eyelids are generally visible to the patient, and can sometimes cause local swelling, a localized loss of the eyelashes, and a red or uncomfortable eye if located on the inner surface of the eyelids.

Papilloma

Definition. Eyelid papilloma is the most common benign tumor of the eyelid. It may be broad-based (sessile) or narrow-based (pedunculated) with a corrugated or cerebriform surface (fig. 4.9).

Etiology. Papilomas are derived from squamous cells. As for most malignant lesions, UV (sun) exposure is the main etiologic factor of eyelid papilloma. They may be nonspecific or related to human papilloma virus. It usually occurs in middle-aged or elderly patients.

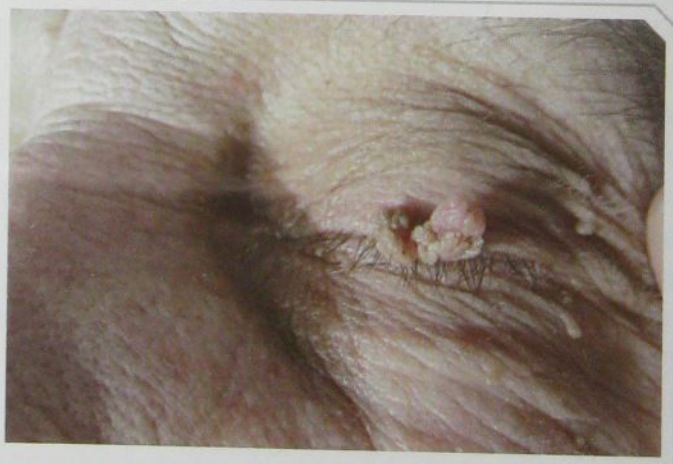


Fig. 4.9. Papilloma

Clinical Picture. Clinically eyelid papilloma can be sessile, pedunculated, solitary or multiple, and is usually of similar color to the adjacent skin. Eyelid papilloma tends to have gradual onset and to progress slowly. Abrupt onset and more rapid growth suggest another diagnosis.

Symptoms. Protrusion from the skin that does not affect vision but brings aesthetic discomfort.

Signs. Broad-based or narrow-based protrusions with irregular surfaces, skin-colored.

Methods of Examination. Slit-lamp examination, external examination for additional lesions, histologic examination is required.

Differential Diagnosis. Basal cell carcinoma, squamous cell carcinoma, seborrheic keratosis, sebaceous gland carcinoma.

Treatment. The standard of treatment for most eyelid papillomas is surgical excision. Electrocautery, chemical cauterization, and laser ablation can be used to remove a papilloma.

Prognosis is excellent. However, the lesions can recur in the same or different location.

Complications. Surgical scarring and possibly eyelid notching are the only likely complications.

Prophylaxis. Protection from the sun's damaging influence, with hats, sunglasses, and protective lotions, and minimization of exposure to the sun. One should see a medical practitioner if any new lesions appear.

Xanthelasma

Definition. Xanthelasma are slightly raised yellow plaques on the eyelids near the inner canthus (fig. 4.10). They are often bilateral and symmetrical.

Etiology. Xanthelasma rsepresent lipid deposits in histiocytes in the dermis of the eyelids. They are frequent in elderly women and occasionally found associated with diabetes, liver problems, and hypercholesterolemia

Clinical Picture. Yellowish deposit of fat underneath the skin. Xanthelasma can be soft, semisolid, or calcareous. Generally, these lesions do not affect the function of the eyelids, but ptosis has been known to occur.

Symptoms. Xanthelasma are usually not itchy or tender. Patients generally complain of aesthetic concers.

Signs. Yellow plagues, ranging in size from 2 to 30 mm. They are flat-surfaced and have distinct borders.

Methods of Examination. External examination, blood test to check cholesterol levels, biopsy of the growth can be made.

Differential Diagnosis. Wegener granulomatosis, xanthoma, milia cysts.

Treatment. Dietary restriction and pharmacologic reduction of serum li-

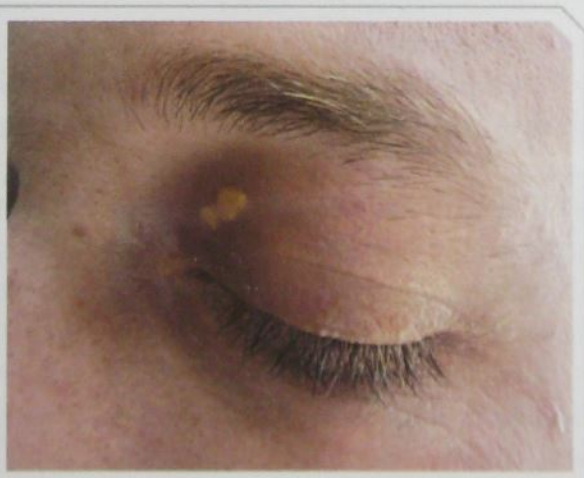


Fig. 4.10. Xanthelasma

pids, although it shows only limited response. Xanthelasma can be removed surgically or treated with a laser.

Prognosis. The condition itself is harmless. The prognosis depends on any association with underlying lipid abnormalities and cardiovascular risk.

Complications. If left untreated, the nodules usually do not give rise to any complications. Problems may arise if conditions, such as high blood cholesterol, are not addressed. Patients can be at risk of heart ailments and other disorders that are associated to high blood lipid levels.

Prophylaxis. Making changes to the diet and taking medication may be recommended to reduce the cholesterol level.

Molluscum Contagiosum

Definition. Molluscum contagiosum is a viral skin infection that is characterized by the formation of small white of reddish nodules on the skin with a central opening.

Etiology. The disease is caused by a poxvirus (molluscum contagiosum virus) and typically affects young or immunocompromised persons. This is a highly contagious disease that can easily spread from one person to another through close skin to skin contact. People may also get infected with this virus by sharing personal items with an infected person. Clothes, towels, and infected clothing articles are often found to be the cause of infection.

Clinical Picture. Dome-shaped, waxy epidermal nodules with central umbilication form and, if present on the eyelid margin, may cause a secondary follicular conjunctivitis. There may be a single or numerous nodules. The size ranges from that of a pin head to 1.5—2.0 mm.

Symptoms. Appearance of small bumps that are painless, but itchy and tender.

Signs. Small, dome-shaped bumps with dimpling and white or waxy substance in the center of the lesion.

Methods of Examination. Diagnosis is based on the appearance of the lesion and can be confirmed by biopsy.

Differential Diagnosis. Epidermal cyst, keratoacanthoma.

Treatment. In most cases, molluscum contagiosum doesn't need to be treated. The bumps usually go away on their own in 6 to 9 months. But in some cases, they may last much longer — sometimes even for years. Otherwise the nodule can be removed by means of processing with iodine or brilliant green. Therapeutic modalities include topical application of various medications, antiviral therapy, and immune response stimulation. Individual lesions may be removed surgically, by laser therapy, cryotherapy or curettage.

Prognosis is generally excellent because the disease is usually benign and self-limited. Spontaneous resolution generally occurs within 18 months in immunocompetent individuals.

Complications. Persistense, spread or recurrence of lesions or secondary bacterial skin infection.

Prophylaxis. Personal hygiene, avoiding direct contact with the skin lesions, no sharing of towels or other personal items such as razors and make-up with other people.

Cutaneous Horn

Definition. Cutaneous horn is a funnel-shaped or conical growth from the skin that is composed of compacted keratin.

Etiology. Cutaneous horns usually arise on sun-exposed skin in elderly men, usually in their fifties, but true causes for the formation of cutaneous horns remain unknown as they can appear on other parts of the body.

Clinical Picture. Cutaneous horns are yellow or brown in color and usually range in size from a few millimeters to a few centimeters. They are curved, which is why they are referred to as horns. They are typically twice as long as the width of the base.

Symptoms. There are really no symptoms associated with this medical condition aside from the cone-shaped protrusion from the skin.

Signs. A hard, firm horny outgrowth of epidermal origin.

Methods of Examination. Diagnosis is confirmed with horn biopsy.

Differential Diagnosis. Actinic keratosis, warts, keratoacanthoma, seborrheic keratosis, pyogenic granuloma, basal cell carcinoma, and squamous cell carcinoma.

Treatment. The cutaneous horn should be surgically removed because it can develop into malignant tumor.

Prognosis of the cutaneous horn is generally good.

Complications. If treatment is delayed, cancerous degeneration is possible.

Prophylaxis. No effective preventive measures; sun safety, early detection and early treatment are the key to prevention.

Basal Cell Carcinoma

Definition. Is a type of skin cancer that arises from basal cells, which line the deepest layer of the epidermis. It frequently occurs at the eyelid margins, at the inner corner of the eyes, and on the upper cheeks. It is the most common form of cancer.

Etiology. Ultraviolet rays from the sun or from a tanning bed are the main cause of basal cell carcinoma. It is why most basal cell cancers occur on the skin that is regularly exposed to sunlight or other ultraviolet radiation.

Clinical Picture. Basal cell carcinoma usually appears as a flat, firm, pale area that is small, raised, pink or red, semi-transparent, shiny, and waxy, and the area may bleed following a minor injury. It may have one or more visible and irregular blood vessels, an ulcerative area in the center that often is pigmented, and black-blue or brown areas. Large basal cel carcinoma may have oozing or crusted areas. The lesion grows slowly, is not painful, and does not itch. If located along the lid

margin, eyelashes may be missing in the region of the tumor. Extension to the posterior eyelid surface may be detected by everting the eyelid. Although basal cell carcinoma does not tend to metastasize, it may be locally invasive.

Symptoms. Patients presenting with basal cell carcinoma often report a slowly enlarging lesion that does not heal and that bleeds when traumatized.

Signs. Flat ulcerated lesion with central depression and rolled border, pink and scaly, semi-transparent, telangiectatic vessels.

Methods of Examination. The diagnosis is based on biopsy results.

Differential Diagnosis. Moluscum contagiosum, squamous cell carcinoma, melanoma (see table 4.1).

Treatment is complete surgical resection with histologic control of the margins. Radiation and cryotherapy, which have higher recurrence rates than surgical resection, are used when surgery is not appropriate or possible.

Table 4.1

Differential Diagnosis of Chalazion, Hordeolum, and Basal Cell Carcinoma

Clinical Signs	Hordeolum	Chalazion	Basal Cell Cancer
Rapid beginning	+	–	–
Unnoticeable beginning	–	+	+
Slow development	–	+	+
Local temparature	+	–	–
Local pain	+	–	–
Photophobia	+	–	–
Tearing	+	–	–
Purulent discharge from the eye	+	–	–
Eyelid swelling	+	–	–
Eyelid hyperemia	+	–	–
Purulent focus near the basis of the eyelash	+	–	–
Round-shaped, painless growth not attached to the skin	–	+	–
New growth with dense edges, covered with crust, bleeding after its removal	–	–	+
Yellowish content, surrounded by a net of slightly widened vessels, is seen when looking from the conjunctiva	–	+	–
New growth is localized mainly on the lower eyelid near the inner area of the palpebral fissure	–	–	+

Prognosis. Most of these cancers are cured when treated early. Some basal cell cancers return. Smaller ones are less likely to come back. Basal cell carcinoma almost never spreads to other parts of the body.

Complications. Basal cell skin cancer almost never spreads. But, left untreated, it may grow (spread) into the surrounding areas and nearby tissues and bone.

Prophylaxis. Avoiding the sun and using sunscreen may help protect against basal cell carcinoma.

Eyelid Adenocarcinoma

Definition. Malignant tumor of the eyelids, which covers the eyeball and orbit with metastases to the internal organs.

Etiology. Usually it develops from the meibomian glands, rarely from the Zeiss and Molly glands (fig. 4.11).

Clinical picture. It occurs frequently in men, less frequently in women over the age of 40 years; primarily affects the upper eyelid. Swelling arises imperceptibly as a painless growth under the skin or conjunctiva of the eyelid sized from a millet grain to a pea. In the initial stages it is similar to chalazion. At first there are no changes in the skin. Sometimes there are several nodules. The process is slow. Later in tumor invasion there is observed conjunctival chemosis, ulceration of the tumor from the conjunctiva and eyelid skin. Then there develops an ulcer with black jagged edges. The bottom of the ulcer is bleeding. Often there develops conjunctivitis, simblepharon. With the progression of the process ptosis and limited mobility of the eyelid are observed.

Symptoms. Long-lasting redness or irritation of the eyes.

Signs. Nodal formation reminding chalazion.

Differential diagnoses with chalazion and epithelial skin cancer, which arises from the surface epithelium and has the form of a small nodule on the skin of the eyelids.

Methods of Examination. The diagnosis is based on biopsy results.

Treatment. In the initial stages the tumor is surgically removed within healthy tissues, at relapse — radiotherapy. If the tumor spreads on the eyeball, enucleation is performed; in case of tumor invasion into the orbit — orbital exenteration with pre- and postoperative irradiation.

Methods of Examination. The diagnosis is based on biopsy results.

Prognosis for adenocarcinoma depends on its malignancy. There is tumor of the conjunctiva, sclera, which grows into the eyeball and eye-socket. There are metastases to the parotid and submandibular regional lymph nodes and internal organs.

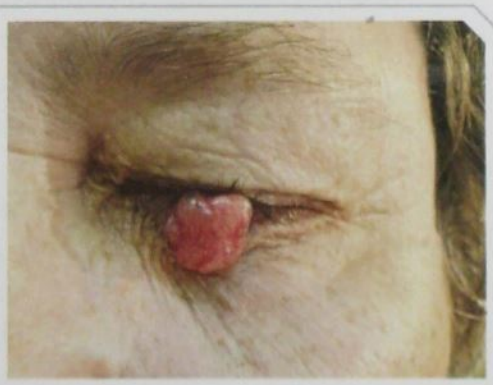


Fig. 4.11. Adenocarcinoma

Complications. It causes death in the late stages.

Prophylaxis. The best way to prevent carcinoma is to protect yourself from exposure to UV rays, practice sun safety, seek shade while being outdoors, avoid sunburn, cover up, wear wide-brimmed hats, use sunglasses with UV protection, and check the skin regularly for new or abnormal growths.

Melanoma

Definition. Melanoma is a relatively rare pigmented eyelid tumor that develops in the cells which produce melanine. It typically appears as a pigmented thickening (tumor) of the eyelid or extension of pigment from the conjunctiva.

Etiology. Melanoma is caused mainly by intense, occasional UV exposure (frequently leading to sunburn), especially in those who are genetically predisposed to the disease. It is a cancer that originates in the pigment-producing cells of the skin (melanocytes).

Clinical Picture. Melanoma can begin as a new, small, pigmented skin growth on a normal skin, most often on sun-exposed areas, or it may develop in preexisting moles. Melanoma readily spreads (metastasizes) to distant parts of the body, where it continues to grow and destroy tissue.

Symptoms. A pigmented skin growth with irregular borders and spots of different colors.

Signs. Typical: asymmetric, irregular borders, changes in color, diameter more than 6 millimeters wide. Some are flat, irregular brown patches containing small black spots. Others are raised brown patches with red, white, black, or blue spots.

Methods of Examination. During examination of the eye, the physician should always evert the eyelid to look for conjunctival involvement. Systemic evaluation for regional or distant metastases is necessary with the diagnosis of melanoma.

Differential Diagnosis. It must be differentiated from nevi and basal cell carcinoma. Suspicious eyelid tumors should be evaluated by biopsy to aid in the diagnosis.

Treatment. Surgical excision with wide margins is the mainstay of early stage melanoma management. Treatment can include systemic chemo- and radiation therapy.

Prognosis. If melanoma is recognized and treated early, it is almost always curable, but if it is not, the cancer can advance and spread to other parts of the body, where it becomes hard to treat and can be fatal.

Complications. While it is not the most common of skin cancers, it causes most deaths.

Prophylaxis. There is no sure way to prevent melanoma, but the best way to lower the risk is to protect yourself from exposure to UV rays, practice sun safety being outdoors — to seek shade, avoid sunburn, cover up, wear wide-brimmed hats, use sunglasses with UV protection, and check the skin regularly for new or abnormal growths.

Review:

1. Key points

Diseases of the eyelids by origin may be congenital or acquired and by the type of the inflammatory process — acute or chronic, by pathogenesis — anomalies of lid position, inflammatory disorders, and tumors. The main symptoms of eyelid diseases are swelling, redness, foreign body sensation, burning, itching, pain. The examination consists of external inspection of the eyelids and eyelid margins, assessment of the eyelid position and the width of the palpebral fissure, palpation, slit-lamp examination, upper and lower lid eversion.

Congenital or developmental anomalies are coloboma, epicanthus, blepharophimosis, ankyloblepharon, distichiasis. The main symptom is anomaly of the palpebral fissure form and size. The main treatment method is surgical reconstruction of structural disorders.

Acquired anomalies of lid position are entropion, ectropion, ptosis, lagophthalmos, trichiasis, etc. In addition to malposition of the eyelids the symptoms are discomfort, pain, irritation, excessive tearing, foreign body sensation, and photophobia. The treatment includes underlying disease cure, surgical correction of anomalies and lubrication or tear preparation instillation for cornea protection.

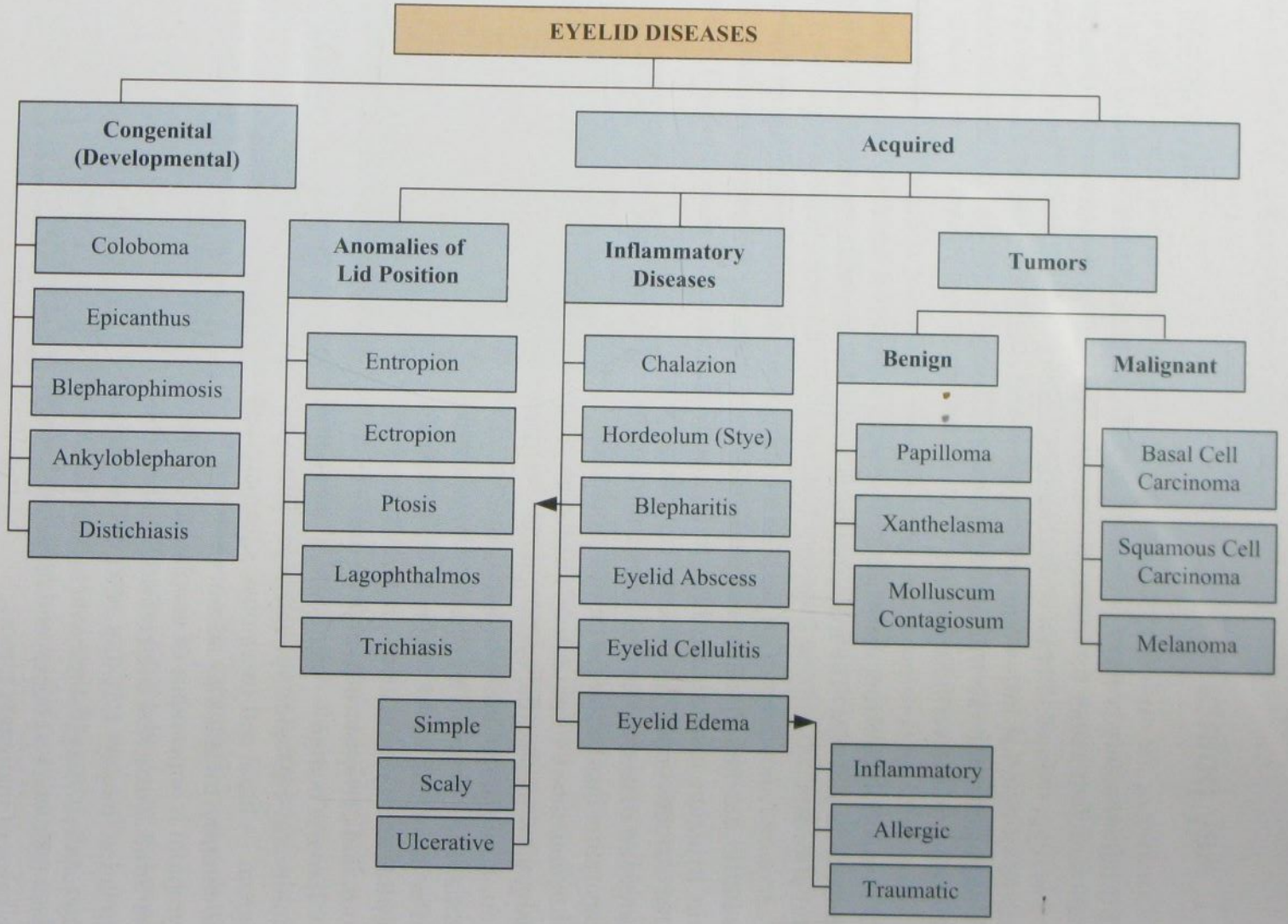
Inflammatory diseases may affect the eyelid margins — blepharitis; eyelid glands — chalazion, hordeolum; skin — edema, abscess, cellulitis. The symptoms and methods of treatment differ according to the origin of the pathology. Topical anti-inflammatory preparations, antibiotics, and corticosteroids may be used.

Tumors: benign — papilloma, xanthelasma, molluscum contagiosum, cutaneous horn; malignant — basal cell carcinoma, squamous cell carcinoma, melanoma, adenocarcinoma. The symptom is appearance of neoplasms of different forms, sizes, and colors. For diagnosis confirmation biopsy, CT, and MRI investigation are required. Surgical removal is used for treatment followed by chemo- and radiation therapy in cases of malignant tumors.

EYE FACTS

The red-eyed tree frog has a third eyelid called the nictitating membrane, which protects the eye but still allows to see through it.

Many species of fish, amphibians, reptiles, birds, and mammals also have third eyelids.


EYELID DISEASES
Congenital (Developmental)
Coloboma
Epicanthus
Blepharophimosis
Ankyloblepharon
Distichiasis
Acquired
Anomalies of Lid Position
Entropion
Ectropion
Ptosis
Lagophthalmos
Trichiasis
Inflammatory Diseases
Chalazion
Hordeolum (Stye)
Blepharitis
Eyelid Abscess
Eyelid Cellulitis
Eyelid Edema
Simple
Scaly
Ulcerative
Inflammatory
Allergic
Traumatic
Tumors
Benign
Papilloma
Xanthelasma
Molluscum Contagiosum
Malignant
Basal Cell Carcinoma
Squamous Cell Carcinoma
Melanoma

3. The Review Questions

A. Control Questions

1. What main examination methods of the eyelids do you know?
2. What eyelid developmental anomalies do you know?
3. What are the main treatment methods of eyelid developmental anomalies?
4. Enumerate anomalies of eyelid position and their etiology.
5. What is blepharitis? Dwell on its etiology, and clinical forms.
6. What are the principles of blepharitis treatment?
7. What are the main features that distinguish chalazion from hordeolum?
8. What complications of hordeolum do you know?
9. What can cause eyelid swelling?
10. What are differential features of inflammatory and non-inflammatory eyelid edemas?
11. What types of eyelid tumors do you know?
12. What is xanthelasma? Dwell on its etiology, methods of treatment and prevention.
13. What is molluscum contagiosum? What are the methods of its treatment?
14. What malignant eyelid tumors do you know? What are their clinical pictures?
15. What are the principles of treatment and prevention of malignant eyelid tumors?

B. Tests

1. **Symptoms of blepharitis are:**
   - A. Inflammation of the eyelid margins
   - B. Loss of eyelashes
   - C. Resistant long duration
   - D. Formation of scales at the root of the eyelashes
   - E. Exophthalmos
   - F. Eyelid swelling

2. **The key to successful treatment of blepharitis is:**
   - A. Elucidation of the etiology of the disease
   - B. Systematic, regular, and long treatment
   - C. Correction of refractive error
   - D. Balanced diet
   - E. Eyelid hygiene
   - F. All of these activities

3. **The main factors that contribute to the emergence of hordeolum are:**
   - A. Paresis of the trigeminal nerve
   - B. Nervous stress
   - C. Beriberi, weakening of the body after infection
   - D. Long-term work at the computer
   - E. Loss of eyelashes
   - F. Pathology of the digestive tract

4. **Which of the following pathological conditions are NOT the cause of blepharitis?**
   A. Pathology of the digestive tract
   B. Endocrine and metabolic disorders
   C. Hookworm infestation
   D. Refractive errors (hyperopia, astigmatism)
   E. Paresis of the oculomotor nerve
   F. Uncorrected refractive anomalies

5. **Congenital anomalies of the eyelids are:**
   A. Blepharochalasis
   B. Ankyloblepharon
   C. Coloboma
   D. Blepharophimosis
   E. All the listed pathologies
   F. None of these diseases

6. **Which complications of entropion and ectropion do you know?**
   A. Dystrophic keratitis
   B. Viral conjunctivitis
   C. Glaucoma
   D. Cataract
   E. Coloboma of the eyelid
   F. Corneal ulcer

7. **Which are the symptoms of eyelid ptosis?**
   A. Maceration of the skin of the eyelids
   B. Wide open eyeball
   C. Ulceration of the ciliary edge of the eyelid
   D. Swelling of the eyelids
   E. Drooping of the upper eyelid
   F. Narrowing of the palpebral fissure

8. **Types of blepharitis:**
   A. Bacterial, viral, fungal
   B. Simple, squamous, ulcer
   C. Simple and complicated
   D. Inflammatory and non-inflammatory
   E. Purulent and simple
   F. Allergic

9. **What factors can cause blepharitis?**
   A. Unfavorable hygienic conditions
   B. Chronic diseases of the gastrointestinal tract
   C. Wide open eyeball
   D. Paresis of the oculomotor nerve
   E. Chronic diseases of the cardiovascular system
   F. Loss of eyelashes

10. **What glands of the eyelids do you know?**
    A. Extradermal and intradermal
    B. Crypts of Henle
    C. Moll, Zeiss, meibomian
    D. Salivary
    E. Sweat
    F. All of these

C. Clinical Cases

Case 1

A patient, 37 years old, complains of the presence of a tumor in the area of the upper eyelid of the left eye. Visual acuity of both eyes is 1.0. Examination of the medial third of the upper eyelid of the left eye showed a clearly demarcated tumor with a diameter up to 4 mm, of tightly elastic consistency, immovable and not painful on palpation. The eyelid skin above it is not changed and mobile, the conjunctiva in this area is yellow. The anterior part of the eye is unchanged. The optical media are transparent. What is the diagnosis?

Case 2

A patient, 36 years old, complains of inability to close the palpebral fissure of the left eye. The patient had a history of neuritis of the facial nerve two years ago. Visual acuity of both eyes is 1.0. Objectively: the palpebral fissure of the left eye is wider than the right one and is not closed. There is an asymmetry of the face. The conjunctiva is normal. The anterior part of the eye is not changed. The optical media are transparent. The fundus is normal. What is the diagnosis?

Case 3

Visual acuity of patient's both eyes is 1.0. The palpebral fissure of the left eye is moderately narrowed. The skin of the eyelid is red, mainly in the outer half. Near the edge of the lid margin there is a limited lump with a central core of pus that is very painful to the touch. The conjunctiva of the eyelid is red, rough, and swollen. The fundus of the eye is unchanged. What is the diagnosis?

Case 4

A 9-year-old patient complains of itching of the eyelids, lacrimation, burning, foreign body sensation, frequent blinking, and fatigue in both eyes lasting for two years. Visual acuity of both eyes is 1.0. Objectively: the edge of the eyelids is thickened, red. On the edge of the eyelids the eyelashes are glued, there are scars and an abnormal growth of eyelashes. The anterior part of the eye is not changed. What is the diagnosis?

Case 5

Patient G., 16 years old, is throwing his head high while walking. The upper eyelids of both eyes are lowered. Visual acuity of both eyes is 1.0. The eyes are healthy. What is the diagnosis?

Case 6

A child is one year old. In the medial third of the eyelid of the right eye there is a clearly demarcated tumor, its palpation is painless. Its movement is limited; the tumor is elastic to the touch. The skin over it is not changed or soldered. What kind of illness do you think it can be?

Case 7

Patient K., 14 years old, went to see a doctor due to edema of the eyelids of the right eye. The swelling appeared suddenly last night. There are no subjective complaints. The day before the patient ate chocolate. There is severe itching of the inner surfaces of the forearms. Current visual acuity of both eyes is 1.0. The eyelids of the right eye are mildly swollen, the skin over it is reddened, of normal temperature (to the touch). The palpebral fissure is narrowed. The conjunctiva is not injected. The anterior segment has no visible changes. The fundus is normal. The left eye is healthy. What is the presumptive diagnosis?

Case 8

Patient A. suffers from pain and edema in the eyelids. About a week ago on the lower eyelid of the right eye a stye appeared, which was treated with heat. The stye doesn't resolve. Currently there is a swelling of the lower eyelid, the skin in this area is tense, hyperemic, hot to the touch. The palpebral fissure is narrowed. There is a purulent discharge from the conjunctiva. Palpation of the outer third of the lower eyelid is sharply painful and determins fluctuation. Visual acuity of both eyes has not changed. Make a diagnosis.

Case 9

Patient S., 16 years old, complains of redness and thickening of the eyelids, itching. Visual acuity of both eyes is 0.8, and with correction with spherical glasses of +1.75 D it is 1.0. The eyelids are hyperemic, thickened. There is frothy discharge in the corners of the eyes. Palpation of the eyelid margins is completely painless. Frequent blinking is noted. The conjunctiva of the eyeball is hyperemic. There meibomian glands are translucent and thickened. What diagnosis can be made?

Case 10

Patient I., 80 years old, had a sudden attack of headache. The eyelids of the left eye are not closing. There is marked facial asymmetry and smoothness of the nasolabial folds to the left. The lower eyelid is not adherent to the eyeball. When you try to close the eyelids, the palpebral fissure stays opened. Visual acuity of both eyes is 1.0. Make a diagnosis.

CHAPTER 5

Disorders of the Lacrimal System

OBJECTIVES

Upon completion of the chapter the students should:

— know the main pathologies of the lacrimal system and their classification;
— know the basic methods of lacrimal system pathology diagnosis;
— know the main symptoms and clinical picture of lacrimal system diseases;
— evaluate and manage patients with diseases of the lacrimal system;
— know the principles of treatment for lacrimal system disorders;
— know the specifics of the clinical course of lacrimal system disease complications.

Plan:

1. CLASSIFICATION OF LACRIMAL SYSTEM DISORDERS
2. SYMPTOMS OF LACRIMAL SYSTEM DISEASES
3. EXAMINATION METHODS
4. DISORDERS OF THE LACRIMAL SYSTEM
   - **4.1. Pathology of the Lacrimal Glands**
     - Disorders of Tear Formation
       - Decreased Tear Formation (Dry Eye)
     - Diseases of the Lacrimal Gland
       - Acute Dacryoadenitis
       - Chronic Dacryoadenitis
       - Tumors of the Lacrimal Gland
   - **4.2. Disorders of the Tear Drainage System**
     - Punctal or Canalicular Stenosis or Obstruction
     - Canaliculitis
     - Diseases of the Lacrimal Sac
       - Acute Dacryocystitis
       - Chronic Dacryocystitis
       - Neonatal Dacryocystitis
       - Tumors of the Lacrimal Sac
   - **4.3. Watering Eye**

1. Classification of Lacrimal System Disorders

Lacrimal system disorders can be classified:
— according to the origin — congenital and acquired;
— according to the nature of the pathologic process — acute and chronic;
— according to the pathogenesis — inflammatory, developmental, tumors, traumas;
— according to the anatomy:
— diseases of the lacrimal glands — dacryoadenitis (acute and chronic), tumors;
— disorders of the tear drainage system — stenosis and obstruction of the lacrimal passages, canaliculitis, eversion of the lacrimal punctum, etc.;
— diseases of the lacrimal sac — dacryocystitis (neonatal, acute, chronic, tumors).

2. Symptoms of Lacrimal System Diseases

— Conjunctival hyperemia.
— Tearing.
— Swelling of the upper eyelid.
— Swelling and redness of the inner corner of the eye.
— Discharge from the lacrimal punctum.
— Mild decrease of vision.

3. Examination Methods

— External examination.
— Inspection of the lacrimal punctum.
— Schirmer's test.

— Tear film break-up time (TBUT).
— Rose bengal staining.
— Fluorescein dye staining.
— Lissamine green staining.
— Probing and irrigation.
— Dacryocystography.

4. Disorders of the Lacrimal System

The lacrimal system consists of the lacrimal gland, accessory lacrimal glands and the lacrimal drainage system, which includes: the puncta, canaliculi, lacrimal sac and nasolacrimal duct (NLD). The normal condition of the system requires that the quantity of secreted tears would be equal to the eliminated quantity. A healthy precorneal tear film is important for clear vision and general health of the ocular surface.

4.1. Pathology of the Lacrimal Glands

Disorders of Tear Formation

Decreased Tear Formation (Dry Eye)

Definition. Dry eye Syndrome (DES), also known as keratoconjunctivitis sicca (KCS), keratitis sicca, xerophthalmia or simply dry eyes is a multifactorial disease, which is characterized by a disturbance of the tear film that lubricates and nourishes the eye surface and surrounding tissues. This condition occurs when the quantity or the quality of the precorneal tear film is insufficient to lubricate the eye, which leads to its damage. Dry eye is a common and often chronic problem, particularly in older adults. Both eyes are usually affected. Dry eye syndrome is more common in women than in men.

Etiology. Dry eye syndrome may be related to:

— decreased tear production caused by damage or dysfunction of the lacrimal glands;
— incorrect composition of tears where the lipid layer of the tear film is insufficient due to meibomian glands dysfunction;
— increased tear evaporation due to eyelid surface anomalies (the eye can't be closed properly), reduced blinking, windy, smoky or dry air.

Risk factors of DES include dry (low humidity) environment, poor hydration, hormone changes, heredity, systemic disorders, some medications, vitamin A deficiency, eyelid problems, rosacea, allergy, LASIK eye surgery and contact lens wear. Dry eye may be a symptom of systemic diseases such as lupus, rheumatoid arthritis, Parkinson's disease, rosacea or Sjogren's syndrome.

Clinical Picture. As identified by the report of the International Dry Eye Workshop (DEWS) dry eye syndrome is classified according to the severity of symptoms and signs as mild, moderate, severe and extremely severe or Level 1, 2, 3, 4 (table 5.1).

Table 5.1

Levels of Severity of Dry Eye Syndrome (according to DEWS*)

Level	Previous Term	Clinical Features
Level 1	Mild	Mild symptoms; mild conjunctival signs; Schirmer score of more than 10 mm/5 minutes; TBUT less than 12 seconds; no other problems
Level 2	Moderate	Moderate symptoms, mild corneal signs; visual signs in the tear film; Schirmer score between 5 and 10 mm/5 minutes; TBUT between 2 and 7 sec
Level 3	Severe	Severe symptoms; marked conjunctival and corneal signs; Schirmer score of less than 5 mm/5 minutes; TBUT less than three seconds
Level 4	Extremely severe	More severe symptoms; conjunctival scarring; severe corneal erosions; Schirmer score of less than 2 mm/5 minutes; TBUT less than three seconds

* International Dry Eye Workshop (DEWS), 2007.

Complaints. Patients complain of burning sensation in the eyes, foreign body sensation (sandy/gritty), redness, transient blurring of vision, excessive stringy mucus, photophobia, intolerance to drafts and winds, constant awareness about the eye itself, decreased tolerance of reading, working on the computer. The symptoms are typically worse during the night time or awakening from sleep (because sleep decreases tear production), and lessen during cool, rainy, or foggy weather and in humid places, such as in the shower.

Signs:

— conjunctival hyperemia, papillary conjunctivitis, appearance of folds on the temporal side, especially on abduction;

— precorneal tear film is with increased amounts of mucus strands and debris in the upper and lower tear menisci (meniscus floaters) (fig. 5.1);

— the marginal tear strip is reduced and contains mucus and debris (the normal lower tear meniscus height is about 1 mm);

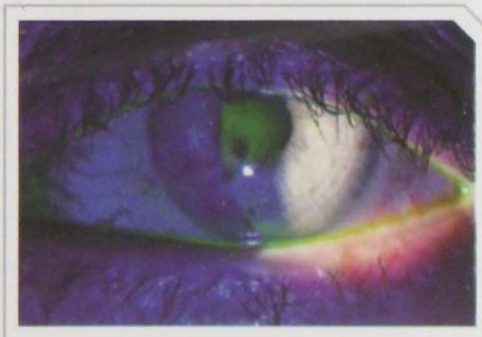


Fig. 5.1. Tear meniscus

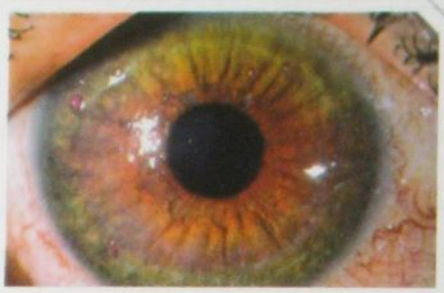


Fig. 5.2. Mucus plaques on the corneal surface stained by the rose bengal

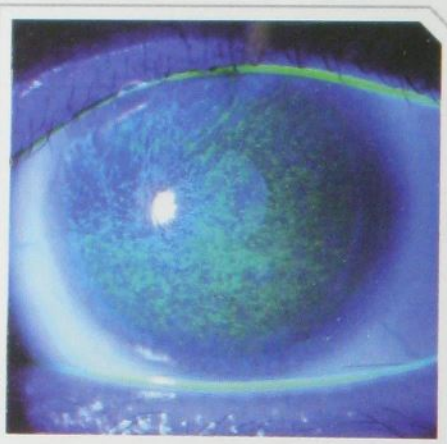


Fig. 5.3. Lissamine green dye in punctated keratopathies

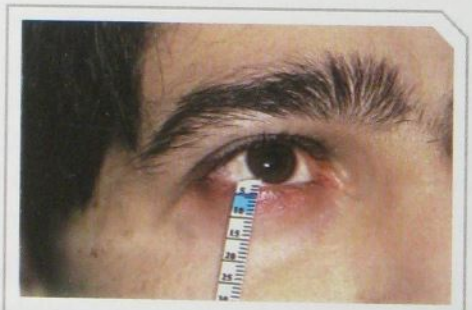


Fig. 5.4. Schirmer's test

— superficial punctate keratitis involving the inferior cornea, which is stained by fluorescein and rose bengal; in severe cases cornea thinning occurs, sometimes with perforation;

— mucus plaques are attached loosely to the corneal surface and are also stained by the rose bengal dye (fig. 5.2);

— filaments are like short 'tails' that hang from the corneal surface.

Methods of Examination. Slit lamp exam, vital dye staining (fluorescein staining, rose bengal staining, alcian blue staining, lissamine green staining (fig. 5.3)), tear film break-up time, Schirmer's test (fig. 5.4), tear osmolarity testing, laboratory tests.

Differential Diagnosis. Atopic and vernal keratoconjunctivitis, blepharitis, contact lens complications, neurotrophic or toxic keratopathy.

Treatment depends on the severity of dry eye syndrome; some people may only require eye lubricants while others may require surgery to help decrease dry eyes (table 5.2). Treatments for dry eyes may include:

1. Environmental management and dietary modification by reduction of the room temperature, usage of humidifiers, avoidance of air-conditioned environment, spending less time on the computer or any monitor, drinking enough water, taking an omega-3 fatty acid.

2. Treatment of associated problems that include ocular allergy, topical and systemic diseases.

3. Artificial tear drops and ointments for tear substitution — Carbomer, Hypromellose, etc.

4. Bandage soft contact lenses — to retain moisture and protect against irritants.

5. Anti-inflammatory agents — topical cyclosporine, topical or systemic corticosteroids, topical or systemic tetracyclines.

6. Stimulation of tear production — bromhexine and eledoisin are sometimes used for stimulation of the lacrimal gland to produce more tears.

7. Autologous serum eye drops made from the patient's own serum.

8. Temporary or permanent blocking of the puncta — to retain tears on the surface of the eye.

9. Major surgical procedures — temporary tarsorrhaphy or botulinum toxin, limbal cell transplantation, conjunctival flap, salivary gland autotransplantation.

10. Psychotherapy — because of the chronic nature of the disease, most patients go through periods of despondency and depression, that's why ophthalmologists must actively encourage these patients to continue to pursue their normal activities.

Table 5.2

Outline of Dry Eye Disease Management (DEWS recommendation)

Level 1	(i) Counselling, (ii) Dietary modification, (iii) Tears substitute (any type) — 4 times/day, (iv) Environmental management, (v) Systemic medication review, (vi) Control of ocular allergy, (vii) Addressing contact lens problems
Level 2	(i) Steps from level one, (ii) Tears supplement — preservative-free (6—8 times), (iii) Oral tetracycline analogues, (iv) Topical anti-inflammatory agents, (v) Topical cyclosporine (0.05 %) — twice daily, (vi) Reversible lacrimal occlusion, (vii) Lid hygiene/nutritional support (omega-3 fatty acid)
Level 3	(i) Steps from levels one and two, (ii) Tears substitute (preservative-free) — 1 hourly, (iii) Autologous serum eye drop (20—50 %), (iv) Bandage contact lens, (v) Permanent punctal occlusion
Level 4	(i) Steps from levels one, two and three, (ii) Tears supplement (P/F) — 1/2 hourly to 1 hourly, (iii) Oral anti-inflammatory medications, (iii) Oral cyclosporine, (iv) Acetyl cysteine 20 % eye drop, (v) Tarsorrhaphy or botulinum toxin, (vi) Limbal cell transplantation (auto/allograft)

Prognosis. The prognosis depends on the underlying cause if there is one. Loss of vision and corneal ulcers are rare.

Complications are uncommon. Conjunctivitis or keratitis sometimes occurs. In severe cases there is corneal ulceration, infection, and corneal perforation (rare).

Prophylaxis. There are some steps to avoid dry eye that include drinking sufficient amounts of water; a healthy diet with vitamins and omega-3 supplements; avoiding windy, dusty and air-conditioned environments, wearing protective glasses, using humidifiers indoors; take "eye breaks" during reading, or working on the computer, or watching TV for a long time; practicing good eye and eyelid hygiene.

Diseases of the Lacrimal Gland

Dacryoadenitis is an inflammation of the lacrimal gland. It may be acute or chronic.

Acute Dacryoadenitis

Definition. Acute dacryoadenitis is an inflammation of the lacrimal gland that is characterized by a rapid clinical course.

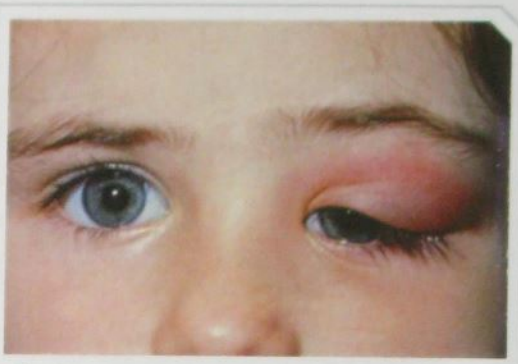


Fig. 5.5. Acute dacryoadenitis

Etiology. Acute dacryoadenitis is a rare inflammatory condition most often seen as a complication of viral or bacterial infection such as mumps, measles, scarlet fever, diphtheria, herpes zoster, Epstein—Barr virus, influenza, staphylococcus, or gonococcus. The blocked duct harbors bacteria and becomes infected.

Clinical Picture. Acute dacryoadenitis usually occurs unilaterally, is characterized by a rapid clinical course (hours to days) and spontaneous healing within ten days. The area around the lacrimal sac is swollen, extremely tender, and red. The upper eyelid exhibits a characteristic S-curve (fig. 5.5). Symptoms such as fever, weakness, preauricular lymphadenopathy or sore throat may be present.

Complaints. The patient complains of pain and swelling in the outer part of the upper eyelid, tenderness, warmth and pain increase upon pressing the swollen region, ocular motility restriction and pain on moving the eye.

Signs. Chemosis, conjunctival injection, erythema of the eyelids, swelling of the lateral third of the upper lid (S-shaped lid), proptosis, mucopurulent discharge, the palpebral lobe of the lacrimal gland is seen as prolapsed and enlarged when the upper lid is everted.

Methods of Examination. External examination of the eye, CT scan.

Differential Diagnosis. Internal hordeolum, eyelid abscess, orbital cellulitis, lacrimal gland tumors.

Treatment depends on the underlying disorder. Warm or cold compresses, UHF-therapy, disinfectant instillation (Furacilin, Sulfacetamide, Myramistin), and local antibiotic ointments (Tobramycin, Gentamicin, Tetracycline) are helpful. In some cases, a short course of oral steroids, oral analgesic may be indicated. In more severe cases oral antibiotics are administered.

Prognosis is good. In most cases acute dacryoadenitis is a self-limiting condition.

Complications. Suppuration may lead to abscess and fistula formation. Degeneration and atrophy of the gland due to acute dacryoadenitis may result in dry eye.

Prophylaxis. Immunization, timely and adequate treatment of underlying factors; some cases cannot be prevented.

Chronic Dacryoadenitis

Definition. Chronic dacryoadenitis is a painless inflammation of the lacrimal gland present for more than a month.

Etiology. Chronic dacryoadenitis is usually caused by chronic conjunctivitis or noninfectious inflammatory disorders such as tuberculosis, sarcoidosis, lymphogranulomatosis, syphilis, thyroid eye disease, leukemia, orbital pseudotumor or Mikulicz syndrome. Chronic dacryoadenitis may be the result of an incompletely healed acute dacryoadenitis.

Clinical Picture. The chronic type is painless, develops slowly and progresses over days to weeks. The lacrimal glands on both sides get enlarged, swelling appears in the outer part of the upper eyelid and keep increasing with time. Usually the upper lid takes a S-curve shape. Tear formation may reduce resulting in dry eyes. The eyelid may droop over the eyes due to swelling, causing difficulty in vision.

Complaints. Swelling in the upper eyelid, redness, burning, difficulty in eye movements, double vision.

Signs. Swelling of the lateral third of the upper lid, enlarged lacrimal gland, which is firm and mobile, mild ptosis, no pain or tenderness over the swelling, conjunctival hyperemia, signs of dry eye.

Methods of Examination. External examination of the eye, CT scan, lacrimal gland biopsy.

Differential diagnosis. Ptosis, chalazion, hordeolum, exophthalmos, eyelid edema, lacrimal gland tumors.

Treatment requires identification of the causative factor and management of the underlying systemic condition. Systemic corticosteroids may be effective in the treatment for unspecific forms. If bacterial infection is present, systemic antibiotics are given. It is rarely necessary to surgically drain the infection.

Local treatment consists of UHF-therapy on the area of the lacrimal gland, instillations of antibiotics, sulfonamides and steroids.

If enlargement of the lacrimal gland persists beyond 2 weeks or the diagnosis is unknown, a biopsy of the gland should be considered.

Prognosis depends on the success of treatment of an underlying systemic condition. Most patients fully recover from dacryoadenitis.

Complications. Abscess of the lacrimal gland.

Prophylaxis. Chronic dacryoadenitis cannot be prevented only by effective treatment of the underlying condition.

Tumors of the Lacrimal Gland

Tumors of the lacrimal gland are relatively rare (about 10—25 % of orbital neoplasms), and show a marked resemblance to parotid gland tumors.

Etiology. The exact underlying causes of lacrimal gland tumors are unknown. These can be genetic or immunologic abnormalities, environmental factors (e.g., exposure to ultraviolet rays, certain chemicals, ionizing radiation), diet, stress, and/or other factors that may play contributing roles in specific types of cancer.

Clinical Picture. Tumors of the lacrimal gland may be benign or malignant. By origin they can be basically grouped into epithelial and non-epithelial lesions, which are subdivided into inflammatory tumors and lymphomas.

The most frequent benign epithelial lacrimal gland tumor is pleomorphic adenoma (benign mixed epithelial tumor). Malignant tumors include adenoid cystic carcinoma and pleomorphic adenocarcinoma.

The clinical picture of lacrimal gland tumors depends on their type. Benign tumors are characterized by slow progressive painless swelling in the upper outer quadrant of the orbit that results in mechanical ptosis. Malignant tumor is a rapidly growing, painful swelling; it causes motility restriction, displacement of the eyeball inferiorly and medially, which can cause double vision, tearing.

Complaints. Unilateral swelling around the eye, fullness of the eyelid, blurry vision, pain around the eye, double vision.

Signs. Proptosis, exophthalmos, a palpable nodule beneath the upper temporal orbital rim, decreased ocular motility, fullness of the upper lid, abnormal (S-shaped) lid contour, diplopia, symptoms of dry eye.

Methods of Examination. External examination, ultrasound studies, CT, MRI, biopsy to confirm what type of tumor it is.

Differential Diagnosis. Exophthalmos, subperiosteal abscess, orbital epidermoid cyst, chronic dacryoadenitis.

Treatment. Most lacrimal gland tumors are removed with surgery followed by symptomatic and supportive treatment, exenteration of the orbit (removal of the entire contents of the orbit) may be necessary. Cancerous tumors need radio- or chemotherapy.

Prognosis depends on the degree of malignancy of the tumor and the stage, at which it is discovered. Adenoid cystic carcinomas have the most unfavorable prognosis. Mortality is increased if the bone is involved.

Complications. Recurrence or malignant change in case of benign tumors.

Prophylaxis. Most cases of lacrimal gland tumors cannot be prevented, but inflammatory lesions can be prevented by timely and adequate treatment of systemic or local inflammatory diseases.

4.2. Disorders of the Tear Drainage System

Punctal or Canalicular Stenosis or Obstruction

Definition. Punctal or canalicular stenosis or obstruction is a condition, in which the external openings of the lacrimal puncta or canaliculi are narrowed or obstructed.

Etiology. These conditions may be congenital or secondary to chronic local inflammations (conjunctivitis, blepharitis, trachoma, herpes simplex); foreign bodies (eyelashes, actinomycosis); scarring (injury, chemical burns, atresia, cosmetics, radiotherapy); certain topical or systemic medications (phospholine iodide drops, systemic 5-FU, etc.), and age-related changes.

EYE FACTS

The normal punctum diameter is 0.3 mm (0.2—0.4 mm). The lower puncta are significantly larger than the upper puncta.

The average diameter of the canaliculi is 0.5 mm.

Clinical Picture. At the beginning the patients notice watery eyes; this condition is progressing over time to the point when it becomes bothersome. The increased tear film causes mild blurring of the vision. There are no other ocular clinical symptoms.

Complaints. Excessive moisture in the eyes, overflow of tears on the cheek, mildly blurred vision.

Signs. Increased tear lake, evident epiphora, punctum stenosis and difficulty in passing the lacrimal dilator or probe.

Methods of Examination. External examination, slit-lamp exam, palpation, fluorescein dye test, probing.

Differential Diagnosis. Nasolacrimal duct obstruction, conjunctivitis, canaliculitis, trichiasis, and ectropion.

Treatment of punctal or canalicular stenosis or obstruction is primarily surgical, depends on the site and grade of obstruction. It involves removal of a foreign body, dilating of the punctum or canaliculus. Several methods can be used to restore drainage of tears: probing; inserting a perforated punctal plug that is kept there for a certain period of time; intubation or stenting; punctoplasty.

If there is a severe or total obstruction of the canaliculi following a major surgical procedure there can be used:

— canaliculodacryocystorhinostomy — the common canaliculus is removed and the remaining canalicular system is directly anastomosed to the lacrimal sac over the silicone stent;

— cojunctivodacryocystorhinostomy (CDCR) — creation of the complete bypass of the tear drainage — a Jones tube is placed through an opening created at the inferior half of the caruncle and then through the osteotomy site into the nasal cavity.

Prognosis. Success rates vary from 40 % to 80 % depending on the etiology.

Complications. Canalicular obstruction may recur following stent removal, ectropion of the lower lid, extrusion of the tube and nasal bleeding may occur.

Prophylaxis includes early ophthalmic consultation and treatment of susceptible patients, especially those with chronic ocular inflammations or those who take topical or systemic treatment with causative medications.

Canaliculitis

Definition. Canaliculitis is an uncommon unilateral inflammation of the lacrimal canaliculus. It affects the lower canaliculus more often than the upper, occurs in adults.

Etiology. Canaliculitis is caused by obstruction within the canaliculus, the presence of a foreign body or the presence of diverticulum, which harbors bacteria within it. The most common cause of canaliculitis is Actinomyces Israelii. However, the other causes can include Aspergillus, Candida albicans, and Herpes simplex virus.

Clinical Picture. Canaliculitis may have two forms — acute or chronic with signs of chronic conjunctivitis, chronic mucopurulent discharge, tearing, ocular surface irritation.

Complaints. A patient complains of a mildly red, swollen and irritated eye, tearing with a slight discharge.

Signs. Epiphora, conjunctivitis, the lacrimal punctum is swollen and inflamed, the canaliculus region is swollen, reddened (especially nasally) and tender to palpation. Pus or granular concrements can be expressed by compressing the canaliculus.

Methods of Examination. Palpation of the medial canthal region and eyelids, slit lamp examination, lacrimal irrigation and instillation of fluorescein.

Differential Diagnosis. Dacryocystitis, nasolacrimal duct obstruction, chronic conjunctivitis.

Treatment. The initial treatment is warm compresses, massage of the local area, topical antibiotics instillation and irrigation of the canaliculus with saline or antibiotic solution.

If the initial treatment unsuccessful, the disease requires surgical treatment — removal of any concretions (canaliculotomy). It is performed by a linear incision into the conjunctival side of the canaliculus, or the eyelid margin, and curetting of the concretions. Silicone stent placement may be also indicated to prevent scarring of the inflamed canaliculus postoperatively. Tincture of iodine may be applied to the lining of the canaliculus after the surgery.

Surgical intervention should be combined with topical antibiotic drops or ointments (Ofloxacin, Levofloxacin, Gentamicin, Tetracycline).

Prognosis. Prognosis is good once the organism is positively identified and appropriately treated.

Complications. Untreated it will result in canalicular stenosis, preseptal cellulitis.

Prophylaxis. There are no specific recommendations for the prevention of canaliculitis, however, proper eyelid and nasal hygiene may decrease the chances of infection.

Diseases of the Lacrimal Sac

Dacryocystitis is an inflammation of the lacrimal sac and the most frequent disorder of the lower lacrimal system. It is often unilateral and always result from a blockage of the nasolacrimal duct. The blocked duct harbors bacteria and becomes infected.

Dacryocystitis may be related to a stenosis within the lacrimal sac, injury, eye infection, nasal inflammation, trauma or age-related changes affecting the eyes and eyelids in older adults. Women are affected 7 times more often than men; the left side is more commonly involved (left : right = 9 : 1).

Dacryocystitis is commonly associated with Staphylococcus aureus, Streptococcus pneumoniae, Pseudomonas species, Mycobacterium, Chlamydia, etc. It may be acute or chronic, by the age of onset — neonatal or adult.

Acute Dacryocystitis

Definition. Acute dacryocystitis is an acute suppurative inflammation of the lacrimal sac.

Etiology. The condition may develop as exacerbation of chronic dacryocystitis or due to direct involvement from the neighboring infected structures such as the paranasal sinuses, dental abscess, tooth caries or poor personal hygiene. The most common

causative microorganisms are Staphylococcus aureus, Streptococcus pneumoniae, Streptococcus pyogenes, Haemophilus influenza, Escherichia coli, etc.

Clinical Picture. It is manifested by the sudden onset of pain, swelling below the inner canthus, often combined with wide-spread cellulitis. It may develop in two days. Symptoms may be accompanied by malaise, fever, and involvement of the regional lymph nodes. The pain may spread as far as the forehead and teeth.

The clinical picture can be divided into three stages: the stage of cellulitis, the stage of lacrimal abscess, and the stage of fistula formation.

Complaints. Excessive tearing, pain, redness, and swelling around the lacrimal sac that is hot, firm and tender, pus discharge from the lacrimal puncta, decreased vision.

Signs. Very tender, tense, red and hot swelling in the region of the lacrimal sac; mucopurulent discharge can be expressed from the punctum.

Methods of Examination. External examination, palpation, CT scan of the orbit and paranasal sinuses, dacryocystography.

Differential Diagnosis. Canaliculitis, orbital or facial cellulitis, hordeolum.

Treatment methods depend on the clinical manifestations of the disease.

The initial treatment at the first stage of the condition consists in application of topical dry warm compresses 3—4 times daily that help to relieve pain and promote drainage accompanied by systemic (Gentamicin, Ciprofloxacin) and topical antibiotics according to the specific pathogens detected (Levofloxacin, Ofloxacin, Ciprofloxacin, Tobramycin). In case of severe pain the patient may need systemic analgesic drugs.

Incision and drainage may be considered if lacrimal abscess is formed. It is best drained by a skin incision, and antibiotics can be changed based on culture results if the initial antibiotic proves ineffective.

After the acute symptoms have subsided, the treatment requires surgery to restore lacrimal drainage — dacryocystorhinostomy, dacryocystectomy.

Dacryocystorhinostomy (DCR) is surgical creation of an opening between the lacrimal sac and the nasal cavity to permit the drainage of tears.

I step — Anesthesia

In adults — local anesthesia of the lacrimal sac and surrounding area:

1. Instillation of 2 drops of 4 % lignocaine hydrochloride into the conjunctival sac.
2. Infiltration: anesthetic agents are injected at the following sites:
   — At the junction of the inferior orbital margin with the beginning of the anterior lacrimal crest: 0.5 ml solution is injected, and then along the line incision to a point of 3 mm above the medial palpebral ligament.
   — The second injection is made at the above point and the needle is directed posteriorly for about 8 mm, and the tissues around the fundus are injected with about 0.5 ml. The needle is then advanced further downwards to the upper half of the posterior lacrimal crest, and 0.5 ml solution is injected.
3. The nasal mucosa of the inferior and middle meatus is anaesthetized with 4 % lignocaine and adrenaline (with a spray or a nasal pack).

In children — general anesthesia.

II step — Surgery Procedure

Nasal pack: In DCR, before starting the operation, a nasal pack is applied on the same side with a roller gauze soaked with 4 % lignocaine, adrenaline and a hemostatic agent. This is for hemostasis during the operation, and to prevent postoperative bleeding.

1. A slightly curved incision with its concavity towards the inner canthus is made (care is taken not to injure the angular vein). The incision is about 2.0 cm, and one-third of its lies above the medial palpebral ligament.
2. Separation of the skin and the orbicularis oculi along the line of incision with a lacrimal dissector, and then retraction with Muller's retractor.
3. The medial palpebral ligament is then exposed, and disinserted from the anterior lacrimal crest with a rougine.
4. The lacrimal sac is separated from the medial wall and floor only. The lacrimal fossa is now exposed.
5. A 10 mm bony ostium is made by cutting the lacrimal bone, part of the adjacent nasal bone, and frontal process of the maxilla. The nasal mucous membrane of the middle meatus is thus exposed.
6. Two horizontal H-shaped incisions are made on the nasal mucosa and the medial wall of the lacrimal sac; and thereby, two anterior flaps and two posterior flaps are created.
7. First the posterior, and then the anterior flaps are sutured with 6-0 or 8-0 chromic catgut or vicryl. Sometime, it is difficult to suture both flaps. In that case, only two anterior flaps are sutured together, while the posterior flaps are excised.
8. The skin is sutured with 5-0 or 6-0 silk.
9. The nasal pack is removed after 48—72 hours.

Dacryocystectomy (DCT) is partial or complete excision of the lacrimal sac.

I step — Anesthesia (local) of the lacrimal sac and surrounding area.

1. Instillation of 2 drops of 4 % lignocaine hydrochloride into the conjunctival sac.
2. Infiltration: anesthetic agents are injected at the following sites:
   - At the junction of the inferior orbital margin with the beginning of the anterior lacrimal crest 0.5 ml solution is injected, and then along the line incision to a point of 3 mm above the medial palpebral ligament.
   - The second injection is made at the above point and the needle is directed posteriorly for about 8 mm, and the tissues around the fundus are injected with about 0.5 ml. The needle is then advanced further downwards to the upper half of the posterior lacrimal crest, and injected with 0.5 ml solution.
3. The nasal mucosa of the inferior and middle meatus is anaesthetized with 4 % lignocaine and adrenaline (with a spray or a nasal pack).

II step — Surgery Procedure

1—3. Same as dacryocystorhinostomy.

4. The lacrimal sac is dissected from the floor, at the fundus, and from its connection with the canaliculi.

5. After the sac is well dissected, it is grasped with straight artery forceps up to its lower end, and twisted until it is torn off from the nasolacrimal duct.

6. The torn end of the nasolacrimal duct is curetted and then cauterized with iodine solution.

7. The skin is closed by interrupted or continuous sutures with 5-0 or 6-0 silk.

8. A pad and a pressure bandage are applied for 24—48 hours. The sutures are removed after 6—7 days.

Prognosis is good if managed promptly and if the surgery is not delayed once the acute phase has resolved.

Complications. Acute inflammation that has spread to the surrounding tissue of the eyelids and cheek entails a risk of lid abscess, orbital cellulitis, facial cellulitis, osteomyelitis of the lacrimal bone, ethmoiditis, sepsis, and cavernous sinus thrombosis, which is a life-threatening complication.

Prophylaxis. There are no specific recommendations for the prevention of dacryocystitis, however, proper eyelid and nasal hygiene may decrease the chances of infection.

Chronic Dacryocystitis

Definition. Chronic dacryocystitis is a persistent or recurring inflammation of the lacrimal sac that persists over weeks or months.

Etiology. Chronic dacryocystitis develops due to an obstruction in the nasolacrimal duct followed by infection. Predisposing factors may be tuberculosis, syphilis, sarcoidosis — originating in the surrounding bones or the nose. Chronic inflammation of the conjunctiva, chronic rhinitis or sinusitis, turbinate hypertrophy, nasal polyps, nasal septum bending, trauma, a foreign body, tumors are common causes. Chronic dacryocystitis may follow an untreated or poorly managed acute dacryocystitis. The most common infecting organism in chronic dacryocystitis, Streptococcus pneumoniae or, sometimes, a fungus such as Candida albicans, is responsible for the infection.

Clinical picture develops slowly and may be divided into four stages:

1. The stage of chronic catarrhal dacryocystitis that is characterized by mild inflammation of the lacrimal sac. In this stage the only symptom is watering eye and sometimes mild redness in the inner canthus.

2. The stage of lacrimal mucocele (distension of the lacrimal sac). It is characterized by constant epiphora associated with a swelling just below the inner canthus, mucous discharge on pressing the swelling.

3. The stage of chronic suppurative dacryocystitis due to pyogenic infections. The condition is characterized by epiphora, mild erythema of the overlying skin of the swelling and purulent discharge.

4. The stage of chronic fibrotic sac (due to persistent infection the lacrimal sac becomes small and fibrotic). Persistent epiphora and discharge are characteristic of this stage.

Complaints. Excessive tearing, painless swelling at the inner corner of the eye, mucous or purulent discharge from the punctum upon pressing the swelling area.

Signs. Epiphora, hyperemia of the inner canthus, pressure to the inflamed lacrimal sac causes extrusion of mucopurulent material through the lower punctum.

Methods of Examination. External examination, palpation, fluorescein dye test, dacryocystography, CT.

Differential Diagnosis. Ethmoid sinusitis, allergic rhinitis, cellulitis, eyelid ectropion, punctual ectropion, lacrimal sac or sinonasal tumor.

Treatment. The purpose of the treatment is to treat an infection and to restore communication between the lacrimal sac and the nasal cavity.

Conservative treatment involves antibiotic (e.g., Ciprofloxacin, Ofloxacin, Levofloxacin) and astringent (e.g., zinc sulphate) eye drops to obtain symptomatic relief.

To remove nasolacrimal duct obstruction a surgery is needed. This involves:

— surgical probing — inserting a fine metal probe via the punctum and canalicular system and passing it into the nasolacrimal sac, past the obstruction;

— dacryocystorhinostomy (DCR) — the surgical procedure of choice, in which a direct communication between the lacrimal sac and the nasal mucosa is created;

— dacryocystectomy (DCT) — partial or complete removal of the lacrimal sac. It should be performed only when DCR is contraindicated or in extreme old age, in cases when the lacrimal sac is shrunken and fibrotic, tumors of the sac;

— balloon dacryoplasty has become popular in the last few years, but may have lower success rates in the long term. It is suitable for patients with focal stenosis or occlusions of the nasolacrimal duct.

Prognosis is generally good with timely treatment, but in cases of treatment delay or in patients with a poorly functioning immune system (immunocompromised) serious complications may develop.

Complications. Chronic conjunctivitis, corneal ulcer, mucocele, or hydrops of the lacrimal sac, acute dacryocystitis, lacrimal abscess, lacrimal fistula, orbital and facial cellulitis, cavernous sinus thrombosis, lacrimal osteomyelitis, etc.

Prophylaxis. Timely treatment of underlying causes, good eyelid and nasal hygiene.

Neonatal Dacryocystitis

Definition. Neonatal or congenital dacryocystitis is an inflammation of the lacrimal sac that usually occurs in newborn infants. The term *dacryocystitis* is not quite correct as it's not a true inflammation of the sac wall but an infection of the retained excretions from the conjunctival sac due to nasolacrimal duct (NLD) blockage.

Etiology. During the prenatal period the NLD is closed by the thin mucous membrane, or the so-called gelatinous tube that reduces by the time of birth or at first weeks of life. If the membrane retains, this causes NLD stenosis or blockage at the distal end of the duct, resulting in tear fluid retention, which provides ideal growth conditions for bacteria, particularly staphylococci, streptococci, and pneumococci.

Clinical Picture. Neonatal dacryocystitis develops shortly after birth when a baby starts to cry with tears. Parents notice lacrimation, swelling in the inner angle of the eye, mucus or pus collection in the inner angle of the eye. In acute stage a child may have weakness, fever, and headache.

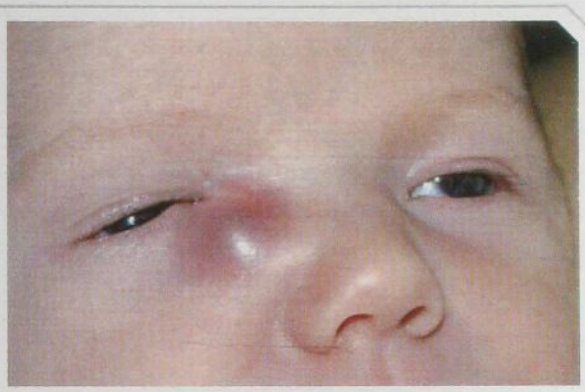


Fig. 5.6. Acute congenital dacryocystitis

Complaints. Tearing, redness, and swelling at the inner corner of the eye, mucus or pus discharge from the lower punctum.

Signs. Epiphora, edema, and redness in the region of the medial epicanthus that is tender, mucopurulent discharge from the puncta on pressing the region of the lacrimal sac (fig. 5.6).

Methods of Examination. External examination, palpation, fluorescein dye test, nasal endoscopy.

Differential Diagnosis. Congenital dacryocystocele, neonatal conjunctivitis, congenital glaucoma.

Treatment depends on the age of the child as in the first month the gelatinous plug may resolve by itself spontaneously.

1. Treatment of dacryocystitis in newborns starts with a massage of the lacrimal passages. It is performed by vibrating or jerky movements with moderate pressure on the area of the lacrimal sac in the direction from top to bottom (from the inner corner of the eye downwards) — 8—10 movements. The aim of the massage is to break down the gelatinous blockage. Massage should be given at least 4 times per day with cleaning of any discharge.

Massage is followed by instillation of disinfectants and antibiotic drops. This conservative treatment cures obstruction in about 90 percent of the infants.

2. If the blockage is not resolved after several weeks to months of this therapy, an ophthalmologist may attempt forceful lacrimal irrigation with normal saline and antibiotic solution. It should be added to the conservative treatment if the condition is not cured up to the age of 2 months. Lacrimal irrigation helps to open the membranous occlusion by exerting hydraulic pressure.

3. If the condition is not cured by the age of 3—4 months (some surgeons prefer to wait till the age of 6—8 months), surgical probing of the NLD is used under general anaesthesia. A thin Bowman's probe is inserted via the lower or upper punctum and canaliculi through the nasolacrimal duct to open any obstruction. Sterile saline is then irrigated through the duct into the nose to make sure that there is now an open path. If there is no improvement after 4 weeks, probing may be repeated.

4. If the initial surgical probing is unsuccessful or if a child is older or has a particularly difficult blockage, another surgical treatment may be recommended — silicone tube intubation, in which a silicone tube is placed in a tear duct to stretch it.

The tube is kept in place for as long as 6 months and then removed in another short surgical procedure.

5. If all the abovementioned methods were not successful by the age of 4 years, dacryocystorhinostomy (DCR) operation should be performed — it helps to create a direct opening from the lacrimal sac into the nasal cavity.

Prognosis is quite good if the condition is managed promptly and surgery is not delayed.

Complications. When not treated in time, it may be complicated by recurrent conjunctivitis, lacrimal sac abscess, and fistulae formation. In aggressive cases it can lead to orbital abscess, meningitis, sepsis, and death.

Prophylaxis. Dacryocystitis is not a preventable disorder, but maintaining good eyelid and nasal hygiene may help.

Tumors of the Lacrimal Sac

Tumors of the lacrimal sac are extremely rare compared to other orbital tumors.

Etiology. The causes of lacrimal sac tumors are not clear; these may be chronic inflammations of the lacrimal sac and surrounding tissues.

Clinical Picture. Approximately 45 % of lacrimal sac tumors are benign and 55 % are malignant and represent a potentially life-threatening condition, so early diagnosis and treatment are particularly important. Most neoplasms are papillomas, mucocele, cysts, polyps, squamous cell carcinomas, transitional cell carcinomas, and adenocarcinomas.

The triad of malignancy are:

— a mass below the medial palpebral ligament;
— a chronic dacryocystitis that irrigates freely;
— expression of bloody mucopus.

In benign tumors the mass grows slowly and is elastic with distinct margins, freely movable under the skin. Malignant tumors grow faster, the mass is firm and fixed to the underlying tissue. Some patients may complain of pain. In advanced cases the involvement of the preauricular, submandibular and cervical lymph nodes can be diagnosed as well as proptosis and limitation of ocular motility.

Complaints. Excessive tearing, swelling, and redness of the inner corner of the eye, sometimes bloody discharge on pressing the swollen region and bleeding from the nose.

Signs. Epiphora, swelling located both above and below the medial canthal tendon, the mass is firm, solid, and incompressible.

Methods of Examination. External examination, palpation, dacryocystography, CT, MRI.

Differential Diagnosis. Chronic dacryostenosis or dacryocystitis.

Treatment. These tumors may carry a vital risk and should be immediately managed with maximum care. The treatment is complete excision of the sac followed by radiotherapy, or chemotherapy.

Prognosis. Benign tumor treatment has a good prognosis if completely excised, while malignant tumors respond to radiotherapy and chemotherapy, have a variable

prognosis depending on the extent of the disease and the type of the tumor. The most dismal prognosis is that of malignant melanoma, which is often fatal in a short period of time in spite of aggressive treatment.

Complications. In any case, all lacrimal sac tumors require careful lifelong follow-up, as recurrence and metastasis may occur many years after initial treatment.

Prophylaxis. Most cases of lacrimal sac tumors cannot be prevented, but timely treatment of chronic inflammation conditions of the surrounding tissues and organs may help.

4.3. Watering Eye

Watering (watery) eye is a condition that is characterized by excessive tearing causing tears run down the cheek. It is not a disease; it is a common symptom of ocular disorders or certain systemic pathology.

Etiology. There are two main causes of watering eye — excessive secretion of tears due to reflex stimulation of the lacrimal gland (*lacrimation*) or poor tear drainage (*epiphora*).

Lacrimation (hypersecretion) may happen naturally in response to emotions, or to cold, windy weather. Otherwise, dry eyes, eye irritation, exposure to bright light, smoke, allergies, foreign body, trauma, and infections can trigger tearing.

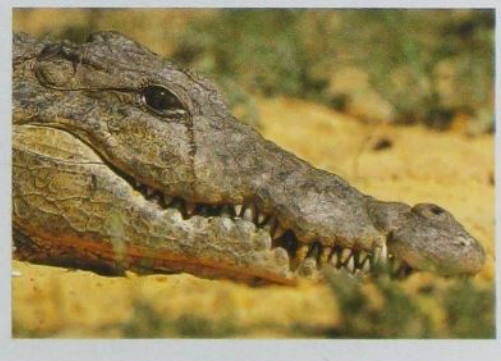
Epiphora (defective drainage) can be caused by:

— mechanical obstruction in the drainage system — narrowed or blocked lacrimal puncta, canaliculi or tear duct, canaliculitis, dacryocystitis, neoplasms of the lacrimal sac, nasal pathology, like polyps, tumors in the inferior meatus of the nose;

— lacrimal pump failure due to lower lid laxity, weakness of the orbicularis oculi, Bell's palsy or ectropion due to other causes.

Methods of Examination. External examination, slit-lamp exam, palpation, syringing, fluorescein dye test, probing test, dacryocystography, CT, nasal endoscopy.

EYE FACTS



CROCODILE TEARS SYNDROME

Spontaneous tearing, usually unilateral, on eating or anticipation of food (parallel with the normal salivation of eating). It occurs when the nerve fibres destined for a salivary gland are damaged and by mistake regrow into a tear gland.

The crocodile tears syndrome most often follows facial paralysis. It is also called Bogorad's syndrome, named after the Russian neuropathologist who described the syndrome.

Treatment. The type of treatment depends on the cause of the watering eye. If dry eye syndrome is a cause of lack of lubrication, artificial tears can help to keep the surface of the eye better lubricated and healthier. In case of infection antibiotic eye drops or ointments are prescribed. If the eyes are watering because of an allergy, antihistamine medication may be prescribed to help reduce the inflammation. There are also some things that can be done to prevent watery eyes, such as wearing protective goggles or sunglasses. If the cause is defective drainage, surgery may be needed. Its type depends on the site and severity of obstruction.

Complications. Undiagnosed and untreated watering eye substantially decreases the patients' quality of life, visual acuity, and impairs social contacts.

Review:

1. Key Points

The lacrimal system consists of the lacrimal gland, accessory lacrimal glands and the lacrimal drainage system, which includes the puncta, canaliculi, lacrimal sac, and nasolacrimal duct. Diseases of the lacrimal system can be caused by disorders of the lacrimal gland functioning (tear formation) or disorders of the tear drainage organs that are classified according to the origin as *neonatal or acquired*, according to the nature of the pathologic process — *acute and chronic*, and also according to the applied anatomy. The main symptoms of lacrimal system diseases are conjunctival hyperemia, tearing or dryness, swelling and redness of the affected areas, swelling of the upper lid, mild decrease of vision. The examination consists of external examination, palpation, slit lamp exam, inspection of the lacrimal punctum, dye staining, Schirmer's test, tear film break-up time, probing, irrigation, dacryocystography.

Dry eye syndrome is a disturbance of the tear film, the main symptoms are redness, burning, foreign body sensation, photophobia, intolerance to drafts and winds. The main treatment methods are dietary modification, treatment of associated problems, artificial tear drops and ointments.

Disease of the lacrimal gland is *dacryoadenitis*, which can be acute or chronic. The main symptom is swelling in the outer part of the upper eyelid (S-shaped lid). The treatment includes UHF-therapy, instillation of disinfectants, antibiotics, systemic corticosteroids and antibiotics according to the underlying factors and severity of the condition.

Punctal or *canalicular stenosis* or *obstruction* is characterized by increased tearing. The main treatment method is surgical reconstruction of the tear drainage system.

Canaliculitis and *dacryocystitis* develop due to obstruction resulting in infection development. Dacryocystitis can be acute, chronic, neonatal. Tearing, swelling of the inner corner of the eye, mucous or purulent discharge from the punctum upon pressing on the swelling area. The main methods of treatment are instillation of antibiotic eye drops and surgical methods such as forceful lacrimal irrigation, probing, dacryocystorhinostomy.

2. Diagrams

LACRIMAL SYSTEM DISORDERS
Disorders of Tear Formation
Dry Eye
Disorders of the Lacrimal Gland
Dacryoadenitis
Acute
Chronic
Tumors
Benign
Malignant
Disorders of the Tear Drainage System
Canaliculitis
Stenosis or Obstruction of the Puncta or Canaliculi
Dacryocystitis
Acute
Chronic
Neonatal
Tumors of the Lacrimal Sac
Benign
Malignant

3. The Review Questions

A. Control Questions

1. What examination methods of the eyelids do you know?
2. Dry eye syndrome, its etiology and clinical picture.
3. What are the principles of dry eye treatment?
4. What is dacryoadenitis, its types, clinical symptoms and treatment methods?
5. What are the main principles of management of punctal and canalicular stenosis or obstruction?
6. What is canaliculitis, its clinical picture and treatment methods?
7. Describe the clinical course of acute dacryocystitis and methods of its treatment.
8. Describe the clinical course of neonatal dacryocystitis and methods its treatment.
9. What are the main causes of tearing, the principles of its treatment?
10. What tumors of the lacrimal system and principles of their treatment do you know?

B. Tests

1. Symptoms of dry eye syndrome are:
A. Redness
B. Burning
C. Mucopurulent discharge
D. Blurring of vision
E. Photophobia
F. Foreign body sensation

2. The treatment methods for dry eye syndrome are:
A. Topical antibiotics
B. Dietary modification
C. Treatment of associated problems
D. Lubricants
E. Dacryocystorhinostomy
F. Tarsorrhaphy

3. Inflammatory diseases of the lacrimal system are:
A. Canaliculitis
B. Papillitis
C. Dry eye
D. Dacryocystitis
E. Dacryoadenitis
F. Meibomitis

4. What are the symptoms of dacryoadenitis?
A. Swelling in the upper eyelid
B. Hyperemia of the inner canthus
C. Tearing
D. Mucopurulent discharge
E. Proptosis
F. S-shaped upper eyelid

5. Signs of acute dacryocystitis are:
A. Tearing
B. Purulent discharge from the lacrimal puncta
C. Proptosis
D. Swelling below the inner canthus
E. Pain of the swollen region
F. Fever

6. **The treatment methods for adult dacryocystitis are:**
   A. Dacryocystectomy
   B. UHF-therapy
   C. Systemic analgesic drugs
   D. Topical antibiotics
   E. Dacryocystorhinostomy
   F. Corticosteroids

7. **The main cause of neonatal dacryocystitis development is:**
   A. Foreign body
   B. Canaliculi stenosis
   C. Lacrimal puncta atresia
   D. Unreduced mucous membrane
   E. Scarring
   F. Nasolacrimal duct stenosis

8. **Initial treatment of congenital dacryocystitis is:**
   A. Probing
   B. Dacryocystorhinostomy
   C. Massage of the lacrimal passages
   D. Antibiotic drops
   E. Forceful lacrimal irrigation
   F. No treatment required

9. **The causes of punctal or canalicular stenosis or obstruction are:**
   A. Dysfunction of the lacrimal gland
   B. Blepharitis
   C. Systemic 5-FU
   D. Foreign bodies
   E. Environmental factors
   F. Scarring

10. **Enumerate the causes of watering eye.**
    A. Ectropion
    B. Allergy
    C. Excessive secretion of tears
    D. Dry eye
    E. Stenosis or obstruction in the drainage system
    F. Nasal polyps

C. Clinical Cases

Case 1

A patient of 15 years of age complains of pain in the upper eyelid and behind the eyeball. During the last 4 hours he notices swelling of the outer part of the upper eyelid, which exhibits a characteristic S-curve. Palpation detects enlarged preauricular lymph nodes. A day before the patient had symptoms of measles. What is the diagnosis?

Case 2

A patient of 53 years of age with a history of sarcoidosis (granulomatous) complains of progressive painless swelling in the outer part of the upper eyelid, drooping of the upper lid. Examination showed swelling of the lacrimal gland that is firm and mobile, mild ptosis, conjunctival hyperemia. What is the diagnosis?

Case 3

A patient of 55 years of age with a history of sarcoidosis (granulomatous) complains of progressive firm painless swelling around the eye, fullness of the eyelid,

forward displacement of the eyeball, blurry vision and double vision. Examination showed proptosis, exophthalmos, a palpable nodule beneath the upper temporal orbital rim, decreased ocular motility, fullness of the upper lid, abnormal (S-shaped) lid contour, diplopia, symptoms of dry eye. CT shows that the mass produces smooth deformities in the adjacent orbital bones. What are the diagnosis and treatment?

Case 4

A 43-year-old patient complains of redness, burning, foreign body sensation in both eyes, intolerance to wind and bright light, blurring of vision. Objectively: conjunctival hyperemia, superficial punctate keratitis, precorneal tear film, Schirmer test — 8 mm/5 min, TBUT — 7 sec. What are the diagnosis and treatment?

Case 5

A mother has noticed in her two-month-old child excessive tearing, swelling in the inner angle of the eye. Examination showed epiphora, edema, and redness in the region of the medial epicanthus that is tender, mucopurulent discharge upon pressing the swelling area. What are the diagnosis and recommendations?

Case 6

A patient of 21years of age during the last 12 hours complains of tearing, a severe pain and marked swelling in the inner angle of the eye that is red, hot, and tender. The swelling spreads to the cheek. Examination also reveals tenderness and enlargement of the regional lymph nodes. What are the diagnosis and treatment?

Case 7

A patient of 31 years of age complains of tearing, slow progressive painless swelling at the inner corner of the eye, mucous or purulent discharge from the punctum upon pressing on the swelling area. What are the diagnosis and treatment steps?

CHAPTER 6

Disorders of the Orbit

OBJECTIVES

To know:

— classification of orbital pathology;
— symptoms and signs of orbital disorders;
— methods of orbital pathology diagnostics;
— causes of orbital diseases;
— management of orbital pathology.

Plan:

1. CLASSIFICATION OF ORBITAL DISEASES
2. CARDINAL SYMPTOMS
3. SPECIFIC EXAMINATION METHODS
4. ORBITAL DISEASES
   - **4.1. Inflammatory Diseases**
     - Periostitis
     - Subperiosteal Abscess
     - Retrobulbar Abscess
     - Orbital Cellulitis
     - Tenonitis
     - Pseudotumor or Idiopathic Orbital Inflammatory Syndrome
   - **4.2. Endocrine Ophthalmopathy**
     - Endocrine Exophthalmos (Thyroid Eye Disease or Graves' Orbitopathy)
   - **4.3. Vascular Disorders**
     - Cavernous Sinus Thrombosis
     - Carotid-Cavernous Fistula
   - **4.4. Tumors**
     - Dermoid Cyst
     - Neurogenic Tumors
     - Optic Nerve Glioma
     - Meningioma
     - Cancer

1. Classification of Orbital Diseases

— By origin — congenital, acquired.
— By the characteristics of the inflammatory process — acute and chronic.
— By pathogenesis — inflammatory, endocrine, vascular, structural, tumors.

1.1. Inflammatory Diseases
- 1.1.1. Infectious — periostitis, subperiosteal abscess, orbital abscess, orbital cellulitis (phlegmon), tenonitis.
- 1.1.2. Non-Infectious
  - 1.1.2.1. Isolated — pseudotumor or idiopathic orbital inflammatory syndrome (IOIS).
  - 1.1.2.2. Systemic — sarcoidosis, Wegener's granulomatosis, systemic lupus erythematosus.
- 1.1.3. Fungal — mucormycosis.
- 1.1.4. Parasitic — trichinosis, echinococcosis, cysticercosis.

1.2. Endocrine Ophthalmopathy — endocrine exophthalmos (thyroid eye disease or Graves' orbitopathy).

1.3. Vascular Disorders — giant cell arteritis, cavernous sinus thrombosis, aneurysms, pulsating exophthalmos.

1.4. Structural Disorders
- 1.4.1. Developmental Deformities — anophthalmia, microphthalmia, craniofascial dysplasia.
- 1.4.2. Traumatic — orbital wall fractures, retrobulbar hemorrhage, intraorbital foreign bodies.

1.5. Tumors
- 1.5.1. Primary
  - 1.5.1.1. Benign — hemangiomas, cysts, osteoma, angioma, optic nerve glioma, meningioma.
  - 1.5.1.2. Malignant — lymphoma, sarcoma, cancer.
- 1.5.2. Secondary and Metastatic — squamous cell carcinoma, mycocele, neuroblastoma, breast carcinoma, prostate carcinoma.

2. Cardinal Symptoms

— Eyeball displacement — exophthalmos, enophthalmos, side displacement.
— Restricted eye movements.
— Diplopia or double vision — due to restriction of the extraocular muscles.
— Decrease of vision — due to compression of the optic nerve or ophthalmic artery, which can eventually affect the patient's eyesight leading to blindness. A reduction in vision also results from central corneal opacification due to exposure keratitis.

Exophthalmos (or proptosis) is abnormal forward displacement (or protrusion, or bulging) of the eyeball out of the orbit (fig. 6.1). It can be either bi- or unilateral. Exophthalmos happens when the muscle and fat lining of the eye socket swells. This can happen with Graves' disease, a thyroid disease that is the most common cause of exophthalmos. A tumor, abnormal blood vessels, and bacterial infection behind the eye can also push the eye forward. It may also result from trauma (such as fracture of the ethmoid bone, which allows air from the sinus to enter the orbital tissue displacing soft tissue and the eyeball).

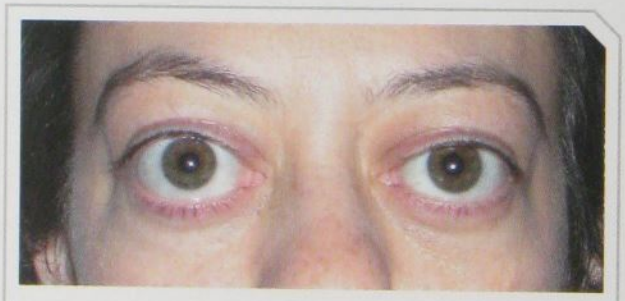


Fig. 6.1. Exophthalmos of the right eye (from http://www.imo.es/en)

Fig. 6.2. Enophthalmos of the left eye (from http://www.imo.es/en)

Exophthalmos is not a separate disease in itself; it is a symptom of many different pathological conditions. It is important that the underlying cause is identified to provide appropriate treatment. If not treated quickly and properly, this infection can spread from the eye socket to the brain. This is an emergency as it may cause permanent loss of sight and can be life-threatening.

Enophthalmos is a relative backward displacement (sinking) of the normal-sized eye in relation to the bony orbital margin (fig. 6.2). It can occur due to an increase of the orbital space or a decrease (contraction) of the orbital content.

Enophthalmos may be uni- or bilateral, but comes to the attention of the clinician more often when unilateral. It is often secondary to orbit-

al trauma, but non-traumatic causes may also occur — structural abnormality, orbital fat atrophy, restrictive muscle syndromes, post-inflammatory restriction and traction due to cicatrization.

3. Specific Examination Methods

— Ocular motility.
— Palpation
— Examination repositioning of the eyeball (orbitotonometry).
— Exophthalmometry.
— Orbital ultrasonography (B-scan, Doppler).
— X-ray study of the orbit (standard X-ray, CT, HCT).
— MRI.
— Contrasting investigative methods (angiography, arteriography, venography).
— Thermography of the orbit.
— Radioisotope scanning of the orbit.
— Diaphanoscopy of the orbit.
— Invasive study of the orbit (intubation, biopsy).
— Consultations of related specialists: ENT, maxillofacial surgeons, neurosurgeons, endocrinologists, oncologists.

4. Orbital Diseases

4.1. Inflammatory Diseases

Inflammatory diseases of the orbit are severe conditions that can lead not only to a loss of visual functions, but also to the death of the patient. The cause of these diseases is usually purulent rhinosinusitis (more than 60 % cases). Moreover, inflammatory diseases of the orbit may develop as a consequence of chronic inflammatory diseases of the maxillofacial area, common infectious diseases (influenza, scarlet fever, syphilis, tuberculosis), and as a result of surgical interventions, traumatic injuries, and panophthalmitis.

Inflammatory diseases of the orbit can be classified as inflammation of the bony walls and periosteum of the orbit (periostitis, osteoperiostitis, subperiosteal abscess), and inflammation of the soft tissues (retrobulbar abscess, orbital cellulitis).

Periostitis

Definition. Periostitis is an inflammation of the periosteum and bony walls of the orbit. It may be acute or chronic. Depending on the stage of inflammation simple and purulent types of periostitis are distinguished. Considering the localization of the pathological focus anterior and posterior types of periostitis are recognized.

Etiology. Most often a trauma or spread of infection from the neighboring structures (paranasal sinuses, eyelids and lacrimal sac lesions), systemic infectious disease caused by migration (tuberculosis, syphilis).

Clinical picture may be of two forms.

Aterior periostitis is characterized by a localized abscess at the orbital margin and possible appearance of a skin fistula.

Posterior (rear, deep) periostitis develops near the top of the orbit. The clinical picture is characterized by infiltration of the eyelids, no axial exophthalmos, chemosis, diplopia, disturbance of the corneal sensitivity, edema and neuritis of the optic nerve with a significant reduction of visual functions.

Complaints depend on the form of the disease. In case of anterior periostitis patients complain of severe pain, tenderness and swelling of the inflamed area. Posterior periostitis is characterized by deep orbital pain, headache, fever, malaise.

Signs. Anterior periostitis: eyelid and conjunctival hyperemia, chemosis of the conjunctiva, restricted ocular motility, fistula formation.

Posterior periostitis: exophthalmos, ptosis, swelling of the eyelids, chemosis, immobility of the eyeball, decreased vision.

Methods of Examination. Diagnosis is confirmed by CT or X-ray.

Differential Diagnosis. Orbital cellulitis, orbital tumors.

Treatment. General treatment of periostitis includes sanation of the primary foci of infection on the background of a systemic combined drug therapy (broad-spectrum antibiotics, sulphanilamides, antihistamines, corticosteroids if necessary). In pain and fever control, analgesic and anti-inflammatory drugs are helpful. In tuberculosis it is required to use anti-tuberculosis drugs, in syphilis — antisyphilitic therapy.

In cases of anterior periostitis local heat and UHF therapy accompanied with local antibiotics are used. When fistula is present, the fistula is resected and bone sequestration is removed. In cases of posterior periostitis, abscess incision and deep drainage of fluid are required.

Prognosis. Generally ends in recovery in case of timely and adequate treatment.

Complications. In severe cases such as backward spread of infection to the brain may be complicated by life-threatening meningitis or cerebral edema.

Prophylaxis. Facial hygiene, timely and adequate treatment of systemic diseases and diseases of the neighboring structures.

Subperiosteal Abscess

Definition. Subperiosteal abscess of the orbit is defined as accumulation of pus between the periosteum and adjacent bony wall of the orbit.

Etiology of subperiosteal abscess includes a wide array of factors: sinusitis, skin infection, bacterial septicemia, orbital/paranasal surgery and penetrating injury. It may be a further step in the development of purulent osteoperiostitis.

The clinical picture of subperiosteal abscess is characterized by acute onset. During the first 48 hours general and local symptoms appear.

Complaints. Eye pain, bulging of the eye, redness of the eye, changes in vision, fever.

Signs. Eyelid swelling, erythema, pyrexia, chills, fatigue, non-axial proptosis and reduction of the eyeball mobility towards abscess localization.

Methods of Examination. CT, HCT or MRI.

Differential Diagnosis. Orbital cellulitis.

Treatment is surgical drainage followed by combined drug therapy (antibiotics, detoxics, disaggregants) and sanation of the primary foci of infection.

Prognosis. Generally ends in recovery in case of timely and adequate treatment.

Complications. In severe cases such as backward spread of infection to the brain may be complicated by life-threatening meningitis or cerebral edema.

Prophylaxis. Facial hygiene, timely and adequate treatment of systemic diseases and diseases of the neighboring structures.

Retrobulbar Abscess

Definition. Retrobulbar abscess is formation of a localized purulent focus posterior to the globe of the eye.

Etiology. Most often a trauma or spread of infection from the neighboring structures (lesions of the paranasal sinuses, eyelids, and lacrimal sac), systemic infectious diseases caused by migration (tuberculosis, syphilis).

Clinical picture of the retrobulbar abscess is characterized by significant signs of intoxication, hyperthermia, and local changes.

Complaints. Pain, eyelid swelling, bulging of the eye, reduction of vision.

Signs manifest as infiltration and hyperemia of the eyelids, proptosis, chemosis, a significant reduction of the mobility, pain in the affected orbit, reduction of visual acuity.

Methods of Examination. CT, HCT or MRI.

Differential Diagnosis. Orbital cellulitis, orbital tumors.

Treatment. Urgent surgical drainage (transcutaneous orbitotomy) on the background of combined drug therapy (antibiotics, sulfonamides, disaggregants, antihistamines, decongestants) and detoxification drugs, sanation of the primary foci of infection.

Prognosis. Generally ends in recovery in case of timely and adequate treatment.

Complications. In severe cases such as backward spread of infection to the brain may be complicated by life-threatening meningitis or cerebral edema.

Prophylaxis. Facial hygiene, timely and adequate treatment of systemic diseases and diseases of the neighboring structures.

Orbital Cellulitis

Definition. Orbital cellulitis, or orbital phlegmon is an acuite diffuse purulent inflammation of the retrobulbar tissue followed by necrosis.

Etiology. It can develop as a result of sinus infection, retrobulbar or subperiosteal abscesses breakthrough, or as a consequence of thrombosis of small orbital vessels.

Clinical Picture. Acute onset with severe symptoms of intoxication and local changes, which are characterized by significant infiltration and hyperemia of the eyelids, incomplete closure of the palpebral fissure, immobility of the eyelids, as well as difficulty of their opening, exophthalmos, full ophthalmoplegia, chemosis (fig. 6.3). Orbital cellulitis is also characterized by a significant decrease in visual function up to blindness (as a result of optic nerve disorders, retinal vascular thrombosis, or choroid).

Complaints. Painful swelling of the eyelids, bulging of the eye, decreased vision, eye pain, fever, painful or difficult eye movements.

Signs. Infiltration and hyperemia of the eyelids, immobility of the eyelids, exophthalmos, full ophthalmoplegia, chemosis, significant decrease of vision.

Methods of Examination. X-ray of the sinuses and surrounding area, CT scan or MRI of the sinuses and orbit. In case of involvement of the eyeball in the inflammatory process it is better to use ultrasound diagnostics (B-scanning).

Differential Diagnosis. Cavernous sinus thrombosis, orbital pseudotumor, Wegener granulomatosis.

Treatment of orbital cellulitis is urgent surgical drainage (transcutaneous orbitotomy) on the background of combined drug therapy (antibiotics, sulfonamides, disaggregants, antihistamines, decongestants, and detoxification drugs) and sanation of the primary foci of infection.

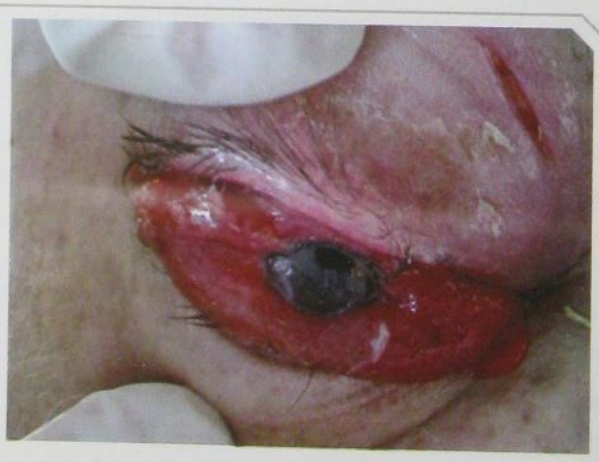


Fig. 6.3. Orbital cellulitis

Prognosis. Orbital cellulitis is a potential life-threating pathology but with prompt treatment the person can recover fully.

NOTE!

Orbital cellulitis is a potentially blinding and life-threatening condition and must not be missed.

Complications. Orbital cellulitis as well as retrobulbar abscess may be complicated by blindness, corneal ulcer, paralysis of the eye muscles, strabismus, ptosis. In case of intracranial spreading of the infection meningitis, encephalitis, brain abscess, cavernous sinus thrombosis, and sepsis can occur.

Prophylaxis. Prompt treatment of the sinus or dental infection may prevent it from spreading to the eyes.

Tenonitis

Definition. Tenonitis is an inflammation of the Tenon's capsule of the eyeball.

Etiology. Tenonitis may be caused by infectious diseases (influenza, measles, scarlet fever, gonorrhea, mumps, tuberculosis, syphilis), systemic diseases (rheumatism, collagen), trauma (including post-operative), inflammation of the eye (ulcers of the cornea, iridocyclitis, endophthalmitis, panophthalmitis, inflammation of the optic nerve).

Clinical Picture. By the nature of exudate tenonitis is classified as serous and purulent, depending on the nature of inflammation — as acute and chronic. Typically, one eye is affected.

Complaints. Feeling of pressure in the orbit, pain that increases with movement of the eyeball.

Signs. Exophthalmos, motility disorders of the eyeball, sometimes diplopia. In case of serous tenonitis, pathological process ends in 3—4 days. In case of purulent tenonitis, exudate breaks into the conjunctival cavity.

Methods of Examination. Clinical ophthalmological examination, MRI.

Differential Diagnosis. Inflammatory disorders of the orbit.

Treatment of tenonitis consists of local and systemic therapy. Dry heat, paraffin, diathermy, radiation quartz, UHF-therapy are effective as local procedures. Instillation of corticosteroids up to 8 times a day, as well as subconjunctival or parabulbar injection are indicated. Also antibiotics, salicylates, antihistamines, vitamins drugs are used. In the case of purulent tendonitis drainage of Tenon's space are carried out.

Prognosis — generally ends in recovery in cases of timely and adequate treatment.

Prophylaxis — facial hygiene, timely and adequate treatment of systemic diseases and diseases of the neighboring structures.

Pseudotumor or Idiopathic Orbital Inflammatory Syndrome

Definition. Idiopathic orbital inflammatory syndrome (IOIS), also known as orbital pseudotumor, is a nonspecific inflammatory process of the orbit without identifiable local or systemic disorders. Nowadays it is divided into three forms: primary idiopathic myositis, vasculitis, and dacryoadenitis. As outcome of all three forms fibrosis of the orbit develops.

Etiology is unknown.

Clinical Picture. The clinical picture is characterized by the development of hyperemia and edema of the eyelids, exophthalmos, restricted mobility reposition of the eyeball, diplopia. The late stage is characterized by exophthalmos, restricted reposition of the eyeball, atrophy of the optic nerve, and affection of the extraocular muscles.

Complaints. Pain in the eye that may be severe, restricted eye movements, decreased vision, double vision, eye swelling (proptosis).

Signs. Painful ophthalmoplegia, exophthalmos, eyelid edema.

Methods of Examination. The most informative methods for the diagnosis are CT, HCT, and MRI. Morphological confirmation of the diagnosis requires a biopsy of the orbital tissue.

Differential Diagnosis. Orbital cellulitis, ruptured dermoid cyst, vasculitis (Wegener's), cancerous tumor.

Treatment is surgical on the background of steroid therapy.

Prognosis. Most cases are mild and outcomes are good. Severe cases may not respond well to treatment and there may be some loss of vision.

Complications. Atrophy of the optic nerve and affection of the extraocular muscles.

4.2. Endocrine Ophthalmopathy

Endocrine Exophthalmos (Thyroid Eye Disease or Graves' Orbitopathy)

Definition. Endocrine exophthalmos is immune-mediated orbitopathy associated with thyroid gland disorders.

Etiology of endocrine exophthalmos is complex and not fully understood. It can affect people with overactivity of the thyroid gland (hyperthyroid), with normally functioning thyroid gland (euthyroid), or even those with underactive thyroid gland (hypothyroid). Thyroid eye disease causes are still unknown, but they are thought to be caused by an abnormal immune response, which targets healthy eye tissues. This can lead to inflammation of the eye socket tissues.

Clinical Picture. There are three main clinical forms of endocrine ophthalmopathy: thyrotoxic exophthalmos, edematous exophthalmos, endocrine myopathy. As a rule, endocrine ophthalmopathy develops in the next direction: the pathological process begins with thyrotoxic exophthalmos, which may progress to swelling, and sometimes can be completed with endocrine myopathy.

The clinical picture of thyrotoxic exophthalmos (fig. 6.4), mainly characterized by a wide-open eye slit caused by retraction of the upper eyelid (Dalrymple's symptom), a decreased amplitude of blinking (Shtelvag symptom), tremor of closed eyelids (Rosenbach symptom), and lag of the upper eyelid when moving the eyes downward (Graefe symptom). Exophthalmos does not exceed 2 mm, usually bilateral (symmetric or asym-

metric). Reposition and movement of the eyeball is not affected. Visual functions are not affected, there are no pathological changes on the eye fundus. There are no structural changes in the soft tissues of the orbit according to computer and magnetic resonance imaging.

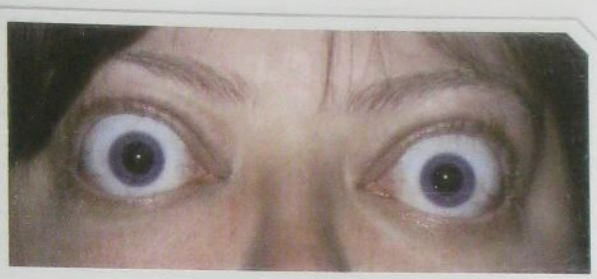


Fig. 6.4. Thyrotoxic exophthalmos (from http://www.eyehealthweb.com)

Edematous exophthalmos is characterized by exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, 'watch-glass' chemosis (chemosis that appears as a large, watery bubble) that along with progression of the pathological changes turns into red, corneal lesions, ophthalmohypertension, optic neuropathy (damage to the optic nerve).

Endocrine myopathy is characterized by a retraction of the upper eyelid, diplopia, strabismus, restricted mobility of the eyeball.

Complaints. Redness of the eyes, swelling watery eyes, sensitivity to light, retraction of the eyelids, protruding eyes, blurry and double vision, reduced vision, limited eye movement, aching (worse in the mornings) behind the eye.

Signs. Exophthalmos, conjunctival injection, chemosis, lid retraction, keratopathy, optic neuropathy.

Methods of Examination The most informative methods for the diagnosis are CT, HCT, MRI, endocrinologist consultation.

Differential Diagnosis. Inflammatory disorders of the orbit, pseudotumor.

Treatment of endocrine ophthalmopathy is a difficult task. It is possible to achieve only a subjective improvement even on the background of intensive treatment (steroid therapy, surgery).

Prognosis depends on how early the disease is diagnosed and how intensive the treatment is. About 1 in 4 people with severe disease will end up with reduced eyesight.

Complications. Atrophy of the optic nerve and affection of the extraocular muscles, corneal affection, and astigmatism.

Prophylaxis. Early detection and treatment of endocrine diseases.

4.3. Vascular Disorders

Cavernous Sinus Thrombosis

Definition. Cavernous sinus thrombosis is an acute clinical syndrome characterized by formation of a thrombus within the cavernous sinus, which can be either septic or aseptic.

Etiology. Septic cavernous sinus thrombosis is a rapidly evolving thrombophlebitic process with an infectious origin (typically from the middle third of the face,

sinuses, ears, teeth, or mouth), affecting the cavernous sinus and its structures. Aseptic cavernous sinus thrombosis is usually a thrombotic process that is a result of traumas, iatrogenic injuries, or prothrombotic conditions.

Clinical Picture. The ophthalmologist will usually diagnose bilateral exophthalmos and episcleral and conjunctival venous stasis in combination with multiple pareses of the cranial nerves. Neurogenic paralysis of all ocular muscles is referred to as total ophthalmoplegia. Where the optic nerve is also involved, the condition is referred to as orbital apex syndrome.

Complaints. Severe pain in the eye and forehead on the affected side, high-grade fever with rigors and vomiting. Swelling and bulging of the eye(s) and surrounding tissues, double vision, restricted eye movements.

Signs. Bilateral exophthalmos, periorbital edema, chemosis, ophthalmoplegia, lateral gaze palsy, mydriasis, decreased visual acuity.

Methods of Examination. CT scan of the head, MRI of the brain, magnetic resonance venogram, sinus X-ray.

Differential Diagnosis. Inflammatory disorders of the orbit.

Treatment. Cavernous sinus thrombosis is treated primarily in the hands of ENT specialists and neurosurgeons. High-dose systemic antibiotic therapy, anticoagulants and corticosteroids are administered. Sometimes surgery is needed to drain the infection. One of the new methods of cavernous sinus thrombosis treatment is endovascular surgery.

Prognosis. Cavernous sinus thrombosis is a life-threating condition if left untreated.

Complications. Meningitis and cerebral abscess may occur, which may lead to death.

Prophylaxis. Timely and adequate treatment of systemic diseases and diseases of the neighboring structures.

Carotid-Cavernous Fistula

Definition. Carotid-cavernous fistula is anastomosis between the internal carotid artery and cavernous sinus. It leads to affection of the venous drainage of the orbit through the upper ophthalmic vein due to increased pressure in the cavernous sinus and throwback of the arterial blood into it.

Etiology. A closed or penetrating head trauma, rupture of a cavernous carotid aneurysm, atherosclerosis.

Clinical Picture. The classic triad of symptoms is considered to be: rapid exophthalmos, pulsating eyes, and blowing noise in the upper inner area of the orbit. However, a third of patients have fixed exophthalmos, in 25 % cases blowing noise can be heard only during phonographic study. Violation of the outflow of venous blood from the orbit leads to red chemosis, increased intraocular pressure, swelling of the retrobulbar fat and extraocular muscles that leads to exophthalmos, reduction of the reposition and mobility of the eyeball (fig. 6.5).

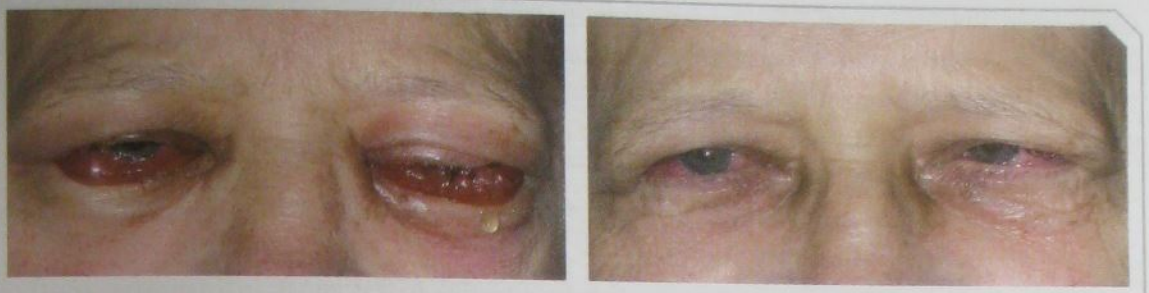


Fig. 6.5. Exophthalmos, chemosis of both eyes due to carotid-cavernous fistula before and after endovascular treatment

There are two types of carotid cavernous sinus fistulas: direct that usually results from a torn carotid artery caused by a head trauma; and indirect that occurs spontaneously and can be caused by a connective tissue disorder.

Complaints. Eye pain or pressure, irritation, bulging eye that may pulse, double vision, hearing a noise in the head (called a bruit).

Signs. Pulsating exophthalmos, chemosis, and subconjunctival hemorrhage, elevated IOP.

Methods of Examination. Doppler ultrasonography, CT scan, MRI.

Differential Diagnosis. Arteriovenous malformation, cavernous sinus thrombosis, cavernous sinus tumors, orbital tumors.

Treatment of carotid cavernous fistula is performed in the neurosurgical department. The mainstay of treatment for carotid cavernous fistula is endovascular therapy. This may be transarterial (mostly in direct cases) or transvenous (most commonly in indirect cases).

Prognosis. Most patients with dural carotid cavernous sinus fistulas are healthy within 6 months after treatment, but patients with direct carotid cavernous sinus fistulas may not experience complete resolution of proptosis, ophthalmoparesis, and visual loss.

4.4. Tumors

Dermoid Cyst

Definition. Dermoid cyst is an overgrowth of normal, non-cancerous tissues in an abnormal location. It is a congenital lesion representing closed cyst lined with ectodermal epithelium and corresponding to the most common orbital tumor in children.

Etiology. No known causes of orbital dermoid exist. Dermoid cysts are believed to arise as embryonic epithelial nests that become entrapped during embryogenesis and pinched at suture lines of bones.

Clinical Picture. Dermoid cysts are present at birth (congenital) and are common. It can be months or years before a dermoid cyst is noticed on a child because the cysts

grow slowly. In the majority of cases it is manifested among children under the age of 5 years. Cyst develops from wandering epithelial cells localized near the bone joints, often subperiosteally in the upper inner quadrant. Content of the cysts is mucous with cholesterol crystals, sometime hair can be present.

Complaints. Painless growth visible in the orbital area.

Sings. A palpable mass that is not attached to the skin, lateral displacement of the eyeball, rarely exophthalmos and ptosis.

Methods of Examination. Computed tomography is the most informative method of research.

Differential Diagnosis. Lacrimal gland tumors, other orbital tumors.

Treatment is surgical. Transcutaneous subperiosteal orbitotomy is indicated.

Prognosis is excellent following successful early surgical intervention.

Complications. Recurrence of tumors with malignancy is a very rare case.

Prophylaxis. There are no preventive methods.

Neurogenic Tumors

Tumors of the optic nerve and its tunics are presented as glioma and meningioma.

Optic Nerve Glioma

Definition. Optic nerve glioma is a tumor of the optic nerve composed of glial cells. Most of these tumors are benign but malignant gliomas of the optic nerve may also occur.

Etiology is unknown. It typically affects children, before age 20. There is a strong association between optic glioma and neurofibromatosis Type 1.

Clinical Picture. The clinical picture of glioma is characterized by slow growth, gradually progressive decrease in visual acuity, axial or lateral painless proptosis, papilledema or atrophy of the optic disc. Glioma of the optic nerve does not affect the dura mater of the optic nerve, but it may spread intracranially.

Complaints. Bulging of the eye, squint, involuntary eyeball movement, vision loss. Sometimes headache, nausea, vomiting, and poor balance depending on the tumor site.

Signs. Exophthalmos, nystagmus, strabismus, decreased vision, swelling of the optic disc, intracranial hypertension.

Methods of Examination. MRI, CT, ultrasound examination.

Differential Diagnosis. Other tumors of the orbit.

Treatment varies depending on the size of the tumor and its localization. Surgery is often used to remove the whole tumor or its part. Although, loss of vision is a common side effect of surgery. Surgery can be followed by radiation and/or chemotherapy if not all of the tumor is removed.

Prognosis is unfavourable for vision.

Complications. Decreased vision, blindness.

Prophylaxis. Genetic counseling may be advised for people with neurofibromatosis-1. Regular eye exams may allow early diagnosis of these tumors before they cause symptoms.

Meningioma

Definition. Meningioma is a tumor that arises from the meninges, especially in the arachnoid cells. Most meningiomas are noncancerous (benign), though rarely a meningioma may be cancerous (malignant).

Etiology is unknown. Most cases are sporadic, appearing randomly, while some are familial.

Clinical Picture. Meningioma is characterized by reduced vision, proptosis, papilledema. Meningioma can spread to the optic nerve sheath, soft tissues of the orbit, and to the intracranial cavity (fig. 6.6).

Complaints. Many people with meningiomas will have no symptoms. Other people develop neurologic symptoms such as seizures, changes in vision or hearing, or arm or leg weakness when the tumor puts pressure on a specific part of the brain.

Signs. Exopthalmos, papilledema, oculomotor palsies, vision loss.

Methods of Examination. CT scan, MRI, and ultrasound scan of the orbit are carried out.

Differential Diagnosis. Other orbital or brain tumors.

Treatment varies depending on the type and location. Surgery is the preferred treatment for most meningiomas, especially for tumors that are large, growing quickly, or causing symptoms. The goal of surgery is to remove as much of the tumor as possible.

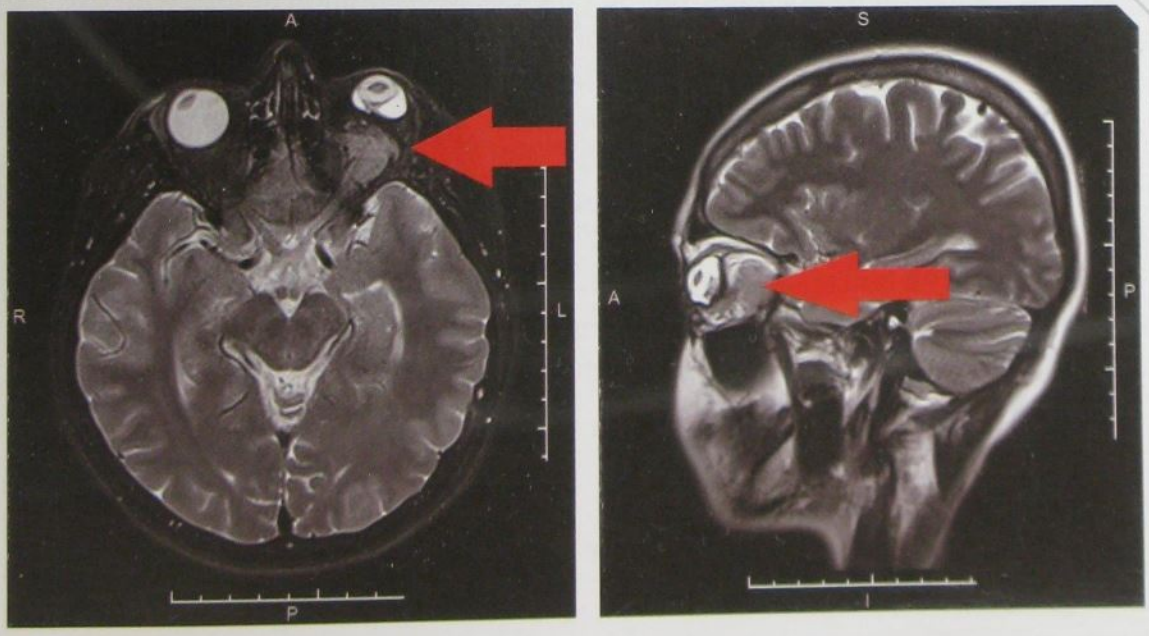


Fig. 6.6. Meningioma of the left orbit on MRI (shown by an arrow)

However, removal of the entire tumor is not always possible, in such cases, the tumor may be only partially removed. In such cases, radiation therapy and chemotherapy are often recommended after surgery to reduce the risk of it coming back.

Prognosis. Completely resected meningiomas usually have an excellent prognosis. Five-year survival for typical meningiomas is more than 80 %; however, this falls to below 60 % in malignant and atypical meningiomas.

Complications. Possible complications of surgery include damage to adjacent normal brain tissue, bleeding, and infection. Other complications include cerebral edema, seizures.

Prophylaxis. There are no preventive methods of meningiomas.

Cancer

Definition. Primary malignant tumor of the orbit is cancer (cancer of the lacrimal gland, primary cancer of the orbit), sarcomas (rhabdomyosarcoma, angiosarcoma, neurosarcoma, liposarcoma, fibrosarcoma, etc.), melanoma, neuroblastoma.

Etiology — unknown.

Clinical picture of primary malignant tumor of the orbit is characterized by diplopia, eyelid edema, and pain in the affected orbit, rapidly growing exophthalmos, degenerative changes in the cornea.

Complaints. Bulging of the eye, vision changes (such as double vision, blurred vision or vision loss), pain, changes to the eye muscle function.

Signs. Exophthalmos, eyelid edema, keratopathy.

Methods of Examination. Diagnosis is based on clinical data, ultrasound scans, computed tomography. Radionuclide scintigraphy and thermography allow differentiating the malignant nature of the tumor. Fine-needle aspiration biopsy with cytology is possible only if the tumor is localized in the anterior part of the orbit.

Differential Diagnosis. Other tumors of the orbit.

Treatment of malignant tumors is combined. Subperiosteal exenteration of the orbit, as a radical method of treatment, as well as external irradiation of the orbit (the most common of them are remote gamma therapy (DHT), X-ray irradiation, and chemotherapy).

Prognosis. The outcome and prognosis ultimately depend on the pathological diagnosis. Most malignant orbital tumors tend to grow into nearby structures.

Prophylaxis. There are no preventive methods of cancer.

Review:

1. Key Points

Diseases of the orbit by origin may be congenital or acquired; by the character of the inflammatory process — acute and chronic; by pathogenesis — inflammatory, endocrine, vascular, structural, tumors.

The main symptoms of orbital diseases are eyeball displacement (exophthalmos, enophthalmos, side displacement), restricted eye movements, double vision, decrease of vision, central corneal opacification due to exposure keratitis. The main diagnostic methods consist of ocular motility examination, palpation, exophthalmometry, orbital ultrasonography (B-scan, Doppler), X-ray study of the orbit, CT, HCT, MRI.

Among *inflammatory processes of the orbit* periostitis, osteoperiostitis, subperiosteal and retrobulbar abscesses, orbital cellulitis, tenonitis, and pseudotumor can be distinguished. Inflammatory diseases of the orbit develop as a consequence of chronic inflammatory diseases of the neighboring structures. They are severe conditions that can lead not only to a loss of visual functions, but also to the death of the patient. General treatment includes sanation of the primary foci of infection on the background of a systemic combined drug therapy.

Endocrine ophthalmopathy can take the form of thyrotoxic exophthalmos, edematous exophthalmos, and endocrine myopathy. Treatment of endocrine ophthalmopathy is a difficult task. It is possible to achieve only a subjective improvement even on the background of intensive treatment (steroid therapy, surgery).

Vascular diseases are cavernous sinus thrombosis, carotid-cavernous fistula. They are acute clinical syndromes and require immediate treatment. The mainstay of treatment is endovascular surgery.

Neoplastic diseases of the orbit include dermoid cyst, optic nerve glioma, meningioma, and cancer. The treatment of the tumors depends on the size of the tumors and their localization. Surgery can be followed by radiation and/or chemotherapy if not all of the tumor is removed during surgery. The outcome and prognosis ultimately depend on the pathological diagnosis.

2. Diagrams:

Orbital Disorders
Inflammation
Periostitis
Subperiosteal Abscess
Retrobulbar Abscess
Orbital Cellulitis
Tenonitis
Pseudotumor
Endocrine Ophthalmopathy
Thyrotoxic Exophthalmos
Edematous Exophthalmos
Endocrine Myopathy
Vascular Diseases
Cavernous Sinus Thrombosis
Carotid-Cavernous Fistula
Tumors
Dermoid Cyst
Optic Nerve Glioma
Meningioma
Cancer

3. The Review Questions

A. Control Questions:

1. What are the cardinal symptoms of orbital diseases?
2. What are the main diagnostic methods of orbital pathology?
3. What inflammatory disorders of the orbit do you know?
4. Orbital cellulitis — clinical picture, methods of examination, treatment basics, prognosis.
5. Pseudotumor — clinical picture, methods of examination, treatment basics, prognosis.
6. Endocrine ophthalmopathy — clinical picture, methods of examination, treatment basics, prognosis
7. Cavernous sinus thrombosis — clinical picture, methods of examination, treatment basics, prognosis
8. Bening tumors of the orbit — clinical picture, methods of examination, treatment, prognosis.
9. Primary malignant tumors of the orbit — clinical picture, methods of examination, treatment, prognosis.
10. Secondary malignant tumors of the orbit — clinical picture, methods of examination, treatment, prognosis.

B. Tests

1. Cardinal symptoms of orbital diseases are:
A. Exophthalmos
B. Enophthalmos
C. Restricted eye movements
D. Side displacement
E. Decrease of vision

2. Exophthalmos (or proptosis) is:
A. Abnormal forward displacement (or protrusion, or bulging) of the eyeball out of the orbit
B. Backward displacement (sinking) of the normal-sized eye in relation to the bony orbital margin
C. Retraction of the upper eyelid
D. Limited mobility of the eyeball
E. Reduction of the reposition and movement of the eyeball

3. Enophthalmos is:
A. Abnormal forward displacement (or protrusion, or bulging) of the eyeball out of the orbit
B. Backward displacement (sinking) of the normal-sized eye in relation to the bony orbital margin
C. Retraction of the upper eyelid
D. Limited mobility of the eyeball
E. Reduction of the reposition and movement of the eyeball

4. For orbital lesion diagnostics the following methods can be used:
A. Exophthalmometry, tonometry
B. Tonometry, tonography, gonioscopy
C. Refractometry, sciascopy, ophthalmometry

D. Visometry, CT, MRI, palpation
E. Ocular motility, exophthalmometry, orbital ultrasonography, X-ray, MRI

5. **Anterior periostitis is characterized by:**
A. Bilateral exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, corneal lesions, ocular hypertension, optic neuropathy
B. Eyelid and conjunctival hyperemia, chemosis of the conjunctiva, restricted ocular motility, fistula formation
C. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis
D. Retraction of the upper eyelid, double vision, strabismus, limited mobility of the eyeball
E. Exophthalmos, reduction of the reposition and movement of the eyeball, redness and swelling of the eyelids, double vision

6. **Tenonitis is characterized by:**
A. Feeling of pressure in the orbit, pain that increases with movement of the eyeball, exophthalmos, motility disorders of the eyeball, sometimes diplopia
B. Eyelid and conjunctival hyperemia, chemosis of the conjunctiva, restricted ocular motility, fistula formation
C. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis
D. Retraction of the upper eyelid, double vision, strabismus, limited mobility of the eyeball
E. Exophthalmos, reduction of the reposition and movement of the eyeball, redness and swelling of the eyelids, double vision

7. **Pseudotumor is characterized by:**
A. Feeling of pressure in the orbit, pain that increases with movement of the eyeball, exophthalmos, motility disorders of the eyeball, sometimes diplopia
B. Eyelid and conjunctival hyperemia, chemosis of the conjunctiva, restricted ocular motility, fistula formation
C. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis
D. Retraction of the upper eyelid, double vision, strabismus, limited mobility of the eyeball
E. Hyperemia and edema of the eyelids, exophthalmos, restricted mobility reposition of the eyeball, diplopia; the late stage is characterized by restricted reposition of the eyeball, atrophy of the optic nerve and affection of the extraocular muscles

8. **Thyrotoxic exophthalmos is characterized by:**
A. Bilateral exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, corneal lesions, ocular hypertension, optic neuropathy
B. Retraction of the upper eyelid, upper eyelid lag when moving

the eyes downwards, bilateral exophthalmos (up to 2 mm), normal visual function

C. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis

D. Retraction of the upper eyelid, double vision, strabismus, limited mobility of the eyeball

E. Exophthalmos, reduction of the reposition and movement of the eyeball, redness and swelling of the eyelids, double vision

9. Orbital cellulitis is characterized by:

A. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis

B. Exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, corneal lesions, ocular hypertension, optic neuropathy

C. Acute onset, unilateral exophthalmos, edema and hyperemia of the eyelids, complete ophthalmoplegia, chemosis, in some cases corneal and optic nerve lesion

D. Retraction of the upper eyelid, diplopia, strabismus, restricted ocular mobility

E. The feeling of pressure and pain in the orbit, exophthalmos, partial ophthalmoplegia, diplopia

10. Cavernous sinus thrombosis is characterized by:

A. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis

B. Exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, corneal lesions, ocular hypertension, optic neuropathy

C. Acute onset, unilateral exophthalmos, edema and hyperemia of the eyelids, complete ophthalmoplegia, chemosis, in some cases corneal and optic nerve lesion

D. Retraction of the upper eyelid, diplopia, strabismus, restricted ocular mobility

E. Bilateral exophthalmos and episcleral and conjunctival venous stasis in combination with multiple pareses of the cranial nerves and also headache, stupor, fever, vomiting

11. Optic nerve glioma is characterized by:

A. Slow growth, a gradual decrease in visual acuity, appearance of axial or lateral painless proptosis, development of optic disc edema or atrophy

B. Exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, corneal lesions, ocular hypertension, optic neuropathy

C. Retraction of the upper eyelid, upper eyelid lag when moving the eyes downwards, bilateral exophthalmos (up to 2 mm), normal visual function

D. Retraction of the upper eyelid, diplopia, strabismus, restricted movement of the eyeball

E. Pulsating exophthalmos, reduction of the reposition and movement of the eyeball, swelling of the eyelids, red chemosis

12. For the treatment of optic nerve glioma the following methods are used:

A. Antibiotic therapy
B. Anti-inflammatory therapy
C. Only surgical treatment
D. Only radiotherapy
E. Surgical removal of the tumor followed by radiotherapy

13. Primary malignant tumor of the orbit is characterized by:

A. Acute onset, unilateral exophthalmos, edema and hyperemia of the eyelids, complete ophthalmoplegia, chemosis, in some cases corneal and optic nerve lesion
B. Exophthalmos (4—5 mm), reduction of the reposition and movement of the eyeball, corneal lesions, ocular hypertension, optic neuropathy
C. Slow growth, a gradual decrease in visual acuity, appearance of axial or lateral painless proptosis, development of edema or atrophy of the optic disc
D. Fast growth, rapid proptosis, diplopia, eyelid edema, pain in the affected orbit, degenerative changes in the cornea
E. Retraction of the upper eyelid, upper eyelid lag when moving the eyes downwards, bilateral exophthalmos (up to 2 mm), normal visual function

14. For the treatment of primary malignant tumor of the orbit the following methods are used:

A. Antibiotic therapy
B. Anti-inflammatory therapy
C. Only surgical treatment
D. Only radiotherapy
E. Surgical treatment followed by radio- and chemotherapy

15. Generally, cancer of the ... metastasizes to the orbit.

A. Brain
B. Liver
C. Rectum
D. Breast and lung
E. Pancreas

C. Clinical Cases

Case 1

A male patient aged 25 complains of pain in the area of the right orbit, edema and redness of the eyelids. Status localis: edema, hyperemia and tensity of the eyelids, redness of the conjunctiva, the palpebral fissure is narrowed, restriction of eyeball motility, exophthalmos. What additional methods of research will help to clarify the type of orbital pathology?

A. Visometry
B. Phosphene diagnostics
C. Orbitotonometry
D. Schirmer's test
E. MRI, CT

Case 2

A patient aged 65 complains of pain in the area of the eyelids and orbit that intensifies during movement of the eye, edema and redness of the eyelids of the left eye. There is weakness, chills, headache, fever up to 38.0 °C. Status localis: edema, hyperemia and tensity of the eyelids, the palpebral fissure is closed, restriction of eyeball motility, exophthalmos, acute tenderness during palpation, chemosis. Define the pathology.

A. Carotid-cavernous fistula
B. Cavernous sinus thrombosis
C. Psudotumor
D. Endocrine ophthalmopathy
E. Orbital cellulitis

Case 3

A female patient aged 73 with hypertension complains of rapid exophthalmos, pulsating eyes and blowing noise in the upper inner area of the orbit. Define the pathology.

A. Carotid-cavernous fistula
B. Cavernous sinus thrombosis
C. Pseudotumor
D. Endocrine ophthalmopathy
E. Orbital cellulitis

Case 4

A female patient aged 32 with diabetes mellitus was admitted to the intensive care unit in stupor, with headache, vomiting, and fever. Status localis: bilateral exophthalmos and episcleral and conjunctival venous stasis in combination with multiple pareses of the cranial nerves, total ophthalmoplegia, edema, hyperemia and tensity of the eyelids, the palpebral fissures are closed. Define the pathology.

A. Carotid-cavernous fistula
B. Cavernous sinus thrombosis
C. Pseudotumor
D. Endocrine ophthalmopathy
E. Orbital cellulitis

Case 5

A male patient aged 45 with Graves' disease complains of a wide-open eye slit. Status localis: retraction of the upper eyelid, a decreased amplitude of blinking, tremor of closed eyelids, upper eyelid lag when moving the eyes downwards, unilateral exophthalmos (2 mm), the visual functions are not affected, there are no pathological changes on the eye fundus. Define the pathology.

A. Thyrotoxic exophthalmos
B. Edematous exophthalmos
C. Pseudotumor
D. Endocrine myopathy
E. Orbital cellulitis

Case 6

A male patient aged 45 with Graves' disease complains of proptosis. Status localis: reduction of the reposition and movement of the eyeball, 'glass' chemosis, ocular hypertension, optic neuropathy. Define the pathology.

A. Thyrotoxic exophthalmos
B. Edematous exophthalmos
C. Pseudotumor
D. Endocrine myopathy
E. Orbital cellulitis

Case 7

A male patient aged 45 with Graves' disease complains of proptosis and diplopia. Status localis: retraction of the upper eyelid, strabismus, restricted mobility of the eyeball. MRI revealed enlargement of all the rectus muscles, without involving its tendons. Define the pathology.

A. Thyrotoxic exophthalmos
B. Edematous exophthalmos
C. Pseudotumor
D. Endocrine myopathy
E. Orbital cellulitis

Case 8

A female patient aged 52 complains of proptosis, redness of the eyelids, diplopia. Status localis: hyperemia and edema of the eyelids, exophthalmos, restricted mobility and reposition of the eyeball. MRI revealed enlargement of the medial rectus muscles, including the tendon. Define the pathology.

A. Thyrotoxic exophthalmos
B. Edematous exophthalmos
C. Pseudotumor
D. Endocrine myopathy
E. Orbital cellulitis

Case 9

A female patient aged 4 complains of painless formation in the upper inner quadrant of the orbit. Define the pathology.

A. Optic nerve glioma
B. Meningioma
C. Pseudotumor
D. Endocrine myopathy
E. Dermoid cyst

Case 10

A female patient aged 5 complains of proptosis, a decrease in visual acuityof the left eye. Status localis: lateral painless proptosis, papilledema of the optic disc. CT-scanning revealed enlarged portion of the optic nerve near the apex of the orbit. Define the pathology.

A. Optic nerve glioma
B. Meningioma
C. Pseudotumor
D. Endocrine myopathy
E. Dermoid cyst

CHAPTER 7

Diseases of the Conjunctiva

OBJECTIVES

Upon completion of the chapter the student should:

— know the classification of conjunctival diseases;
— know the basic diagnostic methods;
— be able to establish the diagnosis of conjunctivitis differentiating it from other causes of red eye;
— know the main symptoms and clinical picture of acute and chronic conjunctivitis;
— identify the cause(s) of conjunctivitis; establish appropriate therapy;
— know the principles of emergency care of conjunctival diseases;
— know the treatment methods of inflammation diseases of the conjunctiva;
— know the types of conjunctiva degeneration;
— know the specific features of the clinical course of conjunctival diseases complications.

Plan:

1. CLASSIFICATION OF CONJUNCTIVAL DISORDERS
2. SYMPTOMS OF CONJUNCTIVAL DISEASES
3. EXAMINATION METHODS
4. DISEASES OF THE CONJUNCTIVA
   - **4.1. Inflammatory Diseases of the Conjunctiva**
     - Infectious Conjunctivitis
       - Bacterial Conjunctivitis
         - Acute Bacterial Conjunctivitis
         - Chronic Bacterial Conjunctivitis
         - Gonococcal Conjunctivitis
         - Diphtheritic Conjunctivitis
       - Viral Conjunctivitis
         - Adenoviral Conjunctivitis
       - Chlamydial Conjunctivitis
         - Paratrachoma
         - Trachoma
     - Non-infectious Conjunctivitis
       - Allergic Conjunctivitis
         - Vernal Keratoconjunctivitis
         - Atopic Keratoconjunctivitis
         - Giant Papillary Conjunctivitis
   - **4.2. Degeneration of the Conjunctiva**
     - Pinguecula
     - Pterygium
   - **4.3. Tumors of the Conjunctiva**
     - Benign Tumors
     - Malignant Tumors

1. Classification of Conjunctival Disorders

Conjunctival disorders can be classified:
— according to the origin — congenital or acquired;
— according to the clinical presentation — mild, moderate or severe;
— according to the onset of the pathological process — acute, subacute or chronic;
— according to the pathogenesis — degenerative, inflammatory, tumors.

1.1. Inflammatory Diseases (Conjunctivitis)
1.1.1. Infectious Conjunctivitis
— Bacterial — acute bacterial conjunctivitis, chronic bacterial conjunctivitis, gonococcal conjunctivitis, diphtheritic conjunctivitis.
— Viral — adenoviral keratoconjunctivitis, epidemic keratoconjunctivitis.
— Chlamydial — chlamydial inclusion conjunctivitis (paratrachoma), trachoma.
— Parasitic.
— Fungal.
1.1.2. Non-infectious Conjunctivitis
— Allergic — vernal keratoconjunctivitis, atopic keratoconjunctivitis, giant papillary conjunctivitis, allergic rhinoconjunctivitis.
— Toxic.
— Irritant.
— Endogenous or autoimmune.

1.2. Degenerations — pinguecula, pterygium, xerosis, etc.

1.3. Tumors:
1.3.1. Benign — dermoid, lipodermoid, papilloma, nevus, hemangioma.
1.3.2. Malignant — carcinoma, lymphoma, Kaposi's sarcoma.

2. Symptoms of Conjunctival Diseases

— Redness of the eye.
— Itching.
— Burning.
— Photophobia.
— Tearing.
— Discharge.
— Foreign body sensation.

3. Examination Methods

— External examination with eversion of the upper and lower eyelids.
— Slit lamp examination.
— Staining.
— Bacteriological laboratory investigation of the discharge.

4. Diseases of the Conjunctiva

4.1. Inflammatory Diseases of the Conjunctiva (Conjunctivitis)

Conjunctivitis is an inflammation of the conjunctiva and is one of the most common eye disorders worldwide that affects children and adults. Conjunctivitis is often called *'pink eye'* or *'red eye'* because when the conjunctiva becomes irritated or inflamed, the blood vessels, which supply it, enlarge and become much more prominent,

so the eye turns red. Conjunctivitis can be present in just one eye, or it can affect both eyes at the same time. Usually conjunctivitis is a minor eye inflammation, but sometimes it can develop into a more serious condition.

Conjunctivitis is most often caused by a virus but also can be caused by bacterial infection, allergies (e.g., cosmetics, pollen) and chemical irritation (contact lens, air pollution, smoke, dust or chemical irritants).

Conjunctivitis can be classified as mild, moderate, severe according to the clinical presentation of the disease. Depending on the onset and duration it can be classified as acute, subacute or chronic.

Clinical signs which have to be considered in the differential diagnosis of conjunctival inflammation are:

1. *Hyperemia (injection)* due to dilatation of the conjunctival blood vessels:
   - — *conjunctival injection* — bright red, clearly visible distended vessels that move with the conjunctiva, decreasing toward the limbus;
   - — *pericorneal injection* — superficial vessels, circular or circumscribed in the vicinity of the limbus;
   - — *ciliary injection* — not clearly discernible, brightly colored nonmobile vessels in the episclera near the limbus;
   - — *mixed injection.*
2. *Discharge (exudate)*:
   - — *watery(serous)* — acute viral, allergic or toxic inflammations;
   - — mucoid — chronic allergic inflammation;
   - — mucopurulent — mild bacterial and chlamydial infection;
   - — purulent — acute bacterial infection.
3. *Conjunctival response:*
   - — *follicular* — viral, chlamydia infections, hypersensitivity to topical medications;
   - — papillary — vernal disease, bacterial infection, contact lens related problem.
4. *Presence of:*
   - — *pseudomembranes* — acute adenoviral infection, gonococcal conjunctivitis, autoimmune conjunctivitis;
   - — true membranes — streptococcal infection, diphtheria.
5. *Presence or absence of lymphadenopathy.*
6. *Presence or absence of eyelid or conjunctival (chemosis) edema.*

Infectious Conjunctivitis

The normal conjunctiva contains microorganisms. Inflammation usually occurs as a result of infection from direct contact with pathogens (such as from a finger, towel, or swimming pool) but also from complicating factors (such as a compromised immune system or injury) or through other means such as sinus or ear infections. Infectious conjunctivitis is a very common condition, and is responsible for 35 % of all eye-related problems. It is most common in children and the elderly.

Bacterial Conjunctivitis

Bacterial conjunctivitis is inflammation of the conjunctiva resulting from bacterial infection. Bacteria may come from the patient's own skin or upper respiratory tract or they may be caught from another person with conjunctivitis. Bacterial conjunctivitis usually begins in one eye and often spreads to the other. The severity of the infection depends on the type of bacteria involved. Organisms that can cause conjunctivitis include: Streptococci; Staphylococci; Pneumococci; Neisseria gonorrhoeae; Neisseria meningitis; Haemophilus influenzae; Corynebacterium diphtheriae; Escherichia coli and so on.

Bacterial infections cause a red eye, which is associated with a grey or yellowish pus or mucopurulent discharge from the eye that may cause the lids sticking together, especially after sleeping. The patient experiences a gritty discomfort rather than pain, foreign body sensation and blurring of vision due to mucous discharge. Clinically bacterial conjunctivitis may be characterized as acute, hyperacute or chronic.

Bacterial conjunctivitis is usually diagnosed on the basis of clinical symptoms, but laboratory examination of the discharge must be obtained to identify the pathogen. Treatment of bacterial conjunctivitis includes irrigation of the eye with normal saline to remove the purulent discharge that accumulates and use of topical and systemic antibiotics.

Acute Bacterial Conjunctivitis

Definition. Acute bacterial conjunctivitis is an infection of the eye that has a rapid onset of signs and symptoms.

Etiology. The most common causative bacteria are: Staphylococcus aureus, Koch-Weeks bacillus in adults and Haemophilus influenzae or Streptococcus pneumoniae in children.

Transmission occurs through direct contact with discharge from the eyes or upper respiratory tracts of infected people, or indirectly through contaminated fingers, clothes, and use of contaminated articles, such as shared eye makeup applicators, towels and topical eye medications. Infection can occur at all ages, but children under 5 are most frequently affected.

Clinical Picture. Acute bacterial conjunctivitis usually begins unilaterally with severe conjunctival hyperemia, lid edema, irritation, tearing, mucopurulent discharge. The second eye typically becomes involved 1—2 days later. The symptoms last less than three or four weeks.

Often Haemophilus influenza is associated with systemic infection, including upper respiratory infection and fever.

Complaints. Redness of an eye, discomfort and foreign body sensation, mild photophobia, yellow or green purulent discharge, that makes lid margins stick together during sleep, slight blurring of vision due to mucous flakes on the cornea. These symptoms have an acute onset and initially present in one eye, in 2 days the process becomes bilateral.

Signs. Diffuse conjunctival hyperemia, bulbar conjunctival injection, lid edema, flakes of pus are seen in the canthi and lid margins, the cilia stick together with yellow crusts (fig. 7.1, 7.2).

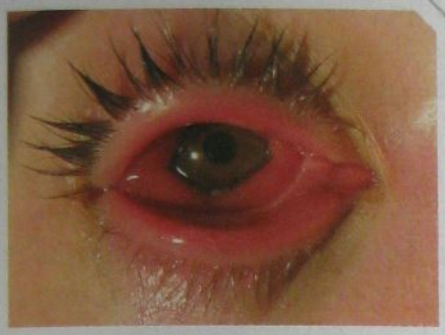


Fig. 7.1. Acute bacterial conjunctivitis with chemosis and purulent discharge

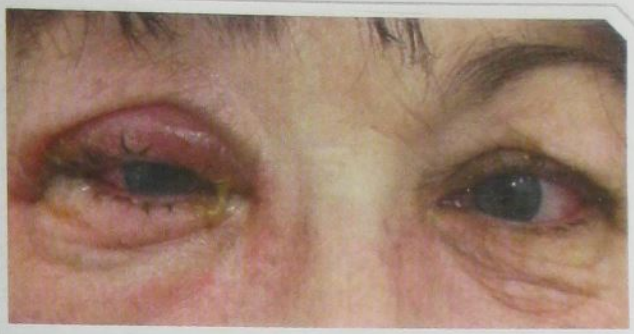


Fig. 7.2. Bacterial conjunctivitis of the right eye with eyelid edema and pus formation

Methods of examination. External examination, slit lamp biomicroscopy, bacteriological laboratory investigation of the discharge and scrapings to diagnose the bacteria type.

Differential diagnosis. Acute allergic or viral conjunctivitis, blepharitis.

Treatment. Treatment with broad-spectrum antibiotics or topical antibiotic combination preparations that cover the full range of pathogens should begin immediately, even before the laboratory results are available.

The eyes at first should be cleaned by irrigation with normal saline to remove the purulent discharge that accumulates and any crusts or stickiness. Then instill antibiotic eye drops or ointments that cover a broad range of Gram-positive and Gram-negative bacteria (Ofloxacin, Ciprofloxacin, Gentamicin, Tobramycin) from three to four times a day for five to seven days.

Prognosis is usually good, although rarely it can progress to complications such as corneal infection or preseptal cellulitis.

Complications. Complications may arise if the cornea is involved, or if the condition is not treated. The complications include: corneal damage, chronic recurrence, chronic blepharitis.

Prophylaxis. Acute bacterial conjunctivitis is a highly infectious disease, capable of spreading in families or organized groups of people. It is best to keep away from work or school if a patient suffers from the condition. Prevention is maintaining good personal hygiene, washing hands before and after contact with the eyes, avoiding hand-eye contact, sharing of pillows, towels, make-up.

Chronic Bacterial Conjunctivitis

Definition. Chronic bacterial conjunctivitis is a bacterial conjunctivitis that lasts for more than 4 weeks.

Etiology. Chronic bacterial conjunctivitis is most commonly caused by Staphylococcus aureus but other bacteria can also be involved. This type of conjunctivitis often develops along with blepharitis or another inflammatory condition, which promotes bacterial growth in the eyelids.

The condition may result from continuation of acute conjunctivitis in the absence of adequate treatment, errors of refraction, nasal or upper respiratory tract catarrh, chronic exposure to dust and smoke, abuse of alcohol, insomnia and metabolic disorders.

Clinical Picture. Symptoms will last for a minimum of four weeks and include frequent episodes.

Complaints. Mild redness of the eye, burning, irritation, foreign body sensation, light sensitivity, morning eyelash crusting or eyelash loss, dryness in the eyes, heaviness in the eyelids, blurry vision.

Signs. Diffused conjunctival hyperemia, congested lid margins, minimal mucopurulent discharge, presence of crusts on the lid margins, lid hyperemia.

Methods of examination. External examination, slit lamp biomicroscopy, bacteriological laboratory analysis of the discharge and scrapings to determine the bacteria type.

Differential diagnosis. Viral conjunctivitis, inclusion conjunctivitis, toxic and chemical conjunctivitis.

Treatment. At first, treatment and elimination of predisposing factors. Warm compresses with calendula or chamomile decoction for 5 minutes once to twice per day may help to soften the crusts. Topical antibiotics — Gentamicin, Tobramycin, Tetracycline, Erythromycin, and Ciprofloxacin in either ointment or drop form (the ointment form is more preferable). The use of lubricating drops may soothe the eyes, and are helpful if the eyes remain dry. If the cornea is inflamed, mild topical steroid treatment may be required.

Prognosis. With appropriate treatment and eye hygiene the outcome is excellent.

Complications are rare as the cornea is not affected otherwise corneal epithelial defects may lead to decrease of vision.

Prophylaxis. Proper hygiene, hand washing and timely adequate treatment of acute bacterial conjunctivitis or other predisposing and causative factors.

Gonococcal Conjunctivitis

Definition. Gonococcal conjunctivitis is a hyperacute severe purulent form of conjunctivitis associated with sexually transmitted infection — gonorrhea.

Etiology. Gonococcal conjunctivitis is caused by Neisseria gonorrhea, a gram-negative intracellular aerobic diplococcus.

Adults acquire this type of infection usually by spreading from the genitalia to the hands and then to the eyes (adult gonococcal conjunctivitis). Individuals in any age group are vulnerable to gonococcal conjunctivitis; but, sexually active individuals are at a high risk. Both male and female genders are equally affected.

Newborns obtain this type of conjunctivitis by passing through the birth canal of an infected mother (neonatal gonococcal conjunctivitis).

Clinical Picture

In case of *adult gonococcal conjunctivitis*, the incubation period lasts for 1—2 days. First signs of the disease: eyelids swell up, become firm so that it is impossible to open them. Discharge is serous bloody, like meat slops (the first stage — infiltration).

On the 4th—5th day the eyelids become softer, the amount of discharge increases, it becomes purulent. Yellow-green pus flows out from the eye in great amounts (the 2nd stage — suppuration). The conjunctiva is hyperemic, swollen, has a rough surface. On the 7th—10th day the edema of the conjunctiva diminishes gradually, papillae appear (the third stage — papillar hypertrophy). In 4—6 weeks the conjunctiva returns to the normal condition.

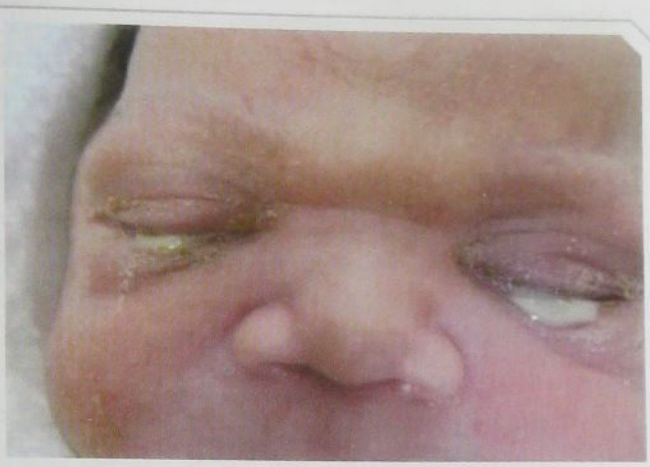


Fig. 7.3. Neonatal gonococcal conjunctivitis (from http://iahealth.net)

Neonatal gonococcal conjunctivitis starts in about 2 to 4 days after birth. It presents with purulent discharge, severe oedema and hardening of the eyelids; with just slight pressure on the lids or when forcing the lids to open purulent material may squirt out of the conjunctival sac. The infant is irritable and his/her conjunctiva is intensely inflamed, chemotic and red. Neonates may also acquire pharyngeal or respiratory infection (fig. 7.3).

Complaints. Acute itching, burning and redness of the eyes, eye pain, eyelid swelling, sensation of a foreign body, purulent discharge of green or yellow color with formation of crusts (usually in mornings on waking up); sticky eyelids which are difficult to keep open, blurred vision.

Signs. Severe purulent sticky discharge, eyelid edema, hemorrhagic conjunctivitis, marked chemosis. The following may also be present: conjunctival papillae, true membrane formation, superficial punctate keratitis and periauricular lymph nodes.

Methods of Examination. External examination, conjunctival scraping for culture testing.

Differential Diagnosis. Acute bacterial conjunctivitis of other etiology.

Treatment. Early diagnosis with prompt and rapid treatment is the key to prevention of permanent damage and blindness from acute gonococcal conjunctivitis.

Medical management of *adult gonococcal conjunctivitis* begins with an intramuscular injection of Ceftriaxone 2 times a day. Ideally, the patient should be hospitalized and given one gram of Ceftriaxone intravenously within 12 to 24 hours. Other antibiotics include orally-administered Tetracycline or Erythromycin four times daily. General treatment is conducted by a venereologist.

Ocular management starts with saline irrigation to clear and remove the mucopurulent discharge from the lids and conjunctiva. Topical Ofloxacin, Gentamicin, Ofloxacin or Ciprofloxacin is indicated even before the culture test re-

NOTE

A bandage on the affected eye in case of bacterial conjunctivitis is contraindicated.

sults are available. Dexpanthenol is appropriate if corneal infection occurs. Corticosteroids are not used, since they worsen the condition.

Neonatal gonococcal conjunctivitis requires frequent eye wash with saline solution to remove discharge and topical Sulfacetamide 20 %, Erythromycin ointments before the culture test is available. Once the diagnosis is confirmed systemic treatment with intravenous Penicillin (or Erythromycin) is vital.

Prognosis. Gonococcal conjunctivitis has a potential to cause blindness and even life-threatening illnesses, such as meningitis and sepsis, if the condition is not quickly diagnosed and properly managed. With early diagnosis and appropriate treatment, the outcome is good.

Complications. Severe complications may arise if the cornea is involved, or if adult gonococcal conjunctivitis is not treated. The complications include corneal ulcer, scar and perforation, symblepharon, loss of vision.

Prophylaxis.

Adult gonococcal conjunctivitis is highly infectious. Prevention the condition spreading consists in maintaining hygiene, washing hands regularly, avoiding sharing of pillows, towels, make-up, and limiting physical contact. To prevent aggravation of the condition a person should stay out of the sun, keep away from dust and smoke and avoid touching or rubbing the eyes.

Prevention of *neonatal gonococcal conjunctivitis* is wiping the eyelids with cotton wool moistened with 1 % solution of boric acid, instillation of Sulfacetamide 20 % in both eyes three times an hour or 0.5 % Erythromycin ointment once shortly after birth.

Diphtheritic Conjunctivitis

Definition. Diphtheritic conjunctivitis is a severe infectious conjunctival inflammation characterized by formation of a true membrane on the palpebral conjunctiva. This disease is quite rare in developed countries due to widespread vaccination against diphtheria but is endemic in many developing countries.

Etiology. The disease is typically caused by Corynebacterium diphtheria. Diphtheritic conjunctivitis mostly occurs in children at the age of two to eight years who are not immunized. It can also be seen in some immunized children, but in a less severe form.

Clinical Picture. The disease begins acutely. A patient has general symptoms that include high fever, weakness, headache. The preauricular lymph nodes are swollen and tender. Diphtheria of the conjunctiva is rarely developing as an isolated disease. More often it is combined with diphtheria of the nose, fauces, larynx.

The clinical picture of the disease can be divided into three stages:

1. *Stage of infiltration.* Acute edema of the eyelids, their skin is red and tense. The eyelids are firm, which makes it hard to open them for eye inspection. Discharge is serous bloody. The conjunctiva is red, infiltrated and covered with a thick grey-yellow membrane. The membrane is tough and firmly adherent to the conjunctiva, which, when removed, bleeds very much and leaves behind a raw area.

2. *Stage of suppuration.* Discharge becomes serous and purulent. Pain decreases and the eyelids become soft. The membrane falls off the conjunctiva leaving granulation.

3. Stage of cicatrisation. Scars in the form of stars appear at the place of granulation tissue, causing entropion, trichiasis and conjunctival xerosis. Commissures may be formed between the palpebral and bulbar conjunctiva — symblepharon. Since the process involves the cornea, it can lead to corneal ulcer.

Complaints. High fever, headache, pain in the eye, swollen, red and tender eyelids, discharge, photophobia.

Signs. The lids are swollen, tense and tender with impaired mobility, a grey-yellow membrane on the palpebral conjunctiva; serous or mucopurulent discharge; the preauricular nodes are enlarged and tender. General signs — fever, nasal and pharyngeal involvement.

Methods of Examination. Diagnosis is made on the basis of typical clinical features and confirmed by bacteriological examination.

Differential Diagnosis. Pseudomembranous conjunctivitis.

Treatment. When suspecting diphtheria, without waiting for the results of bacteriological analysis, a patient should be isolated and injected with antidiphtheric serum (6,000—10,000 AO) repeated in 12 hours, with topical application every other hour.

Locally there are prescribed irrigations with saline solutions, penicillin eye drops (1:10,000 units per ml) with an interval of every half an hour or other broad-spectrum antibiotics and vitamins. Tetracycline in the form of ointment at bed time. Atropine sulfate 1 % ointment should be added if the cornea is ulcerated.

Systemic antibiotic therapy (Penicillin, Erythromycin, Ampicillin, Tetracycline) intramuscularly twice a day for 10 days.

Prognosis in diphtheria is always serious both for the eye and the life of the patient.

Complications. Diphtheritic conjunctivitis can lead to corneal ulceration, conjunctival xerosis, symblepharon, ankyloblepharon, and entropion with trichiasis.

Prophylaxis. Proper and timely immunization in infancy and quick isolation of the infected patient are the usual preventive methods of diphtheria conjunctivitis.

Viral Conjunctivitis

The most common cause of infectious conjunctivitis, especially in older children, is a virus. Viral conjunctivitis is highly contagious, because airborne viruses can be spread through sneezing and coughing. Viral conjunctivitis can also accompany common viral upper respiratory infections such as measles, flu or common cold. Typically, the infection starts in one eye and quickly spreads to the other eye. There are many different serotypes but adenovirus infection is the most common cause of sporadic and epidemic viral conjunctivitis. The most common viruses that cause viral conjunctivitis are adenoviruses, enteroviruses, the measles virus and the herpes simplex virus.

Viral conjunctivitis usually causes a watery discharge, which can be crusty in the morning but is not pus-like; itching; tearing, conjunctival hyperemia, follicle and pseudomembrane formation; lasts from one to two weeks. There may also be cold-like symptoms, such as fever and a sore throat and lymphadenopathy.

There is no specific *treatment* for viral conjunctivitis. It must be fought off by the own body's immune system. However, the symptoms can be relieved with cool compresses and artificial tears. Topical antiviral medications may help as well. For the worst cases, topical corticosteroid drops may be prescribed to reduce discomfort from inflammation.

Adenoviral Conjunctivitis

Definition. Adenoviral conjunctivitis (Ad-CS) also known as "pink eye" is an acute highly contagious inflammation of the conjunctiva caused by adenovirus.

Etiology. Ad-CS is usually caused by adenoviruses of serotypes 3, 4, 7, 10. It can occur equally in men and women; affects kids and young adults. The virus is transmitted through direct contact with items used by the infected individual, by touch (hand-to-eye route), use of shared spaces (like swimming pools), and through respiratory, nasal droplets.

Clinical Picture. There are two major subtypes of adenoviral conjunctivitis:

— pharyngoconjunctival fever (PCF);

— epidemic keratoconjunctivitis (EKC).

The condition begins suddenly and unilaterally with eyelid swelling, pseudoptosis, severe conjunctival edema, hyperemia, marked foreign body sensation and watery discharge (fig. 7.4). Other signs include marked edema of the caruncle, follicle formation (fig. 7.5), conjunctival hemorrhages. Further there may be pseudomembrane formation and corneal lesions.

The fellow eye is generally involved in a few days, but the course is usually milder. The symptoms also include respiratory tract involvement and enlargement and tenderness of the preauricular nodes.

Complaints. Redness of the eye, itching, burning, foreign body sensation, extreme sensitivity to light, watery discharge, swelling of the eyelids, decreased vision, sore throat, runny nose.

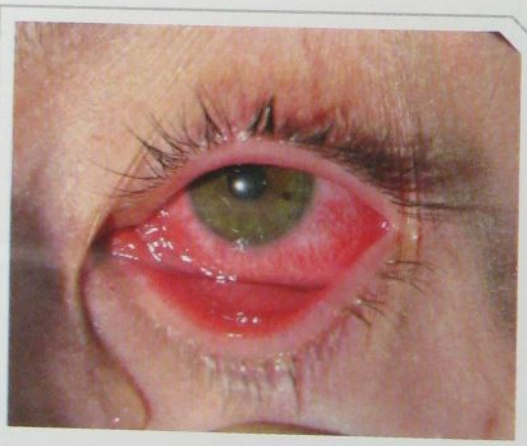


Fig. 7.4. Acute adenoviral conjunctivitis

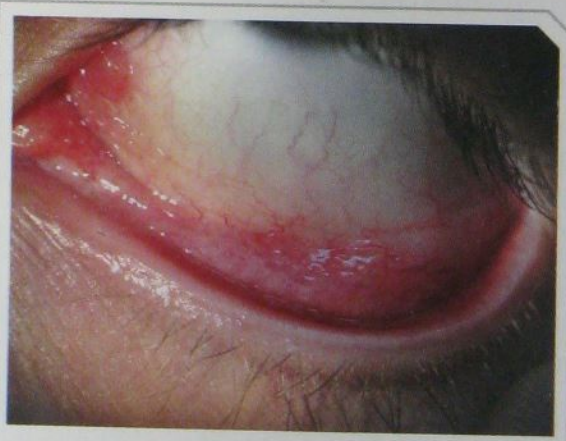


Fig. 7.5. Follicular conjunctivitis

Signs. Eyelid edema, pseudoptosis, follicular conjunctivitis, hyperemia, chemosis, subconjunctival hemorrhage, occasionally a pseudomembranous reaction, epiphora, palpable periauricular lymphadenopathy, respiratory infection.

Methods of Examination. External examination, slit lamp exam.

Differential Diagnosis. Chlamydial conjunctivitis, vernal keratoconjunctivitis, atopic keratoconjunctivitis, trachoma.

Treatment. Adenoviral conjunctivitis is a self-limited process and in most cases it spontaneously resolves. Its treatment is mostly supportive. Cold compresses and lubricants, such as artificial tears (Solcoseryl, Hypromellose, Carbomer) may be used to keep the eyes comfortable. Topical antiviral medications may help as well such as Interferon (drops 6—8 times a day; ointment 2—3 times a day). Topical vasoconstrictors and antihistamines may be used for severe itching but generally are not indicated. For the worst cases, topical corticosteroid drops may be prescribed to reduce discomfort from inflammation.

Prognosis. Common viral conjunctivitis usually resolves completely. Infections involving the cornea may leave a permanent scar that affects vision.

Complications. Complications from Ad-CS may arise if there are secondary conditions — corneal ulcer, scar, possibility of a bacterial infection occurring in addition to viral infection.

Prophylaxis. Adenoviral conjunctivitis is highly contagious. Prevention is possible with good personal hygiene, aseptic procedures, and consuming of immune response modifiers.

Chlamydial Conjunctivitis

Chlamydia is the most common sexually transmitted bacterial disease in the world; it can manifest in the eye in the form of inclusion conjunctivitis and trachoma, although each involves a different subtype of the bacteria. Inclusion conjunctivitis typically occurs in developed countries; in contrast, trachoma is primarily restricted to impoverished regions. While involving different serotypes, both diseases are caused by the same species of Chlamydia — Chlamydia trachomatis.

Paratrachoma

Definition. Paratrachoma or chlamydial inclusion conjunctivitis is an infectious conjunctivitis caused by sexually transmitted Chlamydia trachomatis.

Etiology. Paratrachoma is caused by a strain of Chlamydia trachomatis (serotype D—K). It is also regarded as inclusion conjunctivitis because of the inclusion bodies found in the epithelial cells.

Adult inclusion conjunctivitis results from poor personal hygiene and transmission of contaminated genital secretions to the eye, either via autoinoculation or from a sexual partner. Rarely, adult inclusion conjunctivitis is acquired from contaminated, incompletely chlorinated swimming pool water.

Neonatal inclusion conjunctivitis is acquired when the child passes through the birth canal of an infected mother.

Clinical Picture. In newborns, inclusion conjunctivitis appears between the 2nd and 25th day of life, reaching a maximum intensity in 2 weeks, with swelling of the lids, massive purulent exudation that can be mistaken for gonorrhea. Inclusion conjunctivitis is however benign and unilateral.

In adults after an incubation period of 4 to 12 days, most patients present with unilateral follicular conjunctivitis with minimal suppuration, swollen lymph nodes in front of the ears and conjunctival lymphoid follicles.

Complaints. Itching and redness of the eye, tearing, lid edema, mild eye pain, foreign body sensation, sensitivity to light, blurred vision, pus or watery discharge.

Signs. In case of adult inclusion conjunctivitis — conjunctival injection, follicles on the tarsal upper and lower conjunctiva and fornix, papillary hyperplasia, chemosis, superior micropannus, eyelid edema, mucopurulent or mucus discharge, tender periauricular lymphadenopathy.

In case of neonatal inclusion conjunctivitis — red and inflamed eye, tearing, purulent discharge, swollen eyelids. Follicles are not present; the lymph nodes are not enlarged since the lymphatic system of the newborn is not completely developed.

Methods of Examination. External eye examination, cytological laboratory investigation with staining, consultation of a gynecologist, urologist.

Differential Diagnosis. Chronic follicular conjunctivitis, adenoviral keratoconjunctivitis, allergic conjunctivitis, molluscum contagiosum.

Treatment. In adults, the disorder is treated with Tetracycline or Erythromycin eyedrops or ointment over a period of four to six weeks. The oculogenital mode of infection entails a risk of reinfection. Therefore, patients and sexual partners of treated patients also require treatment with oral Tetracycline, Erythromycin, Amoxicillin.

Children should be treated with oral Erythromycin (50 mg/kg/day in 4 divided doses) for 14 days and topically with Erythromycin ophthalmic ointment for one week. Before applying the ointment the eyes have to be irrigated with normal saline to remove mucopurulent discharge.

Prognosis. With early diagnosis and appropriate treatment, the outcome is good.

Complications may arise if the cornea is involved, or if the pathology is not treated properly. It is also dependent upon the presence of secondary conditions. The complications include: corneal ulcer or scar, entropion, loss of vision. 10—20 % of infected newborns may develop pneumonia during the first six months of life.

Prophylaxis. Patients should follow safe sexual practices. Prevention consists in maintaining hygiene, washing hands regularly, avoiding sharing of pillows, towels, make-up and regular chlorination of swimming pools. The only prevention of neonatal inclusion conjunctivitis is prenatal treatment of pregnant women.

Trachoma

Definition. Trachoma is contagious chronic infectious conjunctivitis caused by the bacterium Chlamydia trachomatis.

Currently, trachoma (from ancient Greek "rough eye") is the leading cause of preventable blindness in the world. Approximately 1.2 million people in the world are

blind because of trachoma. It is endemic in parts of Africa, Asia, the Middle East, Latin America, the Pacific Islands, and aboriginal communities in Australia. Worldwide, an estimated 229 million people in 53 countries live in trachoma-endemic areas.

Etiology. Trachoma is caused by a strain of Chlamydia trachomatis (serotype A—C). It is highly contagious in the early conjunctivitis stage and is spread by direct contact with affected individuals, or contact with their towels and/or washcloths. Insect vectors, especially flies and gnats, play a role in this transmission too.

Clinical Picture. The bacteria Chlamydia trachomatis has an incubation period of 5 to 12 days, after which the affected individual experiences symptoms of conjunctivitis. Left untreated, the disorder progresses through four stages according to its clinical course. Suspicion of trachoma and pre-trachoma is also considered.

Suspicious of trachoma (trachoma dubium — TrD): a diagnosis is made usually at mass prophylactic examinations in those cases, when there are no clear clinical and laboratory data.

Pretrachoma (PrTr): slight hyperemia and insignificant infiltration of the conjunctiva, follicles are not present.

Stage I (stage of conjunctivitis): bilateral, usually subacute conjunctivitis with violaceous discoloration, opacification and loosening of the tissues, lymphoid hyperplasia in the upper tarsus with immature follicles.

Stage II (stage of follicle formation): development of subepithelial mature follicles (trachoma granules).

Stage III (stage of scarring): the follicles burst and heal with scarring and shrinkage of the tarsal conjunctiva.

Stage IV (stage of sequelae and complications): a linear scar on the tarsal conjunctiva, shortening of the fornix, boat-shaped distortion of the tarsus, trachomatous ptosis. The progression is entropion, trichiasis, keratitis, symblepharon, corneal ulceration, perforation, and finally loss of the eye.

Complaints. Irritation and mild itching, discharge from the eyes contains pus, tearing, eyelid swelling, blurred vision, photophobia, pain in the eye.

Signs of trachoma depend on its stage — conjunctival hyperemia, mucopurulent discharge, follicles and papillae, scarring of the conjunctiva, neovascularization, trichiasis, corneal ulcers or opacity.

Methods of Examination. External eye examination with eversion of the upper and lower eyelid, eye swab for laboratory testing.

Differential Diagnosis. Allergic conjunctivitis, epidemic keratoconjunctivitis, pharyngoconjunctival fever, trichiasis.

Treatment. The key to the treatment of trachoma is the SAFE strategy developed by the World Health Organization (WHO) — **S**urgery for treating the trachoma complication; **A**ntibiotics for active disease; **F**acial and hands hygiene; and improvement of **E**nvironmental conditions.

Trachoma may be effectively treated in the early stages with topical and systemic antibiotics (Tetracycline, Erythromycin, Tobramycin, Ciprofloxacin, Ofloxacin) 4—5 times a day for 2—3 months. Topical antibiotics can be administered in the form of

eye drops or ointments. Ointments have the advantage of prolonged contact with the ocular surface and an accompanying soothing effect. Besides, preparations of sulfonamides (Sulfacetamide) are required too.

Eyelid follicles may be removed surgically. When lid scarring is established surgery is required to correct entropion and trichiasis.

If the cornea has become clouded, corneal transplantation is an option that offers some hope of improved vision; however, the results are not good.

Prognosis. Appropriate treatment of the early stage of the disease gives an excellent prognosis. Severe stages may be stabilized but vision may not be improved. Re-infection worsens the prognosis, blindness is possible.

Complications. One episode of trachoma is easily treated with early detection and use of antibiotics. Repeated or secondary infections can lead to complications, including: scarring of the inner eyelid; eyelid deformities, such as an inward folding eyelid (entropion) or ingrown eyelashes (trichiasis); corneal scarring or opacity; partial or complete vision loss.

Prophylaxis. Environmental sanitation and improved personal hygiene are the keys in reducing the spread of trachoma — hand and face washing, controlling flies, proper waste management, improved access to water.

NOTE

Infectious conjunctivitis must be prevented or it can turn into epidemic. The following measures of preventions must be taken:

— Wash your hands often.
— Avoid touching or rubbing the eyes.
— Wash discharge from around the eyes using a fresh cotton ball or tissue.
— Discard used make-up, contact lens solution, and used disposable contact lenses.
— Do not share eye cosmetics or personal eye care items.
— Wash bed linen and towels in hot water and detergent.
— Avoid swimming in a public swimming pool.

Non-infectious Conjunctivitis

This group includes all types of noninfectious inflammatory conjunctivitis caused by mechanical, physical, chemical, or allergic (atopic) factors.

Allergic Conjunctivitis

Allergic conjunctivitis is a common condition responsible for 15 % of all eye-related problems. It occurs when the eye comes into contact with an allergen. An allergen is a particular substance that makes the body's immune system react abnormally, causing irritation and inflammation in the conjunctiva. This is known as an allergic reaction. Common examples of allergens include pollen, dust mites, animal fur or cosmetics. Certain forms of pink eye can be caused by the eye's immune responses,

such as to wearing contact lenses. A reaction to preservatives in eye drops or ointments can cause toxic conjunctivitis.

NOTE

If itching is not present, allergic conjunctivitis is much less likely.

Allergic conjunctivitis usually affects both eyes at the same time. The symptoms of allergic conjunctivitis may appear very suddenly, often immediately after coming into contact with an allergen. In other cases, symptoms will develop 24—48 hours after you have come into contact with the allergen.

Allergic conjunctivitis often causes severe itching, reddening of the eye, irritation, tearing, foreign body and burning sensation. Symptoms can also include watery discharge from the eye, swelling of the eyelid, and edema of the conjunctiva.

People who have allergic conjunctivitis associated with hay fever almost always have other features of allergy, such as sneezing, a runny, itchy nose and itchiness at the back of the throat.

The most important treatment for allergic conjunctivitis is to avoid the substance that causes the allergy. This may be easy if the cause is animal fur, contact lenses or cosmetics, but more difficult if it is pollen. However, keeping windows and doors closed on days when the pollen count is very high can often help reduce symptoms. In mild cases, cool compresses and artificial tears relieve discomfort. In more severe cases, anti-inflammatory medications and antihistamines may be prescribed.

NOTE

Allergy of the eyes cannot be cured, but can be treated.

Allergy can run in families.

Most young people grow out of eye allergies.

There are five main subtypes of allergic conjunctivitis:

- *Seasonal allergic conjunctivitis (SAC)* is the most common form of allergic conjunctivitis and it is found in adults and children. The symptoms are recurrent due to seasonal exposure.
- *Perennial allergic conjunctivitis (PAC)* or year-round conjunctivitis — symptoms occur all year round, and are usually present when you wake up in the morning. The symptoms can be caused by a variety of allergens, such as dust mites, animal fur, mold or other environmental factors that may be present throughout the year.
- *Vernal keratoconjunctivitis (VKC)* is the most common type of conjunctivitis accounting for half of all allergic conjunctivitis cases. Symptoms recur at the same time every year in the spring and summer months when grass, trees, and flowers are in pollen.
- *Atopic keratoconjunctivitis (AKC)* is always associated with inflammation of the skin around the eye, and usually occurs in those using eye drops. This condition causes the skin on your eyelids to become red, cracked, sore and inflamed (dermatitis).
- *Giant papillary conjunctivitis (GPC)* most commonly occurs in people using soft contact lenses. It may also develop following eye surgery.

Vernal Keratoconjunctivitis

Definition. Vernal keratoconjunctivitis (VKC) or spring catarrh is a chronic, recurrent, bilateral conjunctival inflammation that occurs seasonally during the spring and summer months.

Etiology. VKC is caused by an allergic reaction to some exogenous allergens such as grass or trees pollens. It often occurs in people with a strong family history of allergies. These may include allergic rhinitis, asthma, and eczema. It is most common in children and young adults and rarely may persist beyond the age of 25 years. It is more common in males than in females.

Clinical Picture. After contact with an allergen, the eyes start itching intensely and become sensitive to light. Usually both eyes are affected, and cobblestone-like changes appear in the upper palpebral conjunctiva. In other cases, a gelatinous nodule may develop in the limbus (fig. 7.6). In very severe cases, the corneas may scar or clouding of the lens (cataract) may occur leading to reduced vision. The disease may follow a cold or sore throat.

Complaints. Severe burning, itching, tearing, redness of the eye, watery discharge, foreign body sensation, photophobia, blepharospasm and heaviness of the lids.

Signs. Conjunctival injection, giant "cobblestone" papillae on the upper tarsal conjunctiva, gelatinous nodules on the limbus, keratitis.

Methods of Examination. The diagnosis is usually clinical. Eosinophils are present in conjunctival scrapings, which may be taken from the lower or upper tarsal conjunctiva; however, such testing is rarely indicated.

Differential Diagnosis. Atopic keratoconjunctivitis, giant papillary conjunctivitis, viral conjunctivitis, trachoma.

Treatment. Cool compresses and artificial tears such as Dexpanthenol, Carbomer, sometimes relieve discomfort. Artificial tears provide comfort, but they also protect the eye's surface from allergens and dilute the allergens that are present in the tear film.

In more severe cases, anti-inflammatory medications (Diclofenac, Indometacine) and antihistamines may be prescribed. Oral antihistamines that are commonly prescribed include Diazoline, Suprastin, Claritin, Loratadine. Topical antihistamines are Cromoglicate, Lodoxamide, Emedastine Olopatadine. Some patients with persistent allergic conjunctivitis may also require topical steroid drops.

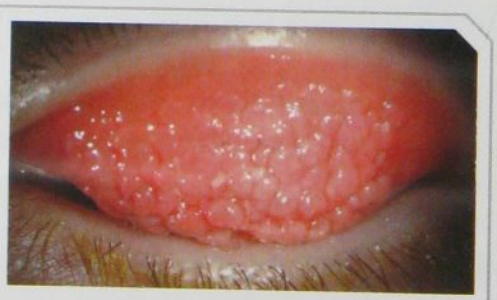


Fig. 7.6. Vernal keratoconjunctivitis with cobblestone-like giant papillae on the upper tarsus (from http://www.eyerounds.org)

Prognosis. Vernal keratoconjunctivitis without proper diagnosis and quick treatment could potentially cause eye damage and loss of vision due to corneal complications. Mild cases of VKC are known to resolve spontaneously. Young children usually outgrow the condition after the onset of puberty.

Complications may arise if VKC is not treated or from unsupervised used of topical corticosteroids. The complications could include corneal damage, scar, and perforation that can lead to loss of vision or secondary bacterial infection.

Prophylaxis. Vernal keratoconjunctivitis is a spontaneous body (eye) response to the presence of an allergen; it cannot be prevented, but only managed. In case of a severe incidence of the condition relocation to colder climates is beneficial, but it may not always be practicable.

Atopic Keratoconjunctivitis

Definition. Atopic keratoconjunctivitis (AKC) is a chronic bilateral allergic ocular disease that occurs most often in patients with a history of atopic dermatitis.

Etiology. AKC is strongly associated with atopic dermatitis, which affects about 3 % of the population. Of these people, it is estimated that 15—25 % have some form of ocular involvement, which is usually AKC. It is more prevalent in men than in women aged 30 to 50 years.

Clinical Picture. The clinical picture mostly presents throughout the year but may worsen during certain seasons, especially in hot, cold seasons. It may affect the eyelid skin and lid margin, conjunctiva, cornea, and lens.

Complaints. Itching of the eyelids that are red and swollen, redness of the eye, burning, mucous discharge, sensitivity to light, blurred vision, dry eyes.

Signs. Eyelid edema and eczema, chronic blepharitis and conjunctivitis, conjunctival injection, chemosis, micropapillae of the tarsal conjunctiva — primarily of the lower eyelid, corneal erosion, cataract, atopic dermatitis.

Methods of Examination. Family history, atopic dermatitis history, clinical investigation — slit lamp and ophthalmoscope exam, blood analysis, scraping of the conjunctival tissue to measure serum IgE level, eosinophil count.

Differential Diagnosis. Vernal keratoconjunctivitis, perennial allergic conjunctivitis, giant papillary conjunctivitis, toxic conjunctivitis (table 7.1).

Treatment of AKC must be co-managed with an allergist or dermatologist to provide guidance and follow-up of systemic therapy.

In early disease, conservative management with combination topical mast cell stabilizers and antihistamine drops, lubricants, and oral antihistamines may keep the ocular condition under control. Application of cold compress can help reduce discomfort.

In more advanced cases, additional topical corticosteroids, cyclosporin, anti-inflammatory eye-drops, and topical ointments are prescribed. When topical treatments fail to induce remission, oral prednisone and cyclosporine are the next treatment options.

Prognosis. If AKC is left untreated, it may result in ulcers and scars forming on the cornea, which may lead to vision loss in the most serious cases.

Complications. Unless treated in time, atopic keratoconjunctivitis can persist for many years and cause severe complications — corneal ulceration, scarring, keratoconus, corneal vascularization and steroid-induced glaucoma.

Table 7.1

Major Differentiating Factors between Vernal Keratoconjunctivitis (VKC) and Atopic Keratoconjunctivitis (AKC)

Characteristics	VKC	AKC
Age at onset	Generally presents in the first decade	Generally presents in the second to third decade
Sex	Males are affected preferentially	Males are affected preferentially
Seasonal variation	Typically occurs during spring months	Generally perennial
Discharge	Thick mucoid discharge	Watery and clear discharge
Eyelid skin involvement	The eyelids are not involved	The eyelids are thickened, red and have eczema signs
Conjunctival scarring	Moderate incidence of conjunctival scarring	Higher incidence of conjunctival scarring
Corneal neovascularization	Not present unless secondary to infectious keratitis	Deep corneal neovascularization tends to develop
Presence of eosinophils in conjunctival scraping	Conjunctival scraping reveals eosinophils to a greater degree in VKC than in AKC	Presence of eosinophils is less likely

Prophylaxis. AKC cannot be prevented, but only managed. In order to preserve the eye condition, the patient should protect the eyes from airborne particles by wearing glasses, maintain hygiene, avoid touching or rubbing the eyes and prevent aggravation of the condition by staying out of the sun, keeping away from dust and smoke.

Giant Papillary Conjunctivitis

Definition. Giant papillary conjunctivitis (GPC) is a conjunctival inflammation characterized by large papillae that form in response to chronic presence of a foreign body in the eye.

Etiology. GPC is most often caused by soft contact lenses, so this condition is also called contact lens-induced papillary conjunctivitis (CLPC). GPC can occur with any type of lens, ocular prosthesis, or even exposed surgical sutures. The rate of GPC in contact lens wearers is about 21 %. Several factors contribute to GPC — mechanic irritation, allergic reaction to a particular material or a contact lens solution, improper contact lens care and hygiene.

Clinical Picture. The condition begins with hyperemia of the tarsal conjunctiva followed by small bumps that arise and grow larger. Patients often find their lenses going upward as they blink. In acute stage papillae are up to 1 mm in diameter, their number, elevation and vascularization increase, which makes tarsal conjunctiva look like cobblestones. Redness, swelling, and mucus production become more severe; and some changes in the cornea may be present.

Complaints. Itchy eyes, irritation, redness, burning, mucous discharge, light sensitivity, foreign body sensation upon removal of contact lenses, swollen eyelids and contact lens intolerance.

Signs. Conjunctival hyperemia, the upper tarsal conjunctiva is irritated and swollen with giant papillae that give a "cobblestone" appearance, mucous discharge.

Methods of Examination. External examination, slit lamp exam with upper eyelid eversion.

Differential Diagnosis. Vernal keratoconjunctivitis, atopic keratoconjunctivitis, chlamydial infection, blepharitis (table 7.2).

Treatment. In most cases, the first step of GPC treatment involves removal of the irritant — discontinuance of the use of contact lenses or removal of surgical stiches.

Cool compresses and artificial tears sometimes relieve discomfort in mild cases. In more severe cases, mass cell stabilizers, nonsteroidal anti-inflammatory drugs (NSAIDs) and antihistamines may be prescribed. Cases of persistent giant papillary conjunctivitis may also require topical corticosteroids.

Prognosis for GPC is generally favorable. With effective treatment, it takes anywhere from 1—6 weeks to resolving completely.

Complications. When left untreated, GPC may cause corneal damage or scarring, bacterial or viral infections that can occur superimposed. GPC can also lead to ptosis.

Prophylaxis. Appropriate contact lens hygiene is an important component in the disease prevention.

Table 7.2

Differential Diagnosis of Conjunctivitis

	Bacterial	Viral	Chlamydial	Allergic
Eye involvement	Unilateral but the other eye becomes involved	Unilateral but the other eye becomes involved	Unilateral but the other eye becomes involved	Bilateral
Type of discharge	Purulent (yellow-green)	Watery (clear)	Mucopurulent	Mucoid (white)
Type of conjunctival reaction	Papillar	Follicular	Follicular and papillar	Papillar
Itching	Mild	Mild	No, or mild	Severe
Chemosis	Severe	Mild	Mild	Mild to severe
Eyelid oedema	Moderate	Moderate	Moderate	Severe
Pannus formation	No	No	Yes	No
Photophobia	Possible	Yes	Yes	Severe
Preauricular adenopathy	Rare	Present	Present	No
Fever	Occasional	Moderate	No	No
Contagious	Yes	Yes	Yes	No

4.2. Degeneration of the Conjunctiva

Pinguecula

Definition. Pinguecula is a yellowish, slightly raised thickening of the conjunctiva on the sclera near the limbus, often at the 3 o'clock or 9 o'clock position from the cornea (fig. 7.7). Sometimes, more than one pinguecula may present in each eye.

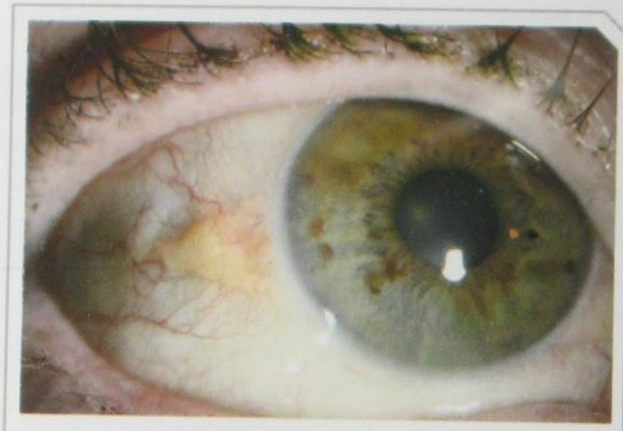


Fig. 7.7. Pinguecula (from http://www.eyerounds.org)

Etiology. Pinguecula usually results from excessive exposure to sun or constant eye irritation from dust, wind or other irritants, causes a change in the normal tissue around the exposed area of the conjunctiva. This condition is more commonly found in adults and older people.

Clinical Picture. Pinguecula results from calcium, fat or protein deposits and may grow larger over the years. It is generally asymptomatic but due to aggravation from dust, wind, dry condition, prolonged exposure to sun light it may become inflamed and cause eye irritation.

Complaints. Some people may have no complaints other than the mass appearing. Others may notice mild burning, itching, foreign body sensation, dryness of the eye, sometimes redness.

Signs. A small yellowish bump on the conjunctiva near the cornea.

Methods of examination. External examination, slit lamp exam.

Differential diagnosis. Pterygium.

Treatment. Usually no treatment is needed. Keeping the eye moist with artificial tears may be helpful in eye lubricating and relieving foreign body sensation.

Pinguecula can lead to localized inflammation and swelling that is sometimes treated with steroid eye drops or non-steroidal anti-inflammatory drugs (NSAIDs).

Pinguecula does not need to be removed as it does not affect vision. Surgical removal of a pinguecula may be considered if it becomes especially uncomfortable, if it interferes with contact lens wear or blinking, or for cosmetic reasons.

Prognosis. The condition is not cancerous and the prognosis is good.

Complications. It may enlarge over the years and develop into a pterygium, which grows onto the cornea and affects vision.

Prophylaxis. Prevention of pinguecula is keeping the eye lubricated, wearing sunglasses and avoiding eye irritants.

Pterygium

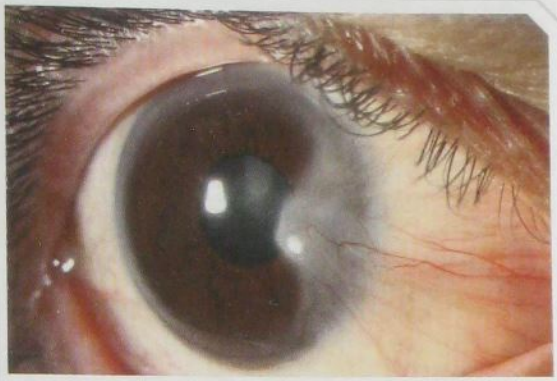


Fig. 7.8. Pterygium (from http://www.eyerounds.org)

Definition. Pterygium is a pinkish triangular or wing-shaped growth of the conjunctiva that grows from the medial portion of the palpebral fissure and extends over the cornea (fig. 7.8).

Etiology. The primary cause of pterygium growth is exposure to the sun; other causes are environmental factors such as wind and dust. Pterygium usually occurs in people aged 20 to 50, and is more common in men.

Clinical Picture. At the beginning a pterygium is a painless area of raised conjunctiva that has blood vessels on the inner or outer edges of the cornea. Having become inflamed it causes burning, irritation, foreign body sensation. If growth extends far enough onto the cornea, vision may be affected.

Complaints. Early pterygium is asymptomatic; as the condition progresses it causes irritation, dryness, and foreign body sensation. If it extends onto the cornea, vision is blurred as the curvature of the cornea is altered.

Signs. A pink fleshy growth on the conjunctiva that is triangular and vascular and spreads to the corneal limbus or even on the cornea.

Methods of examination. External examination, slit lamp exam.

Differential diagnosis. Pinguecula, squamous cell carcinoma.

EYE FACTS

Because pterygium often affects surfers, it is also known as *surfer's eye*.

Treatment depends on the size of the pterygium, whether it is growing, and the symptoms it causes. If pterygium is small, the condition is mild and inflammation occurs, lubricants and mild steroid eye drops may be prescribed. Once pterygium starts to block vision, a surgery may be necessary.

Prognosis. Most often pterygia cause no problems and do not need treatment. If a pterygium affects the cornea, its removal can have good results.

Complications of pterygium are reduction of central vision, astigmatism, corneal scarring.

Prophylaxis. Prevention of pterygium is wearing sunglasses, staying out of windy, dusty and polluted environment or wearing protective glasses as well as keeping eyes lubricated and moist.

4.3. Tumors of the Conjunctiva

Conjunctival tumors are often diagnosed in childhood. They are divided into benign tumors (99 %) and malignant tumors (1 %). Benign lesions may become malignant; for example, a nevus may develop into a malignant melanoma.

Benign tumors

Nevus (birthmark) is a congenital, benign, pigmented, slightly elevated conjunctival growth (fig. 7.9). Most commonly nevus is present at birth and is usually located near the limbus in the temporal part of the palpebral fissure. Conjunctival nevus may increase in size as a patient grows older. They have a low rate of malignancy in terms of conversion to conjunctival melanomas.

Conjunctival nevus only requires periodic clinical observation (with photographs if possible). If there is any concern about tumor growth or change in pigmentation, excisional biopsy with histopathologic analysis should be performed.

Treatment is surgical: complete excision is indicated when a nevus has enlarged to such a degree as to irritate the eye.

Dermoid is a congenital round elevated whitish or pale yellowish solid mass lesion of the conjunctiva. It is mostly located on the limbus involving the conjunctiva and cornea. It may have hair follicles, sebaceous and sweat glands.

Conjunctival dermoid may be present as an isolated lesion or associated with other ocular and systemic pathology as oculo-auriculo-vertebral dysplasia (Goldenhar's syndrome), coloboma of the upper lid, and lacrimal stenosis.

Treatment of dermoid is surgical: for cosmetic reasons or if the mass encroaches towards the central cornea resulting in astigmatism and visual impairment. Surgical excision should remain strictly superficial as complete excision may result in perforation of the eyeball.

Papilloma is a benign epithelial growth on the bulbar or tarsal conjunctiva. Conjunctival papilloma is mostly caused by human papillomavirus, has the form of multiple nodules and is usually combined with the presence of papilloma in other parts of body. It is characterized by a smooth surface, red-pink color on a wide vascular pedicle. Papilloma is benign and does not have a malignant growth.

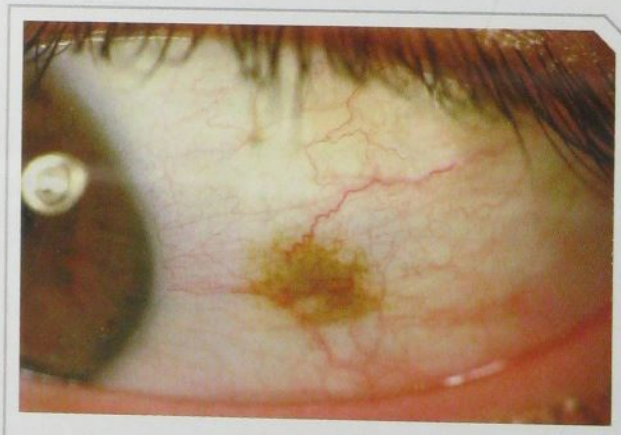


Fig. 7.9. Nevus of the conjunctiva

Depending on the size and location, these lesions can be irritating, cause tearing or blurring of vision, or simply be cosmetically unappealing.

No treatment recommended for asymptomatic lesions because of the high rate of spontaneous resolution. Treatment for recurrent or symptomatic lesions includes surgical excision, cryotherapy, topical mitomycin C or interferon.

Malignant Tumors

Carcinoma is a malignant tumor arising from the squamous epithelium and appears as a gelatinous whitish or yellow-pink nodule on the eye surface near the limbus. This tumor can extend onto the cornea, around the limbus, and rarely into the eye and orbit.

Conjunctival carcinoma is usually found in older Caucasian patients. It also occurs more often in men than in women. Some believe that excessive exposure to sunlight from outdoor activities like sunbathing, golfing, fishing, and other sports can lead to this tumor.

The treatment for squamous cell carcinoma of the conjunctiva is complete surgical removal of the tumor, cryotherapy (freezing), radiation therapy, and more recently, chemotherapy eye-drops.

Lymphoma is an ocular surface tumor that usually appears as a painless, salmon-pink, "fleshy" conjunctival thickening and often occurs in the inferior fornix. Conjunctival lymphoma shows a female predominance and occurs most frequently in the fifth to seventh decade of life.

Conjunctival lymphomas can become large enough to displace the eyeball, and restrict eye movement. Eye movement restriction can cause diplopia. Rarely, and if large enough, orbital lymphoma can press on the optic nerve and cause loss of vision.

Depending on the size of the tumor, treatment can include complete excision, cryotherapy, administration of chemotherapy medicines, radiation, or treatment by intravenous chemotherapy. Sometimes patients take a special antibiotic to prevent long-term involvement elsewhere in the body.

Kaposi's sarcoma is a malignant vascular tumor in the conjunctiva that appears as light to dark red nodular mass, commonly seen in the fornix. It is typically found in patients with acquired immunodeficiency syndrome (AIDS), but can occur in patients with immunosuppression, organ transplantation or human herpes simplex virus-8 (HHV-8) infection.

Treatment depends on the patient's age and the status of general health, current medications and immune status. It may be treated by cryotherapy, surgical excision (if the lesion is small), radiotherapy and/or chemotherapy.

Review:

1. Key points

Diseases of the conjunctiva according to the origin may be *congenital or acquired*, by clinical presentation — *mild, moderate* or *severe*, by the onset of the

pathological process — *acute, subacute* or *chronic*, by pathogenesis — *degenerative, inflammatory, tumors*. The main symptoms of conjunctival diseases are redness of the eye, itching, burning, foreign body sensation, photophobia, discharge. The examination consists of external inspection of the conjunctiva with eversion of the upper and lower eyelids, slit lamp examination, bacteriological laboratory investigation of the discharge.

Inflammatory diseases (conjunctivitis) may be bacterial, viral, chlamydial.

Bacterial conjunctivitis is inflammation of the conjunctiva as a result of bacterial infection. Bacterial infections cause a red eye, which is associated with a grey or yellowish pus or mucopurulent discharge from the eye that may cause the lids to stick together, especially after sleeping. Treatment of bacterial conjunctivitis includes irrigation of the eye with normal saline to remove the purulent discharge and the use of topical and systemic antibiotics.

Viral conjunctivitis is inflammation of the conjunctiva due to viruses and is highly contagious. Viral conjunctivitis usually causes a watery discharge, which can be crusty in the morning but is not pus-like; itching; tearing, conjunctival hyperemia, follicle and pseudomembrane formation. There may also be cold-like symptoms. There is no specific treatment for viral conjunctivitis, it is mostly supportive. Cold compresses, lubricants, topical antiviral medications and corticosteroid drops.

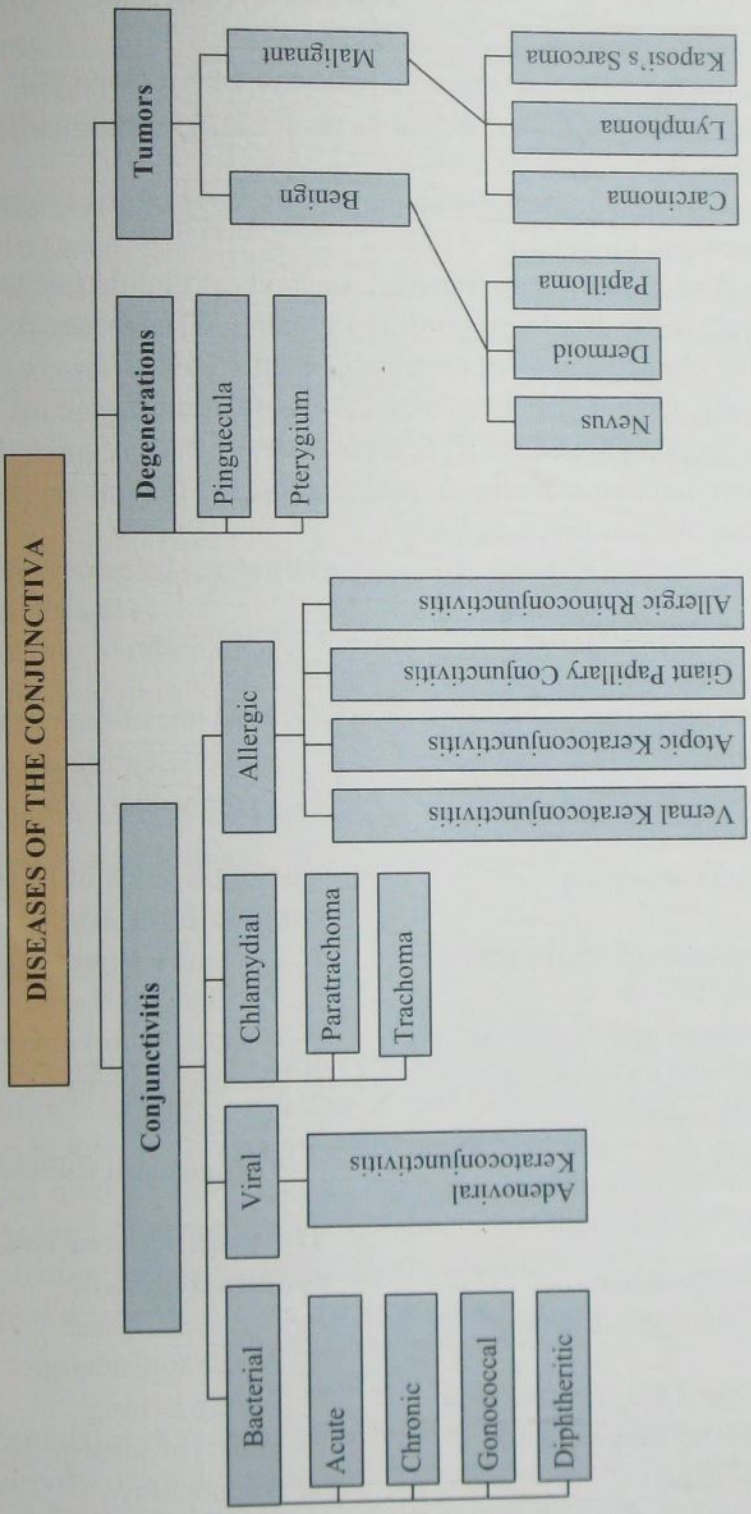
Chlamydial conjunctivitis is a sexually transmitted bacterial disease that can manifest in the eye in the form of inclusion conjunctivitis and trachoma that is the leading cause of preventable blindness in the world. The signs of trachoma depend on its stage — conjunctival hyperemia, mucopurulent discharge, follicles and papillae, scarring of the conjunctiva, neovascularization, trichiasis, corneal ulcers or opacity. The key to the treatment of trachoma is the SAFE strategy developed by the World Health Organization (WHO) — Surgery for treating trachoma complication; Antibiotics for active disease; Facial and hands hygiene and improvement of Environmental conditions.

Allergic conjunctivitis is a type of *non-infectious* inflammatory conjunctivitis. The symptoms of allergic conjunctivitis may appear very suddenly and causes severe itching, reddening of the eye, irritation, tearing, foreign-body and burning sensation. Symptoms can also include watery discharge from the eye, swelling of the eyelid, and edema of the conjunctiva. The most important treatment for allergic conjunctivitis is to avoid the substance that causes the allergy. In mild cases, cool compresses and artificial tears relieve discomfort. In more severe cases, anti-inflammatory medications and antihistamines may be prescribed.

Degenerations of the conjunctiva are pinguecula and pterygium. The symptom is the growth of the conjunctiva; treatment depends on its size and location. If the condition affects vision, surgical removal may be necessary.

Tumors: benign — nevus, dermoid, papilloma; malignant — carcinoma, lymphoma, Kaposi's sarcoma. The symptom is appearance of neoplasms of different form, size and color. Treatment is surgical, cryotherapy, radiation therapy, chemotherapy.

2. Diagrams


DISEASES OF THE CONJUNCTIVA
Conjunctivitis
Bacterial
Acute
Chronic
Gonococcal
Diphtheritic
Viral
Adenoviral Keratoconjunctivitis
Chlamydial
Paratrachoma
Trachoma
Allergic
Vernal Keratoconjunctivitis
Atopic Keratoconjunctivitis
Giant Papillary Conjunctivitis
Allergic Rhinoconjunctivitis
Degenerations
Pinguecula
Pterygium
Tumors
Benign
Nevus
Dermoid
Papilloma
Malignant
Carcinoma
Lymphoma
Kaposi's Sarcoma

3. The Review Questions

A. Control Questions

1. What are the main clinical signs which have to be considered in the differential diagnosis of conjunctival inflammation?
2. What are the main types of conjunctivitis and their pathogens?
3. What is bacterial conjunctivitis and its main signs, symptoms and principles of treatment?
4. Adult and neonatal gonococcal conjunctivitis, its etiology, symptoms, methods of treatment and prevention.
5. What are the symptoms and treatment methods of diphtheritic conjunctivitis?
6. Adenoviral conjunctivitis, its clinical presentation and treatment.
7. What is the clinical course of chlamydial conjunctivitis?
8. What are the general principles of trachoma treatment?
9. What are the complications and consequences of trachoma?
10. Allergic conjunctivitis, it clinical presentation and treatment.

B. Tests

1. **Symptoms of conjunctivitis are:**
   A. Redness of the eye
   B. Itching and burning
   C. Photophobia
   D. Tearing
   E. Discharge
   F. Foreign body sensation

2. **The main methods of conjunctiva diagnostics are:**
   A. External examination with eversion of the upper and lower eyelids
   B. Slit lamp examination
   C. Ophthalmoscopy
   D. Staining
   E. Perimetry
   F. Bacteriological laboratory investigation of the discharge

3. **The main clinical diagnostic signs in the differential diagnosis of conjunctivitis are:**
   A. Conjunctival injection
   B. Conjunctival response
   C. Discharge
   D. Edema
   E. Presence of membranes or pseudomembranes
   F. Lymphadenopathy

4. **The main signs of bacterial conjunctivitis are:**
   A. Unilateral onset
   B. Purulent discharge
   C. Corneal involvement
   D. Watery discharge
   E. Adenopathy
   F. Conjunctival follicles

5. **The main signs of viral conjunctivitis are:**
   A. Unilateral onset
   B. Purulent discharge
   C. Severe itching
   D. Watery discharge
   E. Adenopathy
   F. Conjunctival follicles

6. **The main signs of chlamydial conjunctivitis are:**
   A. Unilateral onset
   B. Mucopurulent discharge
   C. Pannus
   D. Watery discharge
   E. Corneal involvement
   F. Conjunctival follicles

7. **The main signs of allergic conjunctivitis are:**
   A. Unilateral onset
   B. Mucoid discharge
   C. Bilateral onset
   D. Watery discharge
   E. Severe itching
   F. Conjunctival papilla

8. **The keys to successful prevention of conjunctivitis are:**
   A. Avoid talking to other people
   B. Avoid touching or rubbing the eyes
   C. Wash hands regularly
   D. Avoid sharing cosmetics, bed linen, towels and clothing
   E. Environmental sanitation
   F. Discard used make-up, contact lens solution, and used disposable contact lenses

9. **The keys to trachoma treatment (the SAFE strategy) are:**
   A. Topical and systemic antibiotics for active disease
   B. Improvement of environmental condition
   C. Facial and hands hygiene
   D. Surgical treatment of complications
   E. Topical corticosteroids
   F. Lubricants or artificial tears

10. **The main complications of trachoma are:**
    A. Entropion.
    B. Symblepharon.
    C. Trichiasis.
    D. Corneal scarring.
    E. Vision loss.
    F. Glaucoma.

C. Clinical Cases

Case 1

A patient referred to a doctor with complaints of sudden redness of the right eye, development of photophobia, sensation of sand in the eyes, lacrimation which developed three days before, yellow purulent discharge from the eye. Today the same symptoms, but not so marked, developed in the left eye. Objective examination: marked edema of the eyelids, hyperemia of the conjunctiva in the area of the transitional fold. Fine punctate hemorrhages are seen in the conjunctiva of the upper eyelid, scanty lacrimal-purulent discharge. Define the diagnosis.

Case 2

A patient complains of redness of the eye, burning, irritation, foreign body sensation, light sensitivity, morning eyelash crusting, heaviness in the eyelids, blurry vision. Objectively: conjunctival hyperemia, congested lid margins, minimal mucopurulent discharge, presence of crusts on the lid margins, lid hyperemia. Define the diagnosis.

Case 3

A mother has noticed in her 1-day-old child a mucoid discharge and the lids get stuck together when the child sleeps. Examination of the eyes reveals mild conjunctival congestion and blepharospasm. The preauricular lymph nodes are palpable. Define the diagnosis.

Case 4

A patient complains of bad edema, pain, sticking of the eyelids. He also has general symptoms that include high fever, weakness, headache. The preauricular lymph nodes are swollen and tender. Examination: conjunctiva is red, infiltrated and covered with thick grey-yellow membrane that is firmly adherent to the conjunctiva and on removing bleeds. Discharge is serous bloody. What is your diagnosis?

Case 5

A patient complains of sudden onset of eye redness, severe itching, burning, foreign body sensation, extreme sensitivity to light, watery discharge, swelling of the eyelids, and respiratory infection. Examination: eyelid edema, pseudoptosis, conjunctival edema, follicular conjunctivitis, hyperemia, chemosis, palpable preauricular lymphadenopathy. Define the diagnosis.

Case 6

A patient complains of itching and redness of the eye, tearing, lid edema, mild eye pain, foreign body sensation, sensitivity to light, blurred vision, pus or watery discharge. Objectively: conjunctival injection, follicles on tarsal upper and lower conjunctiva and fornix, papillary hyperplasia, chemosis, superior micropannus, eyelid edema, mucopurulent or mucus discharge, tender preauricular lymphadenopathy. Define the diagnosis.

Case 7

A patient complains of pain in the eye, irritation and mild itching, pus discharge from the eyes, tearing, eyelid swelling, blurred vision, photophobia. Objective examination: conjunctival hyperemia, follicles which partly burst with signs of healing, scars on the conjunctiva with shrinkage, corneal ulcers. Define the diagnosis.

Case 8

A patient, 26 years of age, complains of eye itching, irritation, redness, burning, mucous discharge, light sensitivity, foreign body sensation upon removal of contact lenses, and contact lens intolerance. Objective examination: conjunctival hyperemia, the upper tarsal conjunctiva is irritated and swollen with giant papillae that give a "cobblestone" appearance, mucous discharge. What is your diagnosis?

Case 9

A 11-year-old child has blepharospasm, lacrimation, photophobia; he has been ill for several years; the process is exacerbated in autumn and spring. Objective examination: the conjunctiva is hyperemic, edematous, resembling cobblestones; there are yellow-grayish eminences near the limbus. What is your diagnosis?

Case 10

A patient, 50 years of age, complains of severe itching, burning and cutting pain in the eyes, redness of the skin in the external angles of the orbital fissure. Objective examination: the skin at the external angles of the orbital fissure is macerated, eczematous, there are single wet fissures. The eyelid conjunctiva is hyperemic and friable. The discharge is scanty as tenacious mucus. What is your diagnosis?

CHAPTER 8

Diseases of the Cornea

OBJECTIVES

Upon completion of the chapter the students should:

— know the classification of corneal diseases;
— know the basic diagnostic methods;
— know the main symptoms and clinical picture of keratitis;
— be able to establish the diagnosis of keratitis, differentiating it from other causes of red eye;
— know the methods of keratitis treatment;
— know the types of degenerative corneal diseases and anomalies of the corneal curvature;
— know the specifics of the clinical course of corneal diseases complications.

Plan:

1. CLASSIFICATION OF CORNEAL DISEASES
2. SYMPTOMS OF CORNEAL DISEASES
3. EXAMINATION METHODS
4. DISEASES OF THE CORNEA
   - **4.1. Congenital Anomalies**
     - Microcornea
     - Megalocornea
   - **4.2. Corneal Erosion**
   - **4.3. Inflammatory Diseases**
     - Superficial Punctate Keratitis
     - Infectious Keratitis
       - Corneal Ulcer
       - Herpetic Keratitis
       - Acanthamoeba Keratitis
       - Keratomycosis
     - Neurogenic Keratitis
       - Neuroparalytic Keratitis
     - Trophic Keratitis
       - Keratomalacia
     - Autoimmune or Related to Systemic Diseases Keratitis
       - Syphilitic Interstitial Keratitis
       - Tuberculous Interstitial Keratitis
   - **4.4. Degenerative Disorders of the Cornea**
     - Arcus Senilis
   - **4.5. Ectatic Conditions**
     - Keratoconus
     - Keratoglobus
   - **4.6. Dystrophies of the Cornea**
     - Epithelial Basement Membrane Dystrophy
     - Lattice Dystrophy
     - Fuchs' Dystrophy

1. Classification of Corneal Diseases

Corneal diseases can be classified:
— according to the origin — congenital or acquired;
— according to the clinical presentation — mild, moderate or severe;
— according to the onset of the pathological process — acute, chronic or recurrent;
— according to the dynamics of the pathological process — limited or progressive;
— according to the location — central or peripheral;
— according to the depth — superficial, deep, impending perforation, perforated;
— according to the etiology — exogenous or endogenous;
— according to the pathogenesis — traumatic, inflammatory, degenerative, dystrophies, ectatic conditions.

1.1. Congenital Anomalies — megalocornea, microcornea, congenital cloudy cornea, etc.

1.2. Inflammatory Diseases (Keratitis)

1.2.1. Infectious Keratitis
- Bacterial — superficial punctate keratitis, corneal ulcer, corneal ulcer serpens or hypopyon keratitis, marginal or catarrhal ulcer.
- Viral — Herpes simplex keratitis (punctate epithelial keratitis, dendritic keratitis, geographical ulcer, disciform keratitis), herpes zoster keratitis, adenoviral keratitis.
- Parasitic — acanthamoeba keratitis.
- Fungal — mucotic keratitis (keratomycosis or fungal ulcer).

1.2.2. Non-Infectious Keratitis
- Allergic — vernal, atopic.
- Autoimmune or related to systemic diseases — syphilitic interstitial keratitis, tuberculous interstitial keratitis.
- Trophic — keratomalacia (vitamin A deficiency).
- Neurogenic — neuroparalytic keratitis, neurotrophic keratopathy.

1.3. Degenerative Disorders of the Cornea — arcus senilis.

1.4. Dystrophies of the Cornea — epithelial basement membrane dystrophy, Meesmann dystrophy, Reis—Bücklers' dystrophy, gelatinous drop-like cor-

neal dystrophy, lattice dystrophy, Fuchs' endothelial dystrophy, Schnyder corneal dystrophy, granular corneal dystrophy, etc.

1.5. Abnormalities of the Corneal Curvature or Ectatic Conditions — cornea plana, keratoconus, keratoglobus.

1.6. Abnormalities in Corneal Transparency (Corneal Opacities) — nubecula, macula, corneal leukoma.

1.7. Trauma — corneal erosion etc.

2. Symptoms of Corneal Diseases

Corneal syndrome:
— eye pain;
— photophobia;
— tearing;
— blepharospasm;
— foreign body sensation.

3. Examination Methods

— Corneal examination by side illumination.
— Slit-lamp examination.
— Corneal sensitivity.
— Corneal size measurement.
— Dye Staining of the Cornea.
— Pachymetry.
— Keratometry.
— Corneal Topography.
— Microbial Investigation.

4. Diseases of the Cornea

4.1. Congenital Anomalies

Microcornea

Definition. Microcornea is a congenital malformation of the eye's cornea, in which the cornea has a too small diameter, less than 9 mm in newborns or 10 mm in adults (fig. 8.1).

Etiology. Microcornea usually occurs due to improper fetal development of the cornea at the fifth month of pregnancy. In addition, microcornea may also result from various processes leading to subatrophy or severe atrophy of the initially healthy eyeball.

Clinical Picture. The condition may be unilateral, but often bilateral. Rarely it occurs as an isolated anomaly with normal VA. Most often it is associated with other ocular anomalies such as microphthalmia or nanophthalmos and systemic anomalies (Marfan syndrome, myotonic dystrophy, fetal alcohol syndrome, etc.).

The corneal surface is usually flattened, which results in hyperopia. There is an increased risk of angle closure glaucoma development as the normal outflow of the aqueous humor is disturbed by the presence of embryonic mesenchyme fragments with a darkened lens or choroidal defects.

Complaints. Patients complain of a significant reduction in visual acuity.

Signs. Hyperopia, corneal diameter < 9 mm, shallow anterior chamber, congenital cataract, glaucoma, corneal opacification.

Methods of Examination. VA, refractometry, slit-lamp examination, corneal size measurement, keratometry, A- and B-scan ultrasound.

Differential Diagnosis. Microphthalmia, nanophthalmos, sclerocornea.

Treatment. The choice of treatment depends on the associated anomalies and severity of the condition. Hyperopia is treated with glasses or contact lenses that help prevent amblyopia. Surgical treatment of associated cataract, glaucoma or corneal opacification if they are present.

Prognosis. In most cases, the prognosis of this disease is relatively favorable with timely treatment. Otherwise it may lead to amblyopia or irreversible blindness.

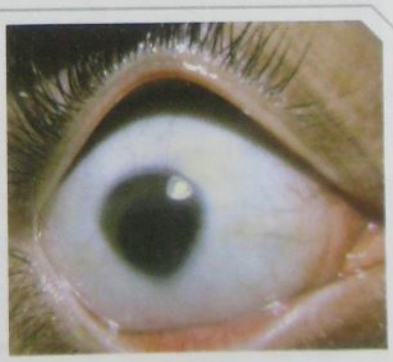


Fig. 8.1. Microcornea with coloboma of iris (from http://www.eyerounds.org)

Complications. Glaucoma, amblyopia.

Prophylaxis. Monitoring children with hyperopia for amblyopia, genetic testing for prediction of microcornea, pregnant women should avoid drinking alcohol.

Megalocornea

Definition. Megalocornea is a congenital anomaly of the cornea characterized by a very large cornea, 12 mm or more in horizontal diameter in newborns and 13 mm in adults (fig. 8.2).

Etiology. Megalocornea is a developmental anomaly related to birth defects. Postulated mechanisms of development include a defect in the formation of the optic cup, in which the anterior tips of the cup fail to fuse, allowing more space for the developing cornea. Quite often it is inherited at the genetic level in the same family. It is also worth noting that megalocornea can be a symptom of hydrophthalmos.

Clinical Picture. Megalocornea is a rare non-progressive condition and is usually bilateral. In spite of the cornea been enlarged, the eyeball is of normal size and the cornea itself is histologically normal and of normal thickness and clarity. The condition may be isolated and characterized by deep anterior chamber, high myopia and astigmatism, trabecular hyperpigmentation, or megalocornea but can be associated with other ocular and systemic findings, including lens subluxation due to zonular stretching, dysgenesis of the iris, Marfan syndrome, Rieger syndrome, Down syndrome, mental and neurologic impairment, etc.

Complaints. Usually none.

Signs. The cornea is enlarged, transparent. Corneal thickness may be slightly decreased or normal; the curvature can be increased or not changed. In megalocornea the cornea has a normal histological structure with a good density of the endothelial cells. High myopia, astigmatism, deep anterior chamber, normal IOP.

Methods of Examination. VA, refractometry, slit-lamp exam, corneal size measurement, gonioscopy, keratometry, A-scan ultrasound biometry, specular microscopy, tonometry.

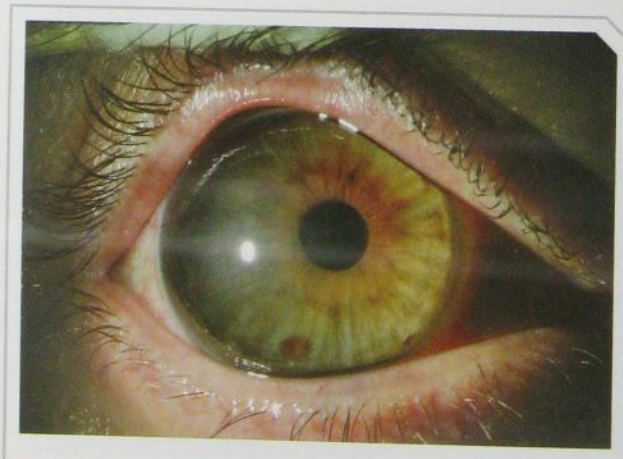


Fig. 8.2. Megalocornea (from http://www.eyerounds.org)

Differential Diagnosis. Hydrophthalmos, buphthalmos, primary congenital glaucoma, keratoglobus.

Treatment. The condition is usually not progressive and requires no treatment. In the presence of refractive errors (myopia, astigmatism) the patient is prescribed glasses or contact correction. Associated cataract requires surgical treatment.

Prognosis is good, but patents must undergo regular examination for the risk of cataract and glaucoma development.

Complications. Glaucoma, posterior subcapsular cataract, lens subluxation.

Prophylaxis. Children should be monitored for refractive amblyopia, genetic testing for prediction of megalocornea.

4.2. Corneal Erosion

Definition. Corneal erosion, also referred to as corneal abrasion or scratched eye, is the damage of the corneal epithelium.

Etiology. The appearance of corneal erosion can be caused by any interference, often mechanical injury — by foreign bodies in the eye, fingernails (scratches), makeup applicators, an edge of paper, contact lenses, sand or debris, etc. You may experience erosion of the cornea as a result of exposure to chemicals, most household chemicals, high temperatures. The eyelashes growing in the wrong direction can cause trauma during blinking of the corneal epithelium with the formation of erosion.

Clinical Picture. The clinical presentation of corneal erosion is usually unilateral as it is associated with trauma. It may be bilateral when associated with dystrophic diseases. Symptoms develop immediately after the occurrence of erosion and are associated with exposure of numerous nerve endings of the cornea.

Complaints. Patients' complains commonly are the following:

— sharp pain;
— tearing;
— photophobia;
— foreign body sensation;
— blepharospasm;
— eye redness;
— blurred vision;
— pain upon blinking and eye movement.

Signs. According to the causative agent they may be a foreign body on the cornea or under the eyelid, incorrect growth of the eyelashes. Slit-lamp exam shows light conjunctival injection, epithelial defects seen with ultraviolet light after fluorescein staining (fig. 8.3).

Methods of Examination. External examination, slit-lamp exam of the cornea and lids, staining of the cornea with 2 % fluorescein instilled into the eye. Examination should be made after an instillation of a topical anesthetic (Proxymetacain) as it is difficult for the patient to open the eye due to sharp pain.

Differential Diagnosis. Acute conjunctivitis, corneal foreign body, corneal ulcer, trichiasis.

NOTE!

Topical anesthetics may be given only to allow examination but should NEVER be used for repeated administration as they are toxic to the corneal epithelium.

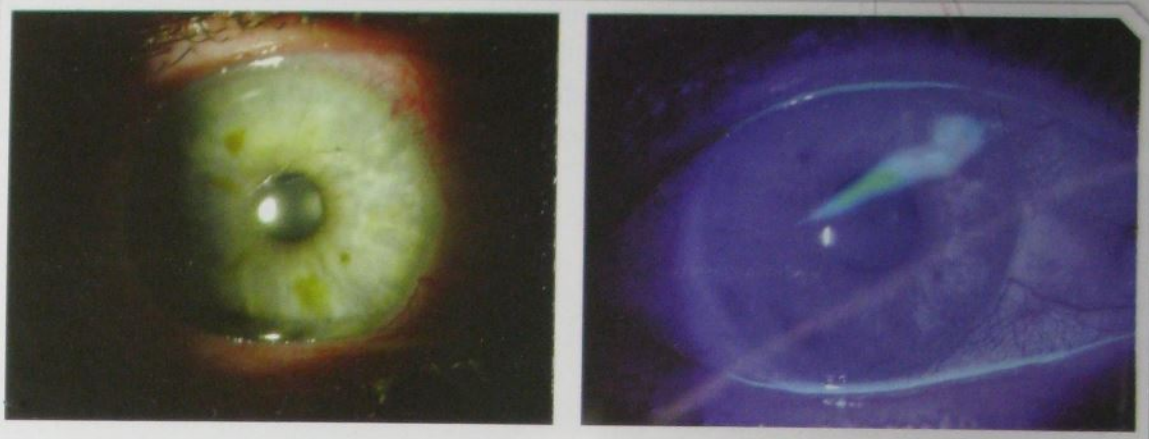


Fig. 8.3. Corneal erosion (direct view and after fluorescein staining)

Treatment. If the condition is caused by a foreign body, it has to be removed.

To prevent infection an antibacterial drug in the form of drops or ointments is usually assigned (Ofloxacin, Tobramycin, Levofloxacin, Tetracycline).

To restore the corneal epithelium, preparations that improve epithelization and keep the eye lubricated are prescribed (Dexpanthenol, Solcoseryl). The drops are administered during the day, and the ointment — at night, as it has a longer duration of action.

In some cases, scratched corneas are treated with a bandage contact lens. When used with eye drops, these special lenses provide pain relief and sometimes can speed healing. Regular contact lenses should be avoided.

For extreme pain or light sensitivity oral pain medicine may be prescribed.

Prognosis. With proper treatment, most superficial corneal erosions heal in a few days without any complications. Deeper erosions that penetrate through Bowman's membrane are more likely to cause permanent corneal scars that can interfere with vision. If necessary, severe scarring can often be treated successfully with a corneal transplant. Untreated corneal erosions can lead to blinding corneal ulcers.

Complications include corneal haze, corneal scarring, infectious keratitis, corneal ulcers and permanently decreased vision.

Prophylaxis. Most corneal erosions can be prevented by wearing protective eyewear to avoid eye injury and careful contact-lens use.

4.3. Inflammatory Diseases

Inflammation of the cornea is **keratitis**. According to etiology keratitis can be exogenous or endogenous. Exogenous keratitis results from either external injury to the cornea, or diseases of the adjacent conjunctiva, eyelids, Meibomian glands, lacrimal sac or invasion of infection from outside. Exogenous processes affect the superficial

cornea. Endogenous corneal diseases are caused by systemic illness, infection from within the patient or allergy. Endogenous keratitis affects the intracorneal processes.

Most forms of keratitis have the same symptomatology that is called *corneal syndrome*. This is severe pain in the eye, photophobia, tearing, blepharospasm, foreign body sensation. The objective signs of keratitis are pericorneal injection, corneal sensitivity, and transparency loss due to infiltration composed of modified corneal cells and leukocytes.

Keratitis is a medical emergency because extensive involvement may lead to blindness. Its treatment depends on the cause of keratitis. It is important to identify the causative agent to provide appropriate therapy. Bacterial and fungal ulcers, for example, require completely different medications.

After healing, superficial keratitis does not generally lead to scarring. Keratitis that has involved deeper layers of the cornea may develop a scar upon healing that impairs vision if it is located on or near the visual axis.

Outcome of keratitis:

— reduction of corneal sensitivity or quite the contrary — keratalgia;
— nubecula — semi-transparent opacity without strict borders. It causes slight blurring of vision if located at the centre;
— spot (macula) — thick opacity that causes considerable worsening of vision;
— wall-eye (leukoma) — thick opaque scar of the cornea.

Superficial Punctate Keratitis

Definition. Superficial punctate keratitis (SPK) is a form of keratitis characterized by punctate defects of the corneal epithelium.

Etiology. SPK can be caused by numerous conditions including contact lens wear, dry eyes, blepharitis, viruses, conjunctivitis, keratitis, trauma, chemical exposure, UV exposure, and even eye rubbing. It can also occur in association with an endogenous disorder such as Thygeson's disease.

Clinical Picture. SPK is characterized by multiple small, pinhead-sized, round lesions in the superficial layers of the cornea. They may vary in size, color and number, be scattered or located according to the causative factor. The condition is often associated with conjunctivitis. Most cases are bilateral, but the presentation is usually asymmetric.

Complaints. Depending on the cause and severity the symptoms vary from a nearly asymptomatic course to severe pain, intense eye redness, lacrimation, photophobia, foreign body sensation, blepharospasm and slightly decreased vision.

Signs. Fine epithelial defects of different color and size seen after fluorescein or Rose bengal staining (fig. 8.4), corneal edema, corneal infiltrates.

Methods of Examination. VA test, slit-lamp exam with fluorescein or Rose bengal staining, determination of corneal sensitivity.

Differential Diagnosis. Dry eye syndrome, herpes simplex keratitis, viral conjunctivitis.

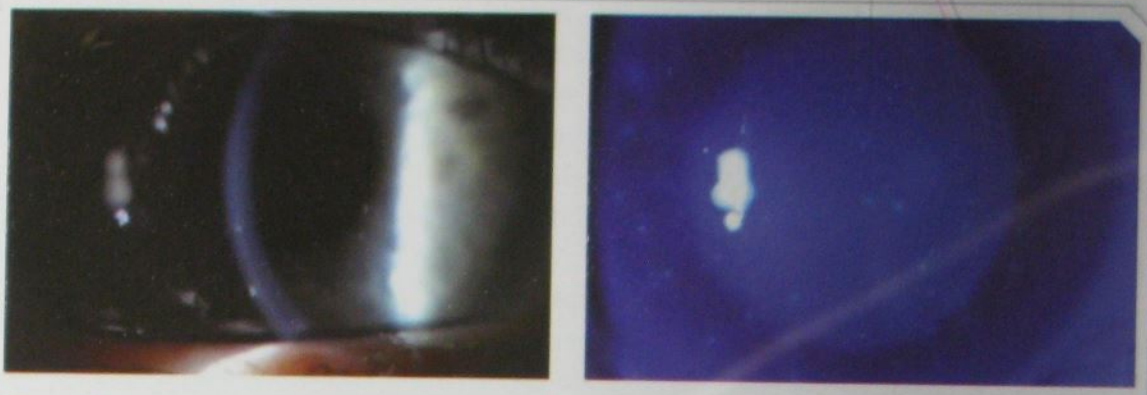


Fig. 8.4. Thygeson's superficial punctate keratitis (view at slit-lamp exam and with fluorescein staining)

Treatment depends on the cause and starts from treating the underlying condition. Other options are determined by the severity of symptoms and may include artificial tears (Carbomer, Hypromellose, Dexpanthenol), corticosteroids, cyclosporin A, soft contact lenses. If infectious etiology is suspected, topical antibiotics are added to the treatment. Patients that regularly use contact lenses should avoid using them until full recovery.

Prognosis for vision is excellent in cases of prompt treatment.

Complications. Mild scarring may develop in recurrent cases.

Prophylaxis. Traumatic causes may be prevented by wearing protective eyewear. Careful adherence to the rules of wear and care when using contact lenses. Timely detection and treatment of chronic blepharitis, conjunctivitis, and correction of immunodeficiency states.

Infectious Keratitis

Corneal Ulcer

Definition. Corneal ulcer, or ulcerative keratitis, is a serious infective inflammation of the cornea that involves damage of its epithelial layer affecting the corneal stroma and looking as an open sore.

Etiology. Corneal ulcers are most commonly caused by an infection with bacteria, viruses, fungi, or Acanthamoeba following trauma to the corneal epithelium. Wearing contact lenses while sleeping or wearing inadequately disinfected contact lenses can cause corneal ulcers too. Some patients with severe dry eye syndrome, allergic eye disease, nutritional deficiencies, immune system disorders and inflammatory diseases such as rheumatoid arthritis, lupus, psoriasis may develop corneal ulcers as a complication of their disease.

The most common bacterial pathogens associated with corneal ulceration are Staphylococcus aureus, Streptococcus pneumoniae, Pseudomonas pyocyanea, E. coli, Proteus, Klebsiella, Neisseria gonorrhoea, Neisseria meningitides, Corynebacterium diphtheriae, and, rarely, diplobacilli.

Clinical Picture. Primarily it should be noted that corneal ulcer may occur in chronic and acute forms. It presents as an infectious and infiltrative loss of corneal substance and looks as a grey-white to yellow-grey disc that can be located centrally, paracentrally or peripherally on the usually transparent cornea (fig. 8.5).

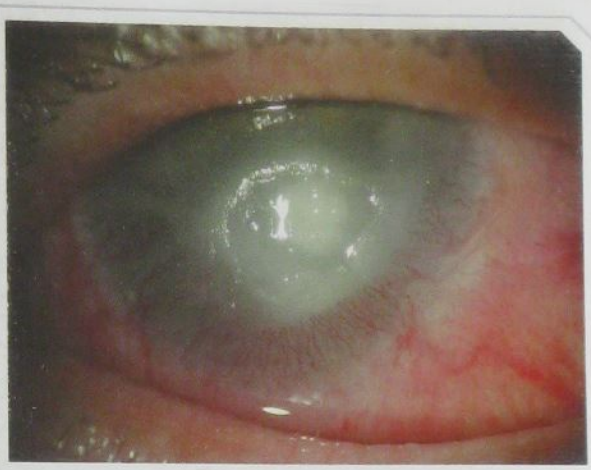


Fig. 8.5. Severe corneal ulcer with infiltration and neovascularization of the cornea

Corneal ulcer develops through 4 stages:

— *stage of progressive infiltration* — infiltration of lymphocytes into the epithelium from the peripheral circulation and the underlying stroma, necrosis of the involved tissue may occur;

— *stage of active ulceration* — necrosis and sloughing of the epithelium, Bowman's membrane and the stroma that can extend and progress by deeper penetration leading to descemetocele formation and possible corneal perforation; hyperemia of the circumcorneal vessels results in accumulation of purulent exudates on the cornea; exudation from the vessels of the iris and ciliary body into the anterior chamber lead to hypopyon formation;

— *stage of regression* — as the result of natural host defence and antimicrobial treatment; a line of demarcation develops around the ulcer; may be accompanied by superficial vascularization that increases the humoral and cellular immune response; the ulcer now begins to heal and epithelium starts growing over the edges;

— *stage of cicatrization* — healing process with progressive regeneration of collagen and formation of fibrous tissue.

Hypopyon Corneal Ulcer (Ulcer Serpens). Hypopyon corneal ulcer is a type of ulcer associated with formation of a hypopyon that is sterile pus in the anterior chamber as the result of iridocyclitis, which settles at the horizontal level.

Ulcer serpens is a characteristic hypopyon ulcer caused by pneumococcus. It appears as greyish white or yellowish disc-shaped ulcer occurring near the center of the cornea and has a tendency to creep over the cornea in a serpiginous fashion. This lesion is elevated along one edge that undermines the corneal stroma (advancing edge). This edge shows active ulceration and infiltration. Another edge is regressive. As the ulcer progresses this edge is being healed, cleared up, and epithelialized. In the course of time vascularization starts from this part.

Ulcer serpens is accompanied with marked iridocyclitis and has a great tendency for perforation.

Complaints. Symptoms of corneal ulcers are severe and include:

— eye pain;
— redness of the eye;
— significant decrease in vision;
— profuse lacrimation;
— photophobia;
— blepharospasm;
— discharge (purulent — in case of bacterial forms of keratitis, watery — viral forms).

Signs of corneal ulcer are:

— conjunctival hyperemia;
— ciliary congestion;
— a grey-white or yellow-grey disc on the cornea;
— chemosis;
— mild lid edema;
— anterior chamber — normal / hypopyon;
— iritis (may not be present).

Methods of Examination. VA test, slit-lamp exam with fluorescein staining, scraping from the conjunctiva and the ulcer itself for determining the causative organisms.

Differential diagnosis. Atopic keratoconjunctivitis, dry eye syndrome, herpetic keratitis, keratomycosis.

Treatment. Corneal ulcer is an emergency condition and requires immediate aggressive treatment in the hospital.

Usually, initial treatment consists of systemic and topical broad-spectrum antibiotics (such as Ofloxacin, Levofloxacin, Lomefloxacin and Gentamicin) until culture results identify the causative organism. As soon as the results of bacteriologic and resistance testing are available, medications may then be changed to more specific to target the cause of the infection. Bacterial corneal ulcer requires intensive treatment with antibiotics; fungal — application of topical anti-fungal agents such as Natamycin; viral — topical antivirals like Acyclovir.

In addition, for improving the epithelization of the cornea Dexpanthenol, Solcoseryl are helpful. Cycloplegics (Atropine) are indicated in the presence of hypopyon to prevent or relieve the ciliary spasm and minimize the complications of accompanying anterior uveitis.

Systemic and topical antihistamine and vitamin medications are indicated as well. Oral pain medication may be prescribed to reduce pain.

Emergency penetrating keratoplasty (PK) is indicated to treat a descemetocele or a perforated corneal ulcer. PK is a surgical replacement of the damaged cornea with a full thickness cornea transplant.

NOTE!

Topical corticosteroids and anesthetics are usually contraindicated in corneal ulcer as they may lead to superinfection with fungi and other bacteria and will often make the condition worse.

Broad areas of superficial necrosis may require a conjunctival flap to accelerate healing. Stenosis or blockage of the lower lacrimal system that may also impair healing of the ulcer should be surgically corrected.

Prognosis. The prognosis depends on the size, location, depth, and etiology of corneal ulcer as well as any pre-existing ocular conditions. With proper treatment, corneal ulcers should improve within two to three weeks. Although most corneal ulcers will cause some degree of scarring, the scar will often not cause any visual loss. If the ulcer is deep, dense, and central, scarring will cause decrease of vision.

Complications. Corneal scarring, irregular astigmatism, secondary cataracts and glaucoma, descemetocele, corneal perforation with secondary infections, loss of vision, loss of the eye.

Prophylaxis. Avoidance of predisposing factors may reduce the risk of corneal infection. Proper education of contact lens use and care can help decrease the risk. The use of protective eyewear for sports and outdoor activities can help prevent trauma and subsequent development of infection.

Herpetic Keratitis

Herpes simplex is a common recurrent infection of the skin or mucous membrane caused by the herpes simplex virus (HSV). A typical feature of the herpes simplex virus is an unnoticed primary infection that often heals spontaneously. Many people then remain carriers of the neurotropic virus, which can lead to recurrent infection at any time proceeding from the trigeminal ganglion. Recurrences may be triggered by exposure to ultraviolet light, stress, menstruation, generalized immunologic deficiency, or febrile infections.

HSV may affect any part of the eye causing a simple infection or a condition that can possibly result in blindness. It typically affects the conjunctiva, cornea, eyelids, and surrounding skin causing conjunctivitis, keratitis, blepharitis, and dermatitis respectively. Sometimes deeper eye structures may be involved in the inflammatory process causing herpetic iridocyclitis or retinitis.

Definition. Herpetic simplex keratitis is inflammation of the cornea caused by the virus of herpes simplex. It is the most common form of viral keratitis and is a common cause of corneal morbidity and blindness.

Etiology. Herpes simplex keratitis is primarily caused by HSV type 1, which can also cause cold sores in and around the mouth and face. Primary HSV infection occurs by direct contact with infected secretions or lesions and then the eye, recurrent cases result from reactivation of the virus that was in a latent form by trigger factors. Type 2 herpes simplex virus is a usual cause of genital herpes, but also can affect the eyes by direct transmission of genital secretions into the eye.

Clinical Picture. HSV keratitis can present with involvement of the corneal epithelium, stroma or endothelium with or without associated inflammation of the anterior chamber. Most cases are unilateral, but bilateral cases may occur, especially in immunocompromised patients.

Epithelial forms of keratitis are caused by the actively replicating virus and can present as one of four different lesions: punctate superficial keratitis, dendritic keratitis, geographic keratitis, and marginal keratitis.

Punctate superficial keratitis is characterized by development of small, raised, clear vesicles that resemble vesicular eruptions seen in the skin or mucous membranes. This is mostly a sign of primary infection. In this case, there may appear foreign body sensation, pain, tearing, sensitivity to light, redness of the eye. These symptoms are associated with damage to the corneal epithelium and irritation of the nerve endings.

As the disease progresses these vesicles burst and coalesce into a dendritic pattern with central location.

Dendritic keratitis is the most common form of recurrent herpetic eye disease. It presents as a characteristic branching (dendritic or serpentine), linear lesion of the corneal epithelium with each branch terminating with a bulb and following the nerve pattern throughout the cornea. The borders of the lesions are swollen and slightly raised, and contain active virus (fig. 8.6). These lesions stain green with fluorescein and branch resembling a tree. Corneal sensation is reduced in approximately 70 % of patients.

The majority of dendritic keratitis cases heal without serious complications but some may progress to involve the corneal stroma. After dendritic epithelial keratitis resolves, a dendritic scar, called a ghost dendrite, may remain in the superficial stroma.

Geographic keratitis is an enlarged dendritic keratitis that is no longer linear but has a much larger epithelial defect with a form that often resembles the shape of a country — hence the term 'geographic'. The borders of the lesions are swollen and contain active virus.

With progression of the infection deeper into the cornea layers, the stroma may become affected. The condition is referred to as stromal keratitis.

Stromal keratitis is an inflammation of the corneal stroma due to progression of HSV epithelial keratitis or immune-mediated response to nonreplicating viral parti-

Fig. 8.6. Herpes simplex virus dendritic keratitis (view at slit-lamp exam and stained with fluorescein)

cles. According to clinical manifestation stromal keratitis can be subdivided into two forms — necrotizing or immune stromal keratitis. In the necrotizing disease, viral replication causes a dense stromal infiltrate, ulceration, and necrosis. It is accompanied by an overlying epithelial defect and a high risk of stromal thinning and perforation. In immune stromal keratitis, also known as non-necrotizing or interstitial stromal keratitis, the epithelium is intact, and the pathology is thought to be driven primarily by immune response.

Stromal keratitis leads to greyish stromal opacity and is often associated with corneal neovascularization, and recurrent episodes can lead to irreversible stromal scarring and vision loss. Keratic precipitates, uveitis, and raised IOP may also occur in conjunction with stromal keratitis.

Endothelial keratitis or endotheliitis is an inflammation of the corneal endothelium and represents an immune reaction to viral antigens. HSV endotheliitis has three clinical presentations — disciform, which is the most common, diffuse, and linear.

Disciform keratitis is characterized by clouding and deep, disc-shaped corneal edema in a central or paracentral region with a clear demarcation between the involved and uninvolved cornea. This condition is associated with folds in Descemet's membrane, anterior uveitis, fine keratic precipitates accompanied by pain, photophobia, injection, and decrease of vision.

Herpes Zoster Keratitis. Proceeding from the trigeminal ganglion, the virus reinfects the region supplied by the trigeminal nerve. The eye is only affected where the ophthalmic division of the trigeminal nerve is involved. In this case, the nasociliary nerve supplying the interior of the eye will also be affected. Hutchinson's sign, vesicular lesions on the tip of the nose, will be present.

Complaints. Common symptoms of ocular herpes include:

— severe pain in and around the eye;
— open sores/blisters around the eye;
— swelling of the eyelids;
— redness of the eye;
— excessive tearing;
— sensitivity to light;
— foreign body sensation;
— irritation and itching;
— blurring of vision;
— haloes around light sources;
— loss of vision.

Signs of herpetic simplex keratitis differ according to the type and severity of the condition and may include:

— corneal injection;
— eyelid edema;
— reduced corneal sensitivity;
— decreased VA;
— photophobia;

— epithelial keratitis — dendritic epithelial defects with terminal bulbs that stain with fluorescein;
— stromal keratitis (non-necrotizing) — deep stromal infiltrate and edema, deep stromal vascularization, immune ring, scarring or thinning of the stroma, intact epithelium;
— stromal keratitis (necrotizing) — all of the above plus epithelial defects, progressive necrosis, and/or ulceration;
— disciform keratitis — central cornea edema, folds in Descemet's membrane, keratic precipitates, elevated IOP.

Methods of Examination. VA test, slit-lamp exam with fluorescein dye, corneal sensitivity, laboratory diagnostics (specific antibodies by enzyme-linked immunosorbent assay (ELISA) and herpes viruses themselves by polymerase chain reaction (PCR)).

Differential Diagnosis. Acanthamoeba keratitis, fungal keratitis, syphilitic keratitis, neurotrophic keratitis, corneal ulcer, allergic conjunctivitis, viral conjunctivitis, epidemic keratoconjunctivitis.

Treatment of herpes simplex keratitis should be started as soon as possible to prevent sight-threatening complications development. Treatment options depend on the type of HSV keratitis or the part of the cornea involved and severity of the condition.

Herpetic epithelial keratitis requires antiviral therapy, topical, and/or oral medications that include:

— topical Idoxuridine 0.1 % solution every hour during the day and every 2 hours during the night until definitive improvement, then 1 drop every 2 hours during the day and every 4 hours during the night, or 0.5 % ointment every four hours during the day and once before bedtime;
— topical Trifluridine 1 % solution every two hours for 7 days in the affected eye until the corneal epithelium is sufficiently healed. Further treatment is then suggested at four times daily for another week;
— topical Ganciclovir 0.15 % gel five times per day until healing, and then 1 drop 3 times per day for another 7 days;
— oral Acyclovir 400 mg five times daily for seven to 10 days (or 3 times daily for recurrent herpes simplex keratitis);
— oral Valacyclovir 500 mg three times daily for seven to 10 days;
— immunocompromised patients may require IV antivirals (e.g., Acyclovir 5 mg/kg IV every 8 hours for 7 to 14 days);
— occasionally, removal of the infected and damaged eye cells with a soft cotton-tipped applicator may help speed healing;

NOTE!

Topical antivirals should not be used longer than 21 days due to their epithelial toxicity.

NOTE!

Topical corticosteroids are contraindicated in cases with epithelial involvement when active viral replication as they can accelerate the spread of viral infections.

— for immunotherapy interferon (leukocyte alpha interferon (200 U/ml) or related drugs — interleukin (10000 ME in 0.1 ml phosphate buffer), reaferon (5000—100 000 ME/ml in distilled water)) and anti-herpetic vaccine are used.

Herpetic stromal keratitis treatment methods depend on the epithelium status. If stromal disease is accompanied with a concomitant epithelial defect, it is treated similarly to epithelial keratitis, with a topical antiviral agent and a cycloplegic agent administered until the epithelium has healed.

Herpetic stromal keratitis without associated epithelial disease or after resolution of the epithelial defect is treated with topical corticosteroids in addition to antiviral drugs:

— corticosteroids Prednisolone 1 %, or Dexamethasone 0.1 % instillation every 2 hours initially, then the interval is extended to every 4 and 8 hours as symptoms improve;
— cycloplegics Atropine 1 % or Scopolamine 0.25 % 1 drop 3 times daily to relieve pain and photophobia with associated uveitis;
— topical Cyclosporine 2 % is used to facilitate epithelial healing if ulceration is present in stromal disease.

Herpetic endothelial (disciform) keratitis is treated with combined corticosteroid and antiviral therapy. Associated elevated intraocular pressure can be treated with timolol and systemic acetazolamide, as necessary.

In patients with severe corneal scarring secondary to herpetic keratitis penetrating keratoplasty is the primary surgical option for visual rehabilitation. But it should not be undertaken until the herpetic disease has been inactive for many months. Postoperatively, recurrent herpetic infection may occur as a result of surgical trauma and topical corticosteroids may be necessary to prevent corneal graft rejection.

Prognosis. The prognosis of keratitis primarily depends on the location and depth of corneal lesions. With timely treatment superficial infiltrates completely dissolve or leave slight opacification, with almost no effect on visual acuity. Deep keratitis, ulcers, especially if they are located in the central zone of the cornea, can lead to a significant reduction of visual acuity due to formation of intensive opacity and rough scars.

Complications. Corneal complications of herpetic eye disease range from epitheliopathy to frank neurotrophic or metaherpetic ulcers. Long-standing disciform keratitis may also result in bullous keratopathy. Late complications of deep vascular stromal scarring include secondary lipid keratopathy. Finally, stromal inflammation may lead to visually significant irregular astigmatism, corneal scarring, and loss of vision.

Prophylaxis. Currently there are no proven methods for preventing HSV keratitis, but some steps may help to avoid its recurrences, for instance

— preventing eye injuries and microtrauma by wearing sunglasses and appropriate eye gear as needed;

— avoid touching the eyes unless you have washed your hands properly, especially if you have a cold sore or a herpes blister;
— routine contact lens hygiene;
— avoid sharing eye makeup;
— balanced diet;
— oral antivirals for preventing recurrences.

Acanthamoeba Keratitis

Definition. Acanthamoeba keratitis is a rare but serious parasitic infection of the cornea by Acanthamoeba, which may cause corneal ulceration leading to blindness.

Etiology. Acanthamoeba keratitis is caused by a microscopic free-living amoeba (single-celled living organism) called Acanthamoeba that is one of the most abundant and ubiquitous protozoa on Earth. It is very common in various water sources (lakes, oceans, and rivers) as well as in untreated domestic tap water, swimming pools, hot tubs, and even drinking water.

In most cases Acanthamoeba keratitis follows minor corneal trauma and occurs in people who wear contact lenses for vision correction, but anyone with a corneal injury is susceptible to developing the infection. Keratitis is usually associated with improper contact lenses hygiene, and swimming in natural water or a swimming pool, especially while contact lenses are worn.

Clinical Picture. Acanthamoeba keratitis presents with nonspecific symptoms similar to bacterial and viral keratitis that makes it difficult to define the diagnosis and can be crucial.

Early disease presents as nonspecific keratitis. Possible findings include epithelial infection that may appear as dendritic, punctuate epithelial erosions, microcysts, and epithelial haze. Later signs include central or paracentral stromal infiltrates that develop into a classic ring-form, disciform, or nummular shape (fig. 8.7).

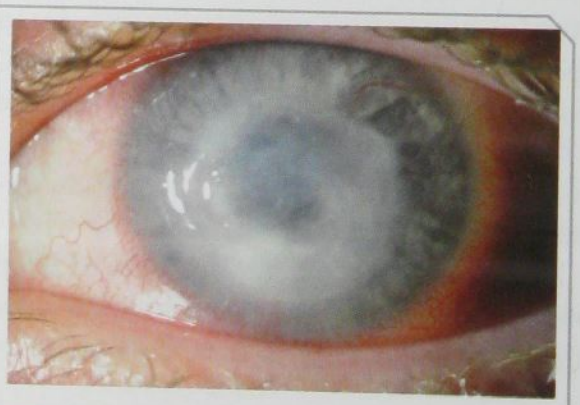


Fig. 8.7. Acanthamoeba keratitis with corneal infiltration, injection and neovascularization

As the disease progresses scleritis, conjunctival hyperemia, radial keratoneuritis, and anterior uveitis with possible hypopyon appear. Advanced signs include stromal thinning and corneal perforation. If left untreated, Acanthamoeba can spread back into the retina and cause serious chorioretinitis.

Complaints of Acanthamoeba keratitis can be very similar to the symptoms of other corneal infections. These symptoms, which can last for several weeks or months, may include:
— eye redness;
— severe eye pain;

— blurred vision;
— decreased vision;
— sensitivity to light;
— foreign body sensation;
— excessive tearing;
— loss of vision.

Signs. Clinical signs vary with the stage of the disease and its progression.

Early:
— corneal injection;
— punctate erosions;
— dendritic epithelial defects;
— mild infiltrates;
— lid edema;

Late:
— classic dense ring-shaped infiltrates;
— radial keratoneuritis;
— anterior uveitis;
— hypopyon;
— cataract;
— corneal thinning, perforation;
— chorioretinitis.

Methods of Examination. VA, slit-lamp exam, fluorescein dye, confocal microscopy, and polymerase chain reaction.

Differential Diagnosis. Herpetic stromal keratitis, fungal keratitis, recurrent corneal erosion.

Treatment. The infection can be difficult to treat due to the resilient nature of the cyst form. Current treatment regimens usually include:

— topical cationic antiseptic agents biguanides such as Polyhexamethylene biguanide (PHMB 0.02 %) or Chlorhexidine (0.02 %) with or without a diamidine such as Propamidine isethionate (0.1 %) or Hexamidine (0.1 %) and aminoglycosides such as Neomycin (1 %). The therapy may last from six months to a year starting at hourly intervals and tapered as the clinical situation improves;
— topical cyclopegics (e.g., Scopolamine 0.25 % three times daily) for pain control;
— oral NSAIDs (e.g., Naproxen 250 to 500 mg orally two times a day) to reduce inflammation;
— corneal debridement in order to prepare topical treatment;
— orally Itraconazole (400 mg), or Ketoconazole (200—600 mg/day), or Voriconazole (200 mg);
— penetrating keratoplasty for visual acuity restoration in case of failure of medical treatment.

NOTE!

The use of topical steroids in Acanthamoeba keratitis treatment is controversial.

All patients with Acanthamoeba keratitis must stop wearing contact lenses.

Prognosis. The prognosis for Acanthamoeba is worse than for many other types of infectious keratitis, and prevention is therefore very important. However, especially if caught early, satisfactory outcomes can certainly be achieved.

Complications. In most cases Acanthamoeba keratitis may lead to cataract, hypopyon, increased intraocular pressure, glaucoma, corneal ulceration, corneal scarring, loss of vision, loss of the eye.

Prophylaxis. Basic preventive measures are to maintain proper contact lens hygiene, not to swim or take a bath or shower while wearing contact lenses, to avoid swimming in open water in the presence of eye injury. Early detection and adequate treatment are important to avoid permanent visual impairment.

Keratomycosis

Definition. Keratomycosis, or fungal keratitis, or mycotic keratitis is an inflammatory disease of the cornea, caused by a fungus.

Etiology. More than 70 different fungi species have been reported to cause fungal keratitis, among them members of the genera Fusarium, Aspergillus and Candida are the most frequent causes of corneal infection.

The most common fungal keratitis follows ocular trauma especially during gardening since soils, plant materials, and dust from the many gardening additives applied for improved growth, all harbor fungi. Contact lens wear is considered to be a significant risk factor for keratomycosis development.

It is reported that the incidence of keratomycosis has risen sharply over the recent years as a result of the increased and often unwarranted use of antibiotics and corticosteroids in ophthalmology. Chronic keratitis, exposure keratopathy, and immunosuppressive diseases may also lead to mycotic corneal infection.

Clinical Picture. Keratomycosis is a rare but severe corneal infection, which leads to a corneal dense grey-white ulcer with a dry base or to a necrotizing infiltration. The infiltrates and ulcer spread very slowly and can penetrate into the deeper layers of the stroma, and further, through intact Descemet's membrane.

The ulcer appears as a fluffy white raised protuberance with ragged borders. When the superficial layers slough off, the base of the ulcer remains dry. Around and beneath the ulcer there is a dense infiltrate extending deep into the corneal stroma. The infiltration often has a fatty, yellow-white appearance and is sharply outlined. Discrete satellite infiltrations and hypopyon may also develop. There is ciliary and conjunctival injection (Fig. 8.8).

If left untreated, the infection will spread into the anterior chamber and result in corneal perforation and loss of the eye.

Complaints. The most common complaints are:

— eye redness;
— severe eye pain;
— blurred vision;
— decreased vision;

— sensitivity to light;
— foreign body sensation;
— tearing;
— mucopurulent discharge.

Signs:

— ciliary and conjunctival injection;
— raised white-grey corneal ulcer with rough dry texture and feathery borders;
— stromal infiltrates with sharp margins;
— satellite lesions;
— endothelial plaque made of leukocytes underneath the lesions;
— hypopyon;
— purulent secretions;
— scleritis,
— endophthalmitis;
— corneal perforation.

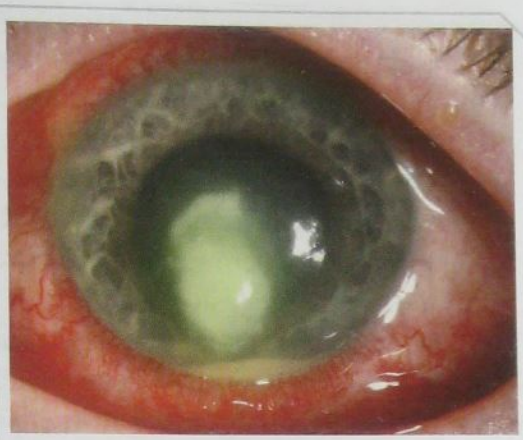


Fig. 8.8. Fungal keratitis with hypopyon in the anterior chamber (from http://www.eyerounds.org)

Methods of Examination. History of ocular trauma (always be suspicious after trauma with vegetable material), VA test, slit-lamp examination, upper lid eversion for excluding the retained foreign body, confocal microscopy, laboratory analysis of the scrapings of the ulcer base, cornel biopsy.

Differential diagnosis. Bacterial keratitis, Acanthamoeba keratitis, herpetic keratitis, recurrent corneal ulcer, conjunctival or corneal foreign body.

Treatment. The condition is difficult to treat. Infiltrates and corneal ulcers of unknown etiology are treated as bacterial, until fungal etiology has been proved, then treatment options are as following:

— topical antimycotic agents Natamycin 5 % (for filamentous fungal infection), Amphotericin B 0.15 % (for candida infection or non-responding cases) or Econazole 1 %, Fluconazole 2 %, Miconazole 1 % and Clotrimazole 1 % solutions initially are given hourly for 48 hours and then reduced as signs permit. Because most antifungals are fungistatic, successful treatment should be continued for prolonged periods at least 6—12 weeks;
— in severe cases, Voriconazole 1 % every hour during the day may be used;
— oral treatment is indicated in severe intraocular involvement with suspected endophthalmitis — Fluconazole (200—400 mg), Itraconazole (200 mg), Voriconazole (200 mg) two times a day;

NOTE!

Among antifungal medicals *Natamycin* is the only commercially available topical antifungal drug; all others are to be specially prepared in sterile conditions by pharmacist from solutions for intravenous injection.

— cycloplegic agents: if there is a hypopyon, Atropine 1 % solution three times a day;
— in bacterial co-infection broad-spectrum topical antibiotics may be recommended;
— corneal debridement every 24—48 hours in order to remove the affected epithelium and facilitate penetration of antifungal drugs to the site of the lesion;
— penetrating keratoplasty in cases not responding to antifungal therapy and infection progression.

NOTE!

Topical steroids must be avoided as they activate the growth of fungi.

Prognosis. The prognosis varies depending on the depth and size of the lesion and the causative organism. In general, small superficial keratomycosis responds well to topical therapy. Deep stromal infections and infections with concomitant intraocular involvement are much more difficult to treat. Approximately in one third of these cases, the treatment of fungal infections of the cornea fails and results in perforations.

Three factors significantly associated with failed treatment of fungal keratitis: large ulcer size (> 14 mm), the presence of hypopyon, and Aspergillus as the causative organism.

Complications. Corneal scarring, glaucoma, endophthalmitis, loss of vision, corneal perforation, loss of the eye.

Prophylaxis. Wearing protective eyewear while gardening or working in an agricultural environment will reduce the risk of ocular trauma, also general hygiene, proper contact lens care, and avoidance of nonessential steroid and antibiotic use can prevent mycotic infection.

Neurogenic Keratitis

Neuroparalytic Keratitis

Definition. Neuroparalytic keratitis, referred to also as neurotrophic keratitis or neurotrophic keratopathy is a degenerative disease of the cornea characterized by reduction or absence of corneal sensitivity resulting from impaired trigeminal innervation.

Etiology. The condition can be developed by the damage at any level of the fifth cranial nerve, from the trigeminal nucleus to the corneal nerve endings. Corneal sensory denervation results in decreased epithelium thickness, poor epithelial wound healing and induction of an inflammatory response.

The most common causes are herpetic keratitis, ophthalmic and neurosurgical procedures that damage the trigeminal ophthalmic branch and result in its paresis or palsy. Other ocular causes of impairment of corneal sensitivity include corneal surgery or trauma, chemical burns, corneal dystrophy, chronic use of topical medications, or contact lens wear. Systemic diseases such as diabetes mellitus, vitamin A deficiency, multiple sclerosis, leprosy, head trauma, cerebral tumors or aneurysms can also cause impairment of trigeminal corneal innervation.

Clinical Picture. Clinical presentation of neuroparalytic keratitis differs according to the severity of corneal damage. It develops through three stages according to the Mackie classification:

— stage 1 is characterized by irregularity of the epithelium, punctate keratopathy, superficial neovascularization, corneal edema, and stromal scarring;
— stage 2 — persistent epithelial defect due to impaired healing;
— stage 3 is characterized by the development of corneal ulceration, melting, and perforation.

Complaints. Since corneal sensory innervation is impaired, the disease is initially asymptomatic, and it is often found by accident. Sometimes patients with neuroparalytic keratitis may complain of redness of the eye and blurred vision due to irregular epithelium or persistent epithelial corneal defects, scarring, or edema.

Signs differ according to the stage of the condition and may include:

— reduction or absence of corneal sensitivity with side and forehead skin hypoesthesia;
— slow blink reflex;
— decreased reflex tearing;
— decreased tear-break time;
— mild conjunctival hyperemia;
— punctate epithelial staining with fluorescein;
— dry and cloudy corneal epithelium;
— corneal edema;
— loss of epithelium with a surrounding rim;
— Descemet's membrane folds and stromal edema;
— corneal ulcer with stromal involvement;
— stromal melting;
— corneal perforation.

Methods of Examination. VA test, slit-lamp exam, corneal sensitivity, fluorescein (lissamine green or rose bengal) staining, Schirmer test, pachymetry, keratometry, confocal microscopy, corneal scrapings and corneal biopsy. If it is necessary to eliminate the tumor compression of the trigeminal nerve, MRI, X-ray of the orbit, skull radiography, radiography of the paranasal sinuses can be conducted.

Differential Diagnosis. Blepharitis, dry eye, punctate corneal keratopathy, superficial corneal vascularization, exposure keratitis, herpetic keratitis, contact lens-related disorders, chemical injury, toxic, or immune corneal ulcers.

Treatment. Neuroparalytic keratitis is one of the most difficult corneal diseases to treat as no medication can improve corneal sensitivity. The primary aim of treatment is identifying and addressing the underlying cause. Topical treatment is directed at protection of the corneal surface, prevention of corneal damage progression and promotion of epithelial healing. It includes the use of:

— frequent application of preservative-free artificial tears eye drops, gels and lubricant ointments to protect the corneal epithelium;

— antibiotic eye drops or ointments can be prescribed to prevent or treat secondary bacterial infection;
— therapeutic soft contact lenses or patching of the affected eye to promote corneal defect healing;
— botulinum A toxin injection in the levator muscle;
— surgical options such as lateral tarsorrhaphy, palpebral spring, amniotic membrane transplantation or conjunctival flap if the lesions do not heal for a long time.

NOTE!

Topical steroids and non-steroidal anti-inflammatory drugs are contraindicated due to their potential of causing corneal melting as a side effect of their anesthetic properties.

Prognosis is generally poor due to impairment in wound healing and depends on the cause and severity of the trigeminal damage, degree of corneal sensitivity reduction, duration of the condition and on the presence of associated ocular surface disease. ••

Complications. Secondary bacterial keratitis, corneal ulcers, corneal scarring, corneal perforation following stromal melting.

Prophylaxis. There are no preventive methods of the condition. Therefore, patients with impaired corneal sensitivity should be instructed to seek the help of an ophthalmologist immediately if the eyes become red or if their vision changes. Patients need to understand that this condition is very serious and may not cause any pain.

Trophic Keratitis

Keratomalacia

Definition. Keratomalacia, also referred to as xerotic keratitis, is a severe condition characterized by cornel drying, softening and clouding due to a vitamin A deficiency and insufficient protein and calories in the diet.

Etiology. Vitamin A deficiency or hypovitaminosis A is the leading cause of preventable blindness in children in developing countries, especially in Africa and South-East Asia. It may be associated with poor nutrition or insufficient absorption of the vitamin due to underlying health condition, such as ulcerative colitis, cystic fibrosis, liver disease, or intestinal bypass surgery and any condition that affects absorption of fat-soluble vitamins. Systemic disorders like measles, pneumonia, or chronic diarrhea can also cause keratomalacia.

Clinical Picture. During the initial stage of the disease, the cornea as well as the conjunctiva becomes extremely dry (xerosis) and as therapy begins the cornea regains its normal appearance in 1—2 weeks.

Left untreated the condition leads to opacification, haziness, wrinkling, and softening of the cornea with the developing of characteristic foamy silver grey deposits on the conjunctiva covering the sclera called Bitot's spots.

With advancing vitamin A deficiency, increasing softening of the cornea may lead to secondary infection, ulceration, full thickness dissolution of the cornea and its rupture (perforation) with possible extrusion of the intraocular contents, degenerative tissue changes (e.g., ocular shrinking), and panophthalmitis resulting in blindness.

Complaints. Patients complaint of poor vision in dim light (night blindness), extreme dryness of the eye, photophobia. They may notice blurring and decrease of vision, dry and scaly skin, foamy deposits on the eye.

Signs.

— nyctalopia;
— conjunctival sclerosis;
— Bitot's spots, or protein deposits;
— corneal xerosis;
— corneal edema and cloudiness;
— corneal ulceration;
— corneal softening and necrosis (melting);
— corneal scarring;
— staphylomas;
— corneal perforation;
— xerophthalmic fundus — numerous small yellowish dots on the fundus of the eye.

Methods of Examination. VA test, slit-lamp exam, corneal staining, ophthalmoscopy, examination of dark adaptation, color blindness test, electroretinography, blood tests to find out the markers, which may suggest vitamin A deficiency such as plasma retinol and retinol binding proteins that are significantly suppressed in vitamin A deficiency.

Differential Diagnosis. Dry eye syndrome, corneal ulcer.

Treatment of keratomalacia includes vitamin A supplements in the form of capsules or injections and protein-rich diet. The dosage is determined by the severity of the condition and amounts according to the WHO recommendations to 50,000 IU for infants below 6 months of age, 100,000 IU for infants 6—12 months of age, and 200,000 IU for those aged 12 months through adulthood, 20,000 IU within 8 weeks of delivery.

Topical treatment includes intensive lubrication of the eyes with eyedrops, ointments or bandage contact lenses to protect the corneal epithelium, and antibiotic eye drops or ointments to prevent or treat secondary bacterial infection. In cases with severe corneal scarring, after the acute situation has settled, keratoplasty may be taken into consideration.

Prognosis. With appropriate early treatment and proper replacement of vitamin A, the prognosis is generally improved. However, as the condition progresses and keratomalacia develops, corneal changes may be irreversible.

Complications. The most common complications of keratomalacia are chronic corneal infection, corneal scarring, corneal rupture, blindness, atrophy of the eyeball.

Prophylaxis. Measures to prevent keratomalacia include balanced diet, vitamin A supplementation and an underlying disease treatment.

Autoimmune or Related to Systemic Diseases Keratitis

Syphilitic Interstitial Keratitis

Definition. Syphilitic interstitial keratitis is a chronic nonsuppurative inflammation of the corneal stroma associated with syphilis.

Etiology. Syphilitic keratitis is an uncommon manifestation of syphilis, it accounts only for 5 % of all cases of syphilitic eye diseases. But prior to antibiotics, syphilis was the most common cause of interstitial keratitis. Today conditions causing interstitial keratitis include herpes simplex virus (HSV), herpes zoster, leprosy, Lyme disease, tuberculosis, and HLA-B27-associated inflammatory conditions.

Most cases (about 90 %) of syphilitic interstitial keratitis are due to congenital syphilis from mothers with primary, secondary, or early latent disease. It is now generally accepted that the disease is a manifestation of delayed local immune-mediated reaction, not a manifestation of an active infection. The inflammation is usually triggered by an injury or an operation on the eye and commonly develops between ages of 5 and 20.

Interstitial keratitis in acquired syphilis is very rare, uveitis and retinitis are more common manifestations of this disease. As with the congenital form, in acquired disease keratitis is most typically a late manifestation, occurring years after the original infection.

Clinical Picture. Congenital syphilitic keratitis is usually bilateral, although both eyes may not be affected at the same time. In congenital syphilis Hutchinson's triad is observed — keratitis, deafness, and barrel teeth. If the syphilitic interstitial keratitis is acquired, it is more likely to affect only one eye.

In both congenital and acquired disease, initial manifestations include a deep focal inflammation of the stroma, typically starting in the periphery. Patients usually complain of pain, eye redness, photophobia, tearing, itching. This *phase of infiltration* usually lasts about one month.

Later in the process of the disease (the *phase of vascularization*) deep stromal vascularization develops. The surrounding inflammatory infiltrate and stromal edema obscure the outline of the vessels, making the stroma appear pink ("salmon patch"). In the inflammation, the iris and ciliary body are involved with keratic precipitates formation. This phase can last up to two months.

Late stage of the disease (the *phase of regression*) is regressive and includes gradual clearing of the cornea from the periphery towards the centre, the stromal vessels begin to recede. This phase of the disease may last from several months to one year. Resolution of the lesions leaves corneal thinning, empty and ghost vessels, stromal opacification, and corneal scarring.

Complaints. Common symptoms are redness of the eye, pain, photophobia, tearing, and gradual loss of vision.

Signs. According to the disease progression the signs may include:

— ciliary injection;

— milky grey areas of stromal opacification that start with a broad base at the limbus and extend towards the corneal centre;
— keratic precipitates;
— stromal edema;
— stromal vascularization;
— "salmon patch" appearance;
— clearing of the vessels that begins at the limbus;
— ghost vessels;
— stromal thinning;
— stromal scarring.

Methods of Examination. Medical history, VA test, slit-lamp examination, confocal microscopy, anterior segment OCT, ultrasound biomicroscopy, tonometry, ophthalmoscopy, serological tests (rapid plasmin reagent test and fluorescent treponema-specific antibody test).

Differential Diagnosis. Tuberculous interstitial keratitis, interstitial keratitis caused by other types of viral and bacterial infection, Cogan syndrome.

Treatment. The treatment for syphilitic interstitial keratitis includes systemic treatment of the underlying disease — syphilis — and topical treatment for keratitis. Both systemic and local treatment should be started in the initial stage of the disease to obtain better results.

Systemic treatment does not treat interstitial keratitis, but is used to treat systemic infection and prevent neurosyphilis. It includes intramuscular or intravenous penicillin G for 10—14 days. In patients with penicillin allergy, several therapeutic alternatives are available, including Tetracycline, Doxycycline, Chloramphenicol, Ceftriaxone and macrolide antibiotics. For children Benzathine penicillin or Penicillin G is used for 10 days.

Systemic corticosteroids (Prednisone) can be added in severe and refractory cases of keratitis.

Local treatment involves instillations of corticosteroid drops (e.g. Dexamethasone 0.1 %, Prednisolone acetate 1 %) every 2—3 hours. As the condition is allergic by origin, corneal clearing occurs with steroids if started well in time and a useful vision is obtained. In cases of anterior uveitis development — cycloplegic drugs are useful (Atropine 1 %). Dark glasses can be used for photophobia.

Sometimes, in persistent corneal opacity and scarring it is necessary to resort to surgical treatment with keratoplasty.

Prognosis. With prompt and adequate treatment of the disease at the initial stage the cornea clears up with recovery of vision. Severe cases with scar formation may need penetrating keratoplasty to obtain good vision.

Complications. Recurrent inflammations, corneal opacity and/or scarring, cataract, secondary glaucoma, decrease of vision.

Prophylaxis. For prophylaxis of the congenital infection antisyphilitic therapy of the mother during the first months of pregnancy. Prevention also consists of avoiding the infection and its thorough treatment and follow-up.

Tuberculous Interstitial Keratitis

Definition. Tuberculous interstitial keratitis is a nonsuppurative inflammation of the corneal stroma associated with tuberculosis infection.

Etiology. Tuberculous interstitial keratitis is caused by an autoimmune reaction to tubercular antigen in aqueous humour occurring in patients with systemic tuberculosis. It is rather a sign of the systemic disease than a diagnosis.

Clinical Picture. Tuberculous interstitial keratitis is a rare and usually unilateral condition. It is more common in middle-aged people who have had previous tuberculosis.

Clinically it has a prolonged course with multiple attacks. The infiltration is usually peripheral and sectoral, sparing the central cornea. The superficial and middle stroma is usually affected. Long-standing disease results in stromal scarring with deep neovascularization or ghost vessels. Regression of the process is less rapid as in case of syphilitic interstitial keratitis, leaving a dense sector-like scar.

Tuberculous interstitial keratitis may be of four clinical forms — deep diffuse, deep focal, sclerosing and phlyctenular. It may be seen as an isolated finding or more often in association with scleritis and uveitis.

Complaints. Redness of the eye, irritation, tearing, foreign body sensation, photophobia, blepharospasm, and blurred vision. Other complaints may include headache, floaters, or flashes. Patients may also be asymptomatic.

Signs. Depending on the form of tuberculous interstitial keratitis the signs may include:

— ciliary injection;
— diffuse infiltration/s of the cornea in the peripheral inferior sector that manifests as a ring-shaped dense nodular opacity;
— stromal edema;
— deep vascularization;
— Descemet's membrane folds;
— keratic precipitates;
— constriction of the pupil;
— small infiltrates of 1 mm (phlyctenas) on the corneal surface;
— phlyctenular pannus;
— stromal opacity;
— stromal scarring.

Methods of Examination. Thorough medical history, VA test, slit-lamp exam, confocal microscopy, ophthalmoscopy, tonometry, anterior segment OCT, complete blood laboratory test, chest X-ray, tuberculin skin test.

Differential Diagnosis. Syphilitic interstitial keratitis, interstitial keratitis caused by other viral and bacterial infection, disciform keratitis, Cogan syndrome.

Treatment. Tuberculous interstitial keratitis requires first of all antituberculous therapy. Topical therapy alone is not curative and must be used only in conjunction with systemic therapy.

Anti-tuberculous treatment involves four-drug treatment with Isoniazid (5 mg/kg/day), Rifampicin (10 mg/kg/day), Pyrazinamide (20—25 mg/kg/day) and Ethambutol (15 mg/kg/day) for two months, followed by a two-drug (rifampicin and isoniazid) regimen for four to seven months.

Topical steroids are used to suppress the disease process (e.g. Dexamethasone 0.1 %, Prednisolone acetate 1 %) ever 2—3 hours.

Cycloplegics are used to avoid the development of posterior synechiae and promote patient comfort.

When the vision is impaired due to stromal opacification and/or scarring, keratoplasty is necessary to restore vision.

Prognosis. Prompt and adequate treatment can preserve the clear cornea and good vision. Prognosis with prolonged duration is questionable to maintain visual function.

Complications. Cataract formation, secondary glaucoma, corneal opacification, corneal scarring, blurry vision, decreased vision.

Prophylaxis. Primary prevention for tuberculous uveitis involves prevention of exposure to actively infected individuals in order to prevent systemic infection. If a patient got infected, prompt and thorough treatment and follow-up.

4.4. Degenerative Disorders of the Cornea

Degeneration of the cornea is gradual deterioration of its tissue characterized by depositions, thinning or vascularization of the corneal tissue. Corneal degenerations are not inherited, they result from the ageing process, local or systemic diseases. Moreover, continuous exposure to environmental conditions, UV stimulation, and in some cases, oxidative stress might be responsible for the onset of degenerative processes in the cornea.

Corneal degeneration commonly presents at middle to older age, and they may be unilateral or bilateral, or even asymmetric, often involve the peripheral cornea and may overlap the limbus and conjunctiva.

Arcus Senilis

Definition. Arcus senilis, or corneal arcus, is an age-related corneal disorder characterized by deposition of lipids and cholesterol at the periphery of the cornea that appears as an opaque, white arc or circular ring.

Etiology. The cause of arcus senilis is deposition of lipids and cholesterol crystals and connective tissue degeneration as a result of fairly normal age-related changes in the eyes. Current studies revealed that these changes may develop from atherosclerosis of the conjunctival vessels and disorders of lipid metabolism. However, the exact cause is not known.

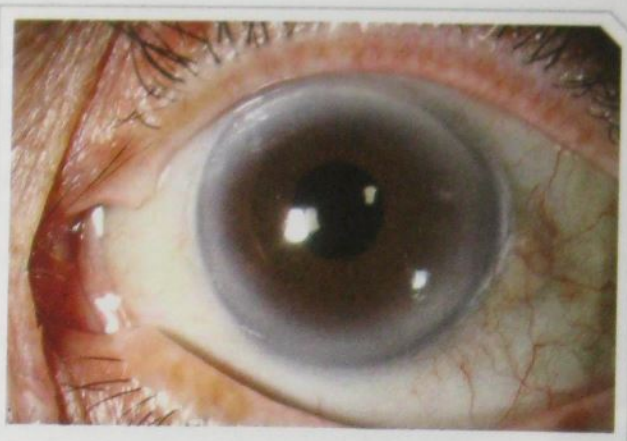


Fig. 8.9. Corneal arcus (from http://www.eyerounds.org)

In patients under 40 years of age it may indicate hyperlipidemia, hypercholesterolemia, or hyperlipoproteinemia and can be a prognostic factor for coronary artery disease in this age group.

Clinical Picture. Arcus senilis is a bilateral condition manifesting as a white ring in the periphery of the cornea. Initially the depositions appear at 6 or 12 o'clock on the corneal periphery (top and bottom of the cornea) and slowly spread to form a complete ring about 1 mm wide (fig. 8.9).

Clinical presentation is asymptomatic, since arcus senilis does not produce any symptoms, nor does it cause any visual disturbances. If there are symptoms, they are not related to corneal arcus.

Complaints. Usually patients do not have complaints related to arcus senilis, except cosmetic appearance of the obvious opaque ring at the periphery of the cornea.

Signs. Lipid deposits or crystals of cholesterol at the level of the stroma that appears as a peripheral annular hazy white arc or ring about 1—2 mm of width and with a clear space between it and the limbus (the lucid interval of Vogt), which is considered one of the hallmarks of this condition.

The arc or ring has a more clearly defined peripheral border and a more diffuse inner border. The deposits are more concentrated in the deeper stroma near Descemet's membrane but in more advanced cases occur in all layers of the stroma.

Additional findings during the examination may include lower central corneal thickness and constriction of the pupils.

Methods of Examination. Slit-lamp exam, blood tests for cholesterol and lipid levels to rule out hypercholesterolemia.

Treatment. Arcus senilis does not require any treatment, especially if it is present in elderly individuals. However, if the person with corneal arcus is under the age of 40, tests for cholesterol and triglyceride should always be performed to diagnose hypercholesterolemia and administer appropriate treatment.

Prognosis. Arcus senilis is a normal ageing process. If it is found in a person under 40 years of age, it may be an indication of an increased risk of coronary artery disease due to atherosclerosis.

Complications. There are no specific complications related directly to arcus senilis.

Prophylaxis. Effective prevention is not currently available, however, controlling the cholesterol levels, regular exercises, and balanced diet are of a great importance.

4.5. Ectatic Conditions

Keratoconus

Definition. Keratoconus is a progressive degenerative condition, in which the cornea thins and slowly protrudes changing its normal round shape to a cone, whichcauses distortion of vision (fig. 8.10). It is also known as conical cornea.

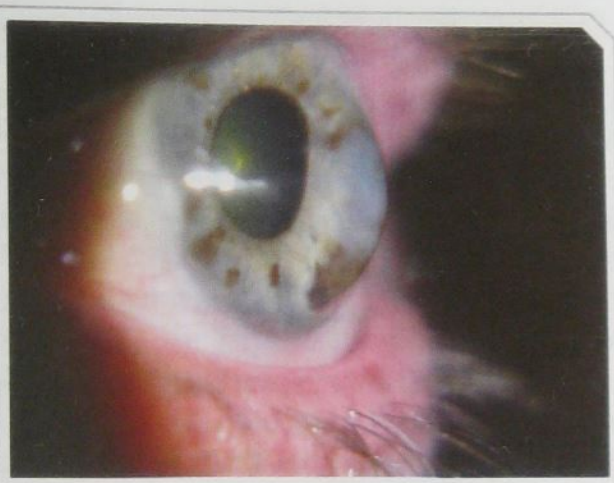


Fig. 8.10. Keratoconus

Etiology. Keratoconus is not caused by inflammation or infection. Its cause is unknown, although experts have speculated on a variety of reasons, including previous medical conditions, heredity, collagen defect, eye allergies, and rubbing the eyes.

Keratoconus is most commonly an isolated disorder, although it may be associated with Down syndrome, Leber's congenital amaurosis, and mitral valve prolapse.

Clinical Picture. Keratoconus is a gradually progressive disease that usually begins during late teens or early twenties and slowly progresses for 10 years or longer and then stabilizes. It is a bilateral condition, but usually one eye is more severely affected than the other.

Ocular signs and symptoms depend on the severity of the condition. It starts from gradual thinning of the central cornea that involves all its layers and remains unnoticed unless corneal topography is performed. By the time the cornea weakens and begins to bulge forward in a cone-like shape. This distortion of the corneal surface results in high myopia and irregular astigmatism with associated reduction of vision that cannot be compensated with glasses. As the condition progresses, the corneal thickness gradually decreases and patients notice acute worsening of the symptoms — photophobia, distortions, double vision, multiple "ghost images". When corneal hydrops develops, eye pain and redness can occur, scarring of the cornea from hydrops can cause decreased vision.

Complaints. In the early stages of keratoconus patients complain of blurring of vision, often worse at night. Over time they notice sensitivity to light, irritation, double vision when looking with just one eye, seeing triple ghost images, distortion, halos, and glare.

Signs. According to the stage of the condition signs may be as following:

— high, irregular myopic astigmatism on exam;

— corneal thinning;
— ectatic cone-shaped protrusion of the cornea typically just below the central visual axis;
— an iron-colored ring surrounding the cone (Fleischer's ring);
— striae in the posterior stroma anteriorly to Descemet's membrane (Vogt's striae);
— scarring at the apex of the cone (apical scarring);
— V-shape deformation of the lower eyelid on looking down (Munson's sign);
— abnormal red reflex on ophthalmoscopy (oil droplet reflex);
— inferior or central steepening (red area) on corneal topography.

Methods of Examination. VA, refractometry, skiascopy, slit-lamp exam, ophthalmoscopy, ultrasound pachymetry, keratometry, corneal topography, computerized corneal tomography.

Differential Diagnosis. Interstitial keratitis, pellucid marginal degeneration, keratoglobus.

Treatment. Treatment for keratoconus depends on the severity of the condition. Mild to moderate keratoconus can be treated with eyeglasses or soft contact lenses. For most people, the cornea will become stable after a few years.

Progressive changes require other treatment options, such as rigid gas permeable (RGP) contact lenses, which can reshape the corneal surface to hide or mask the underlying cone-shaped defect of the cornea. In some individuals, these contact lenses may become uncomfortable and are not tolerated well. In such cases, individuals can use "piggyback" contacts lenses, in which hard contact lenses are placed over soft lenses.

In more severe conditions surgery may be required such as implantation of intracorneal ring segments, collagen cross-linking or corneal transplantation (keratoplasty).

Corneal cross-linking (CXL) is the newest method in keratoconus treatment. It involves treating the cornea with riboflavin and then activating its collagen cross-linking properties with ultraviolet light. CXL increases linking between the collagen fibres, which improves the strength of the cornea and often causes some corneal flattening.

Prognosis. Overall the prognosis is favorable. In most cases vision can be corrected with rigid gas-permeable contact lenses. If corneal transplantation is needed, results are usually good. The recovery period can be long, and patients may often still need contact lenses. Recurrence of keratoconus in the transplanted cornea can occur, but is very rare.

Complications. Corneal hydrops, opacity, scarring, perforation, loss of vision, contact lens-related secondary giant papillary conjunctivitis, ocular discomfort or pain.

Prophylaxis. There are no preventive methods for keratoconus, but there are some measures, which may help to control allergies, to avoid eye rubbing, to prevent eye injures by wearing protective glasses, to be careful with contact lens use, and to undergo regular eye examinations that can help identify keratoconus early, and address any complications of the disease before they become visually disabling.

Keratoglobus

Definition. Keratoglobus is an ectatic degenerative disorder of the cornea characterized by generalized thinning and globular protrusion of the entire cornea.

Etiology. Keratoglobus has been described as both an acquired and a congenital disease and may be associated with various other ocular and systemic syndromes including connective tissue disorders and genetic predisposition.

The congenital form is present at birth associated with Leber's congenital amaurosis, Ehler—Danlos type VI, Marfan syndrome, and blue sclera syndrome. Some concomitant abnormalities reported with keratoglobus include joint hypermobility, dental and skeletal abnormalities, osteal fragility, and deafness.

The acquired form develops in adulthood and may present as end-stage advanced keratoconus. This form of keratoglobus may be associated with chronic marginal blepharitis, vernal keratoconjunctivitis, thyroid ophthalmopathy. Eye rubbing has been proposed to be the major contributing factor in keratoglobus development.

Clinical Picture. Keratoglobus is a rare, non-progressive or minimally progressive bilateral condition, which results in high myopia, irregular astigmatism, scarring, and rarely spontaneous globe rupture.

Clinical presentation is characterized by a generalized corneal bulging secondary to thinning of the corneal stroma and absence of Bowman's membrane. The thinning is greatest in the periphery, which creates an effect of a very flat central cornea and a very steep peripheral cornea.

The cornea is usually clear, although edema may result from a progressive break in Descemet's membrane. The corneal diameter is normal. The cornea may be particularly susceptible to rupture from even minor trauma.

Complaints. The most common complaints are decreased visual acuity, blurring of vision, image distortion, single or multiple episodes of eye pain followed by resolution and decreased visual acuity, photophobia, blepharospasm, epiphora.

Signs:

— high myopia as high as 50—60 diopters;
— irregular astigmatism;
— marked globular protrusion of the cornea;
— generalized thinning of the corneal stroma, most severe peripherally;
— absence of Bowman's membrane;
— folds or breaks of Descemet's membrane;
— irregular retinoscopic reflex;
— the cornea is transparent;
— the corneal diameter is normal or slightly increased;
— deep anterior chamber;
— IOP is normal;
— possible scarring of the stroma;
— common hydrops;
— occasional corneal rupture.

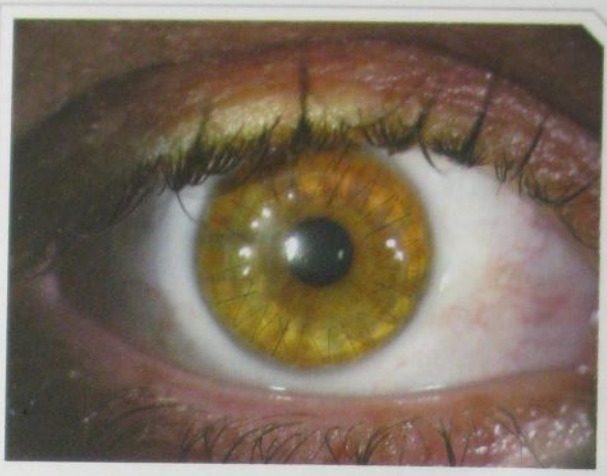


Fig. 8.11. Keratoplasty

Methods of Examination. VA test, refractometry, slit-lamp exam, corneal size measurement, keratometry, keratotopography, pachymetry, OCT, ophthalmoscopy, tonometry.

Differential Diagnosis. Megalocornea, congenital glaucoma, keratoconus, corneal ectasia following refractive surgery, pellucid marginal degeneration.

Treatment. At the early stages keratoglobus is treated with spectacle and contact lens correction of high myopia with or without astigmatism that improves visual acuity and provides protection from ocular trauma. According to the disease progression contact lenses can be soft, rigid gas permeable, hybrid, or scleral.

Further development of the disorder usually leads to a need for corneal transplantation because of extreme thinning of the cornea. Lamellar epikeratoplasty has been used successfully to reinforce thin corneas and, in some cases, to improve vision. It involves partial-thickness excision of the corneal epithelium and stroma so that the deep stroma and endothelium are preserved and replacing them with a donor graft of unaffected corneal tissue.

For acquired keratoglobus, treatment of the associated condition must be initial. For advanced cases, large penetrating keratoplasty (fig. 8.11) may be successful. However, this operation is quite complicated, as there may be some difficulties due to heavy thinning and peripherally damaged cornea.

Prognosis for keratoglobus is poor. Spectacle correction often results in suboptimal BCVA. In addition, surgical correction is difficult and associated with complications.

Complications. Corneal hydrops, corneal perforation and rupture.

Prophylaxis. There are no effective methods of preventing the formation of keratoglobus. However, it is recommended to undergo regular eye exams to be able to diagnose the disease at an early stage of development and start immediate treatment.

4.6. Dystrophies of the Cornea

Corneal dystrophies are a group of rare inherited non-inflammatory opacifying disorders that are caused by progressive accumulation of deposits within the layers of the cornea. These deposits are not caused by inflammation, infection, systemic or local disease, or trauma, but by genetic mutations.

Dystrophies are usually bilateral, symmetric, avascular, have a central location and primarily involve a single corneal layer. They present occasionally at birth, but more usually develop during the first or second decade and sometimes even later in life.

Corneal dystrophies may be asymptomatic in some individuals, but in others they may cause significant vision impairment. The age of onset and specific symptoms vary among the different forms of corneal dystrophy. Treatment of corneal dystrophies depends on the type of the dystrophy and severity of vision impairment. Sometimes it does not require any treatment, in other cases treatment options may include lubricating eye drops, ointments, or special soft contact lenses. In more severe cases laser treatment, cornea scraping or keratoplasty may be necessary.

There are many types of corneal dystrophies, and they are traditionally classified according to the corneal layer affected:

— *anterior dystrophies* (primarily affecting epithelium and Bowman's membrane) — epithelial basement membrane dystrophy, Meesmann dystrophy, gelatinous drop-like corneal dystrophy, Reis—Bücklers' dystrophy;
— *stromal dystrophies* — lattice dystrophy, Schnyder corneal dystrophy, macular corneal dystrophy, granular corneal dystrophy, etc.;
— *posterior dystrophies* (primarily affecting the endothelium and Descemet's membrane) — Fuchs' endothelial dystrophy, posterior polymorphous dystrophy, congenital hereditary endothelial dystrophy.

Some of the most common corneal dystrophies include epithelial basement membrane dystrophy, lattice dystrophy, and Fuchs' dystrophy.

Epithelial Basement Membrane Dystrophy

Definition. Epithelial basement membrane dystrophy (EBMD) is one of the most common forms of genetically transmitted corneal dystrophy that appears as gray patches (maps), creamy white cysts (dots), or fine linear opacities (fingerprints) in the corneal epithelium. It is also known as map-dot-fingerprint dystrophy, or Cogan's microcystic dystrophy.

Etiology. EBMD is not a true dystrophy, this disorder results from abnormal attachment between the epithelium and Bowman's membrane or its thickening, which may cause persistent epithelial defects and recurrent erosions. The corneal dots are intraepithelial microcysts that contain nuclear, cytoplasmic, and lipid debris. The corneal fingerprints are curvilinear clusters of reduplicated and thickened basement membrane.

Clinical Picture. EBMD is characterized by either geographic epithelial changes called maps, opaque irregular opacities called dots, or concentric irregular lines referred to as fingerprints. If corneal changes are mild, the patient is asymptomatic. Occasionally they may be more severe and cause an irregular surface resulting in decreased visual acuity.

Corneal erosions can be a chronic problem and along with altering the cornea's curvature may also expose the nerve endings resulting in moderate to severe pain.

Generally, the pain will be worse on awakening in the morning, or during the night. Other symptoms include sensitivity to light, excessive tearing, and foreign body sensation in the eye.

EBMD usually affects adults between the ages of 40 to 70, although it can develop earlier in life. EBMD tends to occur in both eyes, although it is often asymmetric.

Complaints. Most cases are asymptomatic, although symptoms may include blurry vision, fluctuating vision, flare, or pain, watering, photophobia and blepharospasm associated with recurrent corneal erosions.

Signs. The clinical signs typically include decreased VA, irregular astigmatism, a bilateral presentation of epithelial microcysts and whirling superficial defects, such as corneal ridges and opacities.

Methods of Examination. VA test, refractometry, slit-lamp examination, fluorescein staining, keratometry, corneal topography, ophthalmoscopy.

Differential diagnosis. Recurrent corneal erosion, Fuchs' endothelial dystrophy, corneal trauma with endothelial cell damage.

Treatment. There is no cure for EMBD, therefore the goal is to protect the corneal epithelium or lower the rate of corneal erosion recurrences. In mildly asymptomatic and moderate cases, lubrication with artificial tears, hypertonic saline drops or ointments, patching and therapeutic contact lenses are recommended. In more advanced cases of recurrent corneal erosions antibiotic ointments, topical steroids, and oral doxycycline are needed. Other types of treatment include anterior stromal puncturing (outside the visual axis), epithelial debridement, phototherapeutic keratectomy (PTK).

Prognosis. Most patients are able to maintain sufficient vision and comfort for reading, driving, and other visual tasks, except during episodes of corneal erosions. Other patients may have severe visual impairment due to recurrent corneal erosions.

Complications. Corneal scarring from recurrent erosions.

Prophylaxis. There is no real prevention for EBMD. If recurrent cornea erosions are present with EBMD, they may be prevented with prompt and adequate treatment.

Lattice Dystrophy

Definition. Lattice dystrophy is a most common hereditary stromal dystrophy characterized by accumulation of amyloid deposits or abnormal protein fibres throughout the corneal stroma.

Etiology. Lattice dystrophy is an autosomal dominant condition that results in collagen degeneration.

Clinical Picture. Lattice corneal dystrophy is characterized by the development of lesions that form branching lines resembling cracked glass or lattice. Both corneas are usually symmetrically involved.

Two main types of lattice dystrophies have been identified. In the most common form, lattice corneal dystrophy type I, the refractile lines are more promi-

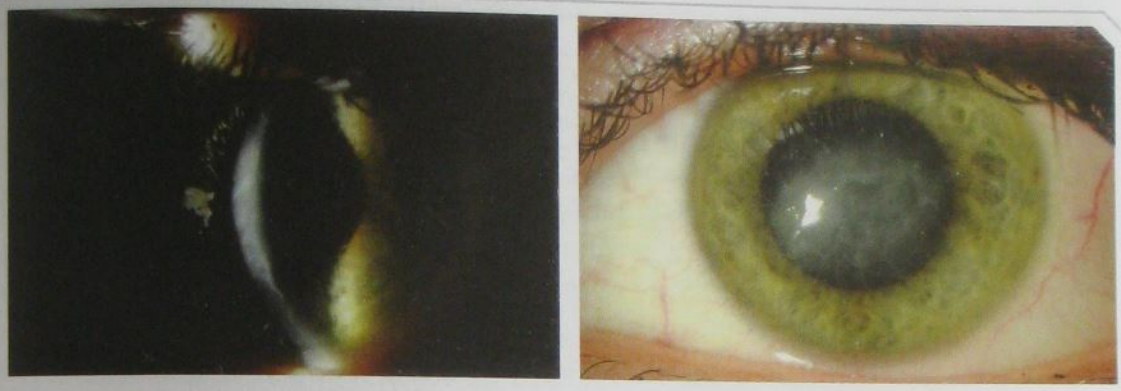


Fig. 8.12. Lattice corneal dystrophy (mild and progressive form)

nent in the central cornea than on the periphery. This is a progressive disorder, in which vision during childhood is often normal but by the fifth and sixth decades most patients have severe visual impairment due to increasing amyloid accumulation and involving more of the stromal tissue. Corneal erosions may occur due to accumulation of abnormal protein fibres under the epithelium causing severe pain.

Type 2 of lattice dystrophy is associated with systemic amyloidosis, ear abnormalities, cranial and peripheral nerve palsies, dry and lax skin.

Complaints. Usually patients remain asymptomatic for years. As the disease progresses, symptoms include blurred and reduced vision, pain, tearing, photophobia from recurrent corneal erosions.

Signs. Decreased visual acuity, irregular astigmatism, deposits in the stroma in the form of clear, comma-shaped overlapping dots and branching filaments that create a "lattice" effect; they become opaque with time and involve more of the stroma (fig. 8.12). In some people — accumulation of abnormal protein fibres under the epithelium.

Methods of Examination. VA test, refractometry, slit-lamp examination, fluorescein staining, keratometry, corneal topography, ophthalmoscopy.

Differential Diagnosis. Recurrent corneal erosion, macular corneal dystrophy, granular corneal dystrophy.

Treatment is symptomatic and depends on the visual impairment of the patient and discomfort. Recurrent erosions are treated with hypertonic agents, artificial tears, or therapeutic contact lenses. Excimer laser PTK is required as an optional treatment for recurrent erosions and superficial opacities. More serious cases with severe visual loss may require penetrating keratoplasty.

Prognosis is good, although protein deposits can develop on the transplanted corneal graft.

Complications. Recurrent corneal erosions, corneal opacity, reduced vision.

Prophylaxis. The disease has no effective preventive measures, early detection and treatment is the key to prevention of its complications.

Fuchs' Dystrophy

Definition. Fuchs' dystrophy is a corneal dystrophy characterized by progressive loss of endothelial cells resulting in corneal edema and decreased vision.

Etiology. Fuchs' corneal dystrophy is a genetic, autosomal dominant disease with high penetrance. However, the condition may also occur in persons without a known family history of the disease due to genetic mutations.

Normally, the endothelial cells are responsible for maintaining the proper amount of fluid in the cornea that keeps it transparent. But with Fuchs' dystrophy, as more and more cells are lost, fluid begins to accumulate in the cornea causing swelling and corneal opacity.

Clinical Picture. Fuchs' dystrophy is a bilateral, asymmetrical condition more frequently developing in women than in men. Vision problems usually do not appear before age 50, although signs of the disease may be seen at ocular examination at an earlier age, usually in the 30s and 40s.

In the early stages a patient with Fuchs' dystrophy notices the appearance of glare and an increased sensitivity to light. As the condition progresses, the patient will wake up with cloudy vision that will gradually clear during the day. This occurs because the cornea is normally thicker in the mornings as it retains fluids during sleep when the eyes are closed. Once the eyes are opened throughout the day, evaporation reduces water content and the thickness of the cornea, allowing for clearing of vision.

In later stages, as the dystrophy worsens, evaporation is not enough to remove accumulated water in the cornea, and swelling and blurred vision last all day.

As the disease gets worse, epithelial swelling may form small "blisters" on the corneal surface, which get bigger and eventually break, causing extreme eye pain. Fuchs' dystrophy can also cause the curvature of the cornea to change, leading to more vision problems (fig. 8.13).

Complaints. Glare and sensitivity to light, eye pain, blurred vision, at first only in the mornings, distorted vision, decreased night vision, seeing colored haloes around lights.

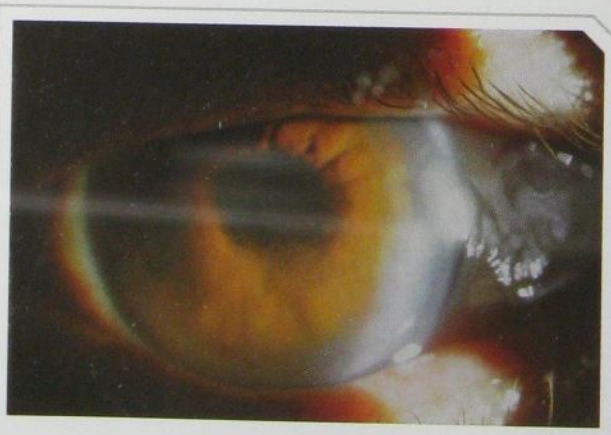


Fig. 8.13. Endothelial dystrophy of the cornea

Signs. Presence of guttae (collagen deposits on Descemet's membrane thought to be secreted by abnormal endothelial cells) which have "beaten metal" appearance at the slit-lamp, stromal edema, cystic epithelial edema, epithelial bullae (blisters), subepithelial scarring, low endothelial cell counts on specular microscopy.

Methods of Examination. VA test, glare test, slit-lamp exam, ultrasound, pachymetry, confocal microscopy, specular microscopy, OCT, tonometry.

Differential Diagnosis. Bullous keratopathy, congenital hereditary endothelial dystrophy, posterior polymorphous dystrophy, corneal edema, corneal erosion.

Treatment. Fuchs' dystrophy cannot be cured as it is impossible to make the endothelial cells work better or become more numerous. However, the symptoms can be controlled with medication.

NOTE!

Cataract surgery can worsen Fuchs' dystrophy because of damage to the endothelial cell layer. For this reason, patients with cataracts and Fuchs' dystrophy requiring surgical intervention are often recommended to undergo cataract surgery before or at the same time as corneal transplantation to ensure the best outcome for the transplant.

At early stages treatment involves instillation of hypertonic solutions and/or ointments to dehydrate the epithelium. With extreme epithelial edema, pain, and photophobia therapeutic soft contact lenses may provide temporary relief. If VA is markedly reduced, penetrating keratoplasty, deep lamellar endothelial keratoplasty or Descemet's membrane endothelial keratoplasty are the treatment of choice.

Prognosis. Fuchs' dystrophy is a progressive disorder, and left untreated it may result in corneal blindness and severe pain. With corneal transplant surgery, the prognosis is good, though there is a possibility that the transplant may be rejected or astigmatism may develop.

Complications. Cataract, glaucoma, corneal blindness is left untreated, epithelial healing problems and ulceration, graft rejection and failure.

Prophylaxis. There is no known prevention, but awareness of the condition and taking certain basic steps to prevent it from deterioration may help.

Review:

1. Key Points

Diseases of the cornea according to the origin may be *congenital* or *acquired*; according to the clinical presentation — *mild, moderate* or *severe*; according to the onset of the pathological process — *acute, chronic* or *recurrent*; according to the location — *central* or *peripheral*; according to the depth — *superficial, deep, impending perforation, perforated*; according to the etiology — *exogenous or endogenous*; according to the pathogenesis — *inflammatory, degenerative, dystrophic, ectatic conditions, abnormalities in corneal transparency, trauma, tumor*. The main symptoms of corneal diseases (corneal syndrome) are eye pain, photophobia, tearing, blepharospasm, foreign body sensation. Other symptoms are blurry vision, refractive anomalies, curvature disorders. The examination consists of corneal examination by side illumination, slit-lamp examination, corneal sensitivity, corneal size measurement, dye staining of the cornea, pachymetry, keratometry, corneal topography, microbial investigation.

Congenital anomalies are megalocornea, microcornea, congenital cloudy cornea, etc. The main symptoms are abnormal size or transparency of the cornea. If the condition does not influence the vision, it requires no treatment, otherwise — surgical reconstruction.

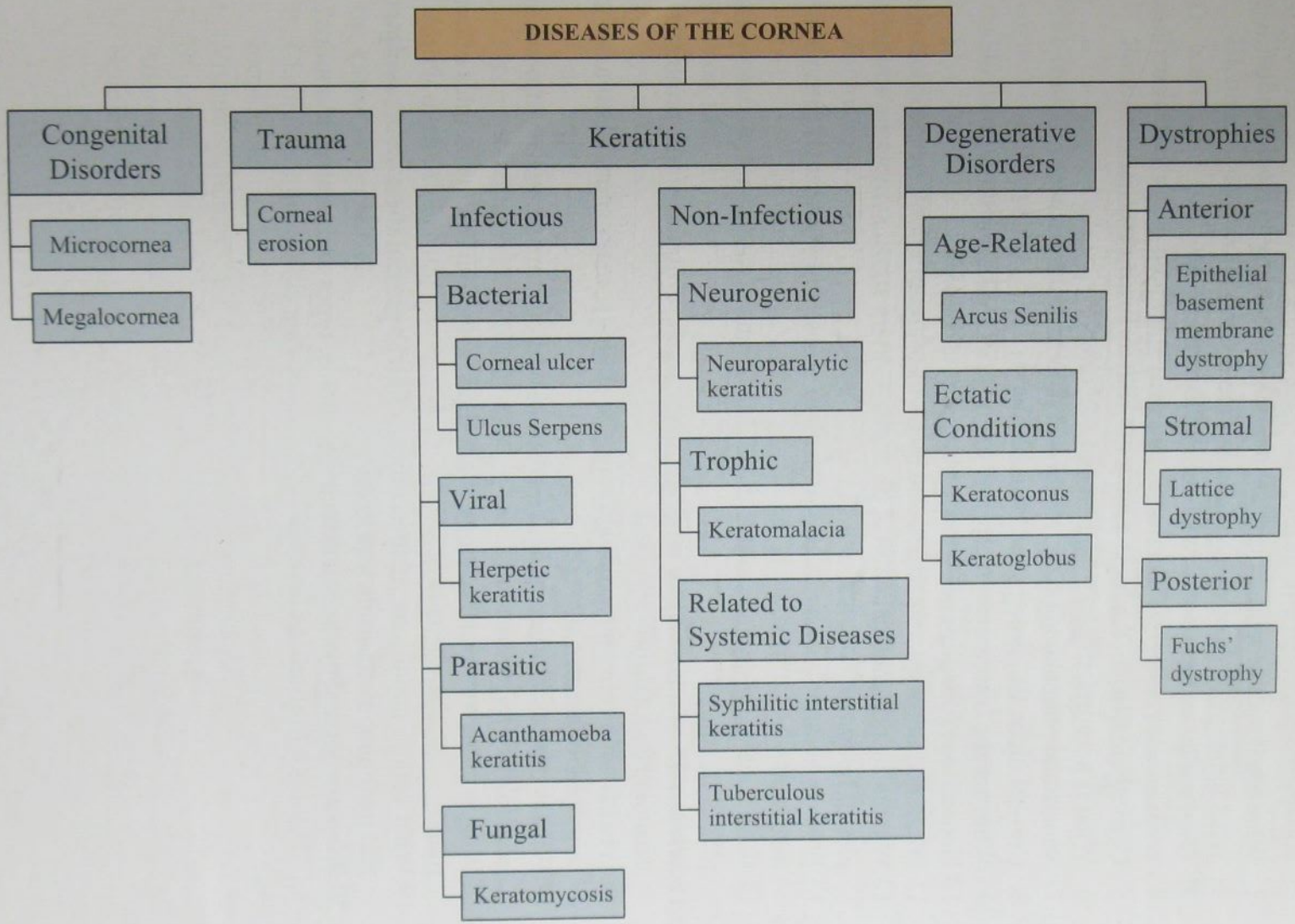
Inflammatory diseases may be exogenous or endogenous, according to the pathogenesis — infectious and non-infectious. Infectious keratitis can be caused by bacteria, viruses, parasites, fungi. Non-infectious keratitis may be allergic, neurogenic, trophic, related to systemic diseases. The symptoms and methods of treatment differ according to the origin of the pathology. Topical treatment may include antibiotics, antivirals, antifungals, corticosteroids, cycloplegics, artificial tears.

Degenerative disorders include arcus senilis, which is age-related degeneration and does not require any treatment.

Ectatic conditions are keratoconus and keratoglobus associated with thinning of the cornea and characterized by its protrusion. Along with severe refractive disorders, they can cause corneal scarring and rupture. Treatment includes therapeutic corneal lenses, lamellar epikeratoplasty, keratoplasty.

Dystrophies of the cornea are inherited progressive disorders which involve loss of corneal transparency. Visual impairment and treatment methods depend on the type and severity of the condition and may include lubricating eye drops, ointments, laser treatment, cornea scraping or keratoplasty.

2. Diagrams


DISEASES OF THE CORNEA
Congenital Disorders
Microcornea
Megalocornea
Trauma
Corneal erosion
Keratitis
Infectious
Bacterial
Corneal ulcer
Ulcus Serpens
Viral
Herpetic keratitis
Parasitic
Acanthamoeba keratitis
Fungal
Keratomycosis
Non-Infectious
Neurogenic
Neuroparalytic keratitis
Trophic
Keratomalacia
Related to Systemic Diseases
Syphilitic interstitial keratitis
Tuberculous interstitial keratitis
Degenerative Disorders
Age-Related
Arcus Senilis
Ectatic Conditions
Keratoconus
Keratoglobus
Dystrophies
Anterior
Epithelial basement membrane dystrophy
Stromal
Lattice dystrophy
Posterior
Fuchs' dystrophy

3. Review Questions

A. Control Questions

1. What are the main types of corneal disorders according to the classification?
2. What main methods of cornea examination do you know?
3. What are the main symptoms of corneal disorders?
4. What corneal congenital anomalies and their treatment options do you know?
5. Corneal erosion, its etiology, symptoms, and treatment methods.
6. What is keratitis, its classification and outcomes?
7. Superficial punctate keratitis, its clinical picture and treatment methods.
8. Corneal ulcer, its etiology and clinical course.
9. What are the complaints, signs, and methods of treatment for corneal ulcer?
10. Herpetic keratitis, its etiology and clinical presentation.
11. What are the complaints, signs, and methods of treatment for herpetic keratitis?
12. What is acanthamoeba keratitis, its etiology, main signs, symptoms and principles of treatment?
13. What is keratomycosis, its etiology, main signs, symptoms and principles of treatment?
14. What is neuroparalytic keratitis, its etiology, main signs, symptoms and principles of treatment?
15. What is keratomalacia, its etiology, main signs, symptoms and principles of treatment?
16. What is syphilitic interstitial keratitis, its etiology, main signs, symptoms and principles of treatment?
17. What is tuberculous interstitial keratitis, its etiology, main signs, symptoms and principles of treatment?
18. What is arcus senilis, its etiology, main signs, symptoms and principles of treatment?
19. What ectatic conditions of the cornea, clinical presentation, main symptoms and treatment methods do you know?
20. What are corneal dystrophies, their symptoms, types and treatment options?

B. Tests

1. **Symptoms of corneal diseases are:**
   A. Eye pain
   B. Photophobia
   C. Blepharospasm
   D. Tearing
   E. Discharge
   F. Foreign body sensation

2. **The main diagnostic methods of cornea conditions are:**
   A. Slit-lamp examination
   B. Dye staining
   C. Corneal sensitivity
   D. Keratometry
   E. Pachymetry
   F. Corneal topography

3. **The main symptoms of corneal erosion are:**
   A. Bilateral onset
   B. Eye redness
   C. Corneal syndrome
   D. Discharge
   E. Blurred vision
   F. Open sores/blisters around the eye

4. **Which of the following pathological conditions are NOT the cause of keratitis?**
   A. Corneal trauma
   B. Chronic conjunctivitis and/or blepharitis
   C. Vitamin deficiency
   D. Systemic diseases
   E. Allergy
   F. Ectatic conditions

5. **What are the main ophthalmic findings in corneal ulcer?**
   A. Conjunctival hyperemia
   B. Ciliary congestion
   C. A grey-white or yellow-grey disc on the transparent cornea
   D. Corneal edema
   E. Discharge
   F. Hypopyon

6. **The main clinical presentations of herpetic simplex keratitis are:**
   A. Punctate superficial keratitis
   B. Dendritic keratitis
   C. Geographic keratitis
   D. Stromal keratitis
   E. Endotheliitis
   F. Neurotrophic keratitis

7. **The main methods of treatment for herpetic simplex keratitis are:**
   A. Topical anti-viral drugs
   B. Systemic immunotherapy
   C. Topical antibiotics
   D. Anesthetics
   E. Artificial tears
   F. Corticosteroids

8. **Topical medications contraindicated in the treatment of corneal epithelium damage:**
   A. Anesthetics
   B. Lubricants
   C. Antibiotics
   D. Corticosteroids
   E. Antifungals
   F. Antivirals

9. What are the most common corneal complications associated with contact lens wearing?

A. Corneal erosion
B. Corneal ulcer
C. Acanthamoeba keratitis
D. Corneal edema
E. Neovascularization
F. Epithelial basement membrane dystrophy

10. What are the treatment methods of interstitial keratitis associated with systemic diseases?

A. Systemic treatment of the underlying disease
B. Topical corticosteroids
C. Systemic corticosteroids
D. Cycloplegics
E. Topical antibiotics
F. Lubricants

11. Degenerative disorders of the cornea are:

A. Arcus senilis
B. Keratoconus
C. Keratoglobus
D. Megalocornea
E. Microcornea
F. Nubecula

12. Surgical methods used in corneal disease treatment are:

A. Epithelial debridement
B. Lamellar epikeratoplasty
C. Lamellar endothelial keratoplasty
D. Descemet's membrane endothelial keratoplasty
E. Penetrating keratoplasty
F. Phacoemulsification

C. Clinical Cases

Case 1

A 19-year-old female patient presented with a complaint of right eye pain. This patient states she was applying makeup and accidentally scratched the eye with an applicator. Her other complaints were blurry vision, foreign body sensation, photophobia, tearing. This patient's vital signs are all within normal limits. Ocular examination: it is painful for the patient to open the eye, conjunctival injection, tearing with no purulent discharge, pieces of mascara under the upper eyelid. Examination with fluorescein stain showed light epithelial defect of the cornea. What are the diagnosis and treatment options?

Case 2

A 23-year-old male patient presented with bilateral photophobia, foreign body sensation, and tearing that is worse in the left eye. He reports a decade-long history of similar relapsing and remitting symptoms in both eyes, but not at the same time. He does not wear contact lenses. Objectively: no redness or discharge, multiple distinct small stellate granular superficial opacities seen without and with fluorescein staining, the stroma is clear and compact. The anterior chamber and fundus are normal. What are the diagnosis and treatment options?

Case 3

A 52-year-old woman presented with complaints of significant pain, ocular burning, foreign body sensation and significant decrease of vision in her left eye which started two days ago; after a few days when she accidentally slept with her soft contact lenses in. Examination: VA OD — 1 with correction, OS — counting fingers. Slit-lamp exam of the left eye revealed eyelid edema, conjunctival chemosis, a central epithelial defect of 2.2 mm × 1.9 mm below and temporally to the center of the cornea. At the base of the defect, there is a dense, white stromal abscess. The anterior chamber — no hypopyon, an infiltration of white blood cells in the anterior corneal stroma surrounding the abscess with a semilunar retrocorneal plaque, KPs, and flare. What are the diagnosis and treatment options?

Case 4

A 37-year-old woman presented to the hospital with severe pain, photophobia, and decreased vision in the right eye. She does not wear contact lenses and denies ocular trauma but is experiencing symptoms of cold. Ocular examination of OD: cold sores, eyelid edema, conjunctival injection, four small dendritic epithelial defects stained green with fluorescein in the visual axis, no basement membrane dystrophy, a clear and compact stroma without vascularization, the anterior chamber and fundus are normal. What are the diagnosis and treatment options?

Case 5

A 42-year-old man presented with severe pain, photophobia, blurring of vision, and haloes around light sources in the right eye. He had a prior history of HSV epithelial keratitis. OD examination: a deep disc-shaped stromal edema in the paracentral region with a clear demarcation between the involved and uninvolved cornea, the folds in Descemet's membrane, keratic precipitates, slightly elevated IOP. What are the diagnosis and treatment options?

Case 6

A 38-year-old man complains of cloudy vision, photophobia, tearing, red and extremely painful right eye. The patient has used disposable soft contact lenses for the past 3 months. OD examination: VA — counting fingers, conjunctival injection, irregular epithelium, punctate epithelial erosions, dendritic epithelial defects, stromal dense ring-shaped infiltrates. What are the diagnosis and treatment options?

Case 7

A 43-year-old woman presented with pain, photophobia, tearing, and decreased vision in the right eye following the entry of some dust particle two days ago when she was working in the garden. OD examination: VA — 0.1, conjunctival injection, a central corneal raised white-grey fluffy ulcer with rough dry texture and irregular feathery borders, diffuse corneal edema, stromal infiltrates with sharp margins, keratic precipitates, hypopyon. What are the diagnosis and treatment options?

Case 8

A 27-year-old man with a month-long history of corneal ulcer in the left eye presented with a decrease of vision and slight redness of his left eye without any irritation or discharge. Ocular examination of OD was normal. OS examination: VA — 0.5, paracentral corneal ulcer with undermined edges without conjunctival injection or aqueous flare, the cornea is insensitive to touch with a cotton tip applicator, the skin is insensitive to touch and pain stimuli in the region of the left trigeminal nerve. The ocular motility, pupil size, and pupil reflexes, the anterior chamber structures and the ocular fundus are normal. The central and peripheral visual fields are normal. What are the diagnosis and treatment options?

Case 9

A 35-year-old man with a history of syphilis acquired 10 years ago presented with decreased vision, eye redness, itching, pain, photophobia, floaters in his left eye lasting over one month. The same symptoms started in the right eye. Objectively: OD — ciliary injection, deep milky-grey opacification of the stroma near the limbus; OS — the same signs plus opacification extending towards the corneal centre, stromal edema, deep stromal vascularization, "salmon patch" appearance, keratic precipitates. What are the diagnosis and treatment options?

Case 10

A 72-year-old woman presented with a white ring in the periphery of the cornea in both eyes. Other ocular conditions were corresponding to her age. The eye exam showed a circular white-grey lipid deposition at the level of the stroma between 4 and 12 o'clock on the corneal periphery about 1 mm wide, the lucid interval of Vogt. She had been treated for chronic pancreatitis and hyperlipoproteinemia. What are the diagnosis and treatment options?

Case 11

A 24-year-old woman presented with complaints of gradual worsening of vision in both eyes for one year, vision is often worse at night, there are halos and distortions. Her BCVA was OD: 0.8 with correction $-6.75\ -6.50 \times 30°$; OS: 0.8 with correction $-6.50\ -7.50 \times 60°$. Examination: OU — high, irregular myopic astigmatism, OD — central corneal thinning, corneal hydrops, Descemet's membrane breaks; OS — prominent Fleischer's ring, Vogt's striae. Corneal topography revealed an irregular cornea of both eyes. What are the diagnosis and treatment options?

Case 12

A 35-year-old man presented with a complaint of blurry vision in both eyes that is worse in the mornings and improves during the day, glare and halos when looking at sources of lights. Examination: VA — 0.8 OU; the anterior chamber, fundus, IOP, and visual field are normal, corneal guttae ("beaten metal" appearance), low endothelial cell count. What are the diagnosis and treatment options?

CHAPTER 9

Diseases of the Sclera

OBJECTIVES

Upon completion of the chapter the students should be able to:
— know the main pathologies of the sclera and its classification;
— know the basic diagnostic methods of scleral examination;
— evaluate and manage the patients with scleral pathology;
— know the principles of scleral disease treatment.

Plan:

1. CLASSIFICATION OF SCLERAL DISEASES
2. SYMPTOMS OF SCLERAL DISEASES
3. EXAMINATION METHODS
4. DISEASES OF THE SCLERA
   - **4.1. Inflammation of the Sclera**
     - Episcleritis
     - Scleritis
   - **4.2. Staphyloma**
   - **4.3. Blue Sclera**
   - **4.4. Tumors of the Sclera**

1. Classification of Scleral Diseases

Scleral diseases can be classified:
— according to the origin — congenital or acquired;
— according to location — superficial or deep; anterior or posterior;
— according to the pathogenesis — inflammatory, degenerative, pigmentation defects, trauma, tumors;
— according to clinical presentation — localized (nodular) or extended (diffuse); non-necrotizing or necrotizing.

1.1. Congenital Disorders — blue sclera, melanosis oculi, ectasia, staphyloma.

1.2. Inflammatory Diseases
1.2.1. Episcleritis
— Simple (or diffuse)
— Nodular
1.2.2. Scleritis
— Anterior
— Non-necrotizing — diffuse or nodular
— Necrotizing — with inflammation or without inflammation
— Posterior

1.3. Degenerative Disorders
1.3.1. Staphyloma — anterior, intercalary, ciliary, equatorial, posterior
1.3.2. Ectasia

1.4. Tumors of the Sclera

2. Symptoms of Scleral Diseases

— Redness of the eye
— Mild to severe ocular pain
— Discoloration of the sclera
— Tenderness on palpation
— Painful eye movements
— Photophobia
— Tearing
— Decrease of vision

3. Examination Methods

— VA
— Slit-lamp exam
— Ophthalmoscopy
— B-scan ultrasonography
— CT scan

4. Diseases of the Sclera

Diseases of the sclera may include congenital, inflammatory, degenerative or pigmentation defects. Scleral disorders may be symptoms of certain systemic diseases or medical conditions, or occur as a result of medications or injury to the eye.

4.1. Inflammation of the Sclera

Inflammation of the sclera usually occurs in association with chronic infections (tuberculosis, syphilis, rheumatoid arthritis), systemic diseases (gout, psoriasis, rosacea), or as a reaction to endogenous toxins.

Episcleritis

Definition. Episcleritis is an inflammatory condition affecting the superficial layer of the sclera (the episclera, the thin vascular tissue that lies between the conjunctiva and the sclera).

Etiology. Most cases of episcleritis have no identifiable cause, although some of them may be associated with connective tissue and vascular diseases, systemic inflammatory conditions, allergy, infection, trauma or certain medications. These include rheumatoid arthritis, Crohn's disease, ulcerative colitis, psoriatic arthritis, systemic lupus erythematosus, ankylosing spondylitis, Reiter's syndrome, Wegener's granulomatosis, Behcet's disease, rosacea, gout, atopy, etc.

It is the most common form of scleral inflammation. Episcleritis usually occurs between the ages of forty and fifty years and affects women more often than men. Episcleritis is very rare in babies and children under the age of five.

Clinical Picture. Episcleritis is characterized by localized or diffuse redness (usually unilateral and in one quadrant of the globe) and varying degrees of discomfort. There are two types of episcleritis:

Simple (or diffuse) episcleritis is the most common condition that is characterized by diffuse inflammation.

Nodular episcleritis is localized inflammation with formation of a nodule, which can be moved slightly over the underlying sclera with a cotton-tipped applicator.

Complaints. Patients usually complain of acute or gradual onset of diffuse or localized eye redness. Some may not report any other symptoms, while others may complain of mild pain, burning, foreign body sensation, photophobia and tearing.

Signs. In diffuse episcleritis sectoral or diffuse redness, mild to moderate tenderness over the area of redness, and dilation of the large episcleral vessels are apparent (fig. 9.1). Nodular episcleritis is characterized by a pink or purple flat nodule near the limbus surrounded by injection. The nodule is firm, tender, covered by freely moving overlying conjunctiva.

Methods of Examination. Slit-lamp examination.

Differential Diagnoses. Scleritis, viral conjunctivitis, superior limbic keratoconjunctivitis.

Treatment. Most cases of episcleritis most often improve without treatment within two to three weeks. However, treatment may make symptoms resolve sooner.

Treatment of episcleritis usually involves the following:

- *in mild-to-moderate cases:*
  - — cold compresses 3 to 4 times per day for pain relief;

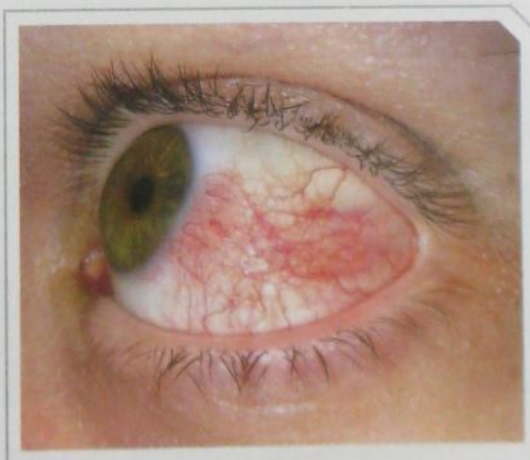


Fig. 9.1. Simple episcleritis

— topical lubricant eye drops (artificial tears) 2—3 times daily to relieve discomfort and irritation;
— topical corticosteroid eye drops (Dexamethasone 0.1 % eye drops 4 times a day);

- *in severe forms:*
  — stronger steroids (Hydrocortisone 2.5 % ointment, Prednisolone 1 % eye drops 4—6 times a day),
  — topical non-steroidal anti-inflammatory drugs (NSAIDs) (Diclofenac 0.1 % eye drops 3 times a day),
  — oral NSAIDs (Ibuprofen 400 mg three times daily or Indomethacin 50 mg twice daily) can be given.

Prognosis is favorable as episcleritis can run its course without treatment and also responds well to treatment.

Complications may be a recurrence of the condition, scleritis, or very rare corneal involvement.

Prophylaxis. There is no primary prevention for episcleritis, but treatment of underlying conditions may help.

Scleritis

Definition. Scleritis is an inflammation of the sclera proper. It is more severe and serious vision-threatening inflammation compared to episcleritis as it occurs throughout the entire thickness of the sclera. Scleritis is a rarer disease, it usually occurs in elderly patients, more often in females.

Etiology. Scleritis is often associated with an underlying connective tissue disorders in up to almost 50 % of patients (rheumatoid arthritis (this disorder is the most frequent), ankylosing spondylitis, lupus, relapsing polychondritis, polyarteritis nodosa, Reiter's syndrome, Wegener's granulomatosis, etc.). Some cases are caused by infection (tuberculosis, syphilis, streptococcal infection, pneumococcal pneumonia, herpes zoster virus, inflammation of the sinuses, corneal ulcer), trauma, may follow recent cataract surgery or retinal detachment surgery, or may be drug induced.

Clinical Picture. Scleritis is classified according to the anatomic location of the inflammation and to the clinical presentation of the disease. Clinical forms of scleritis are the following:

1. Anterior (prevailing in 98 % cases) — inflammation of the sclera involving the anterior segment of the eye:
   a. Non-necrotizing (85 %):
      i. diffuse — a widespread inflammation;
      ii. nodular — development of immobile inflammatory tender deep purple-red nodules.
   b. Necrotizing (13 %):
      i. with inflammation — the most severe of all types, characterized by extreme pain and marked scleral damage;

ii. without inflammation (scleromalacia perforans) — the eye is not red or painful but the sclera is gradually thinned.

2. Posterior (in 2 %) — inflammation of the sclera involving the posterior segment of the eye.

Complaints. Moderate to severe deep eye pain that radiates to the forehead, brow, jaw, or sinuses and increases at night and early morning, tenderness on palpation, painful eye movements, tearing, photophobia, occasionally decreased vision, localized or diffuse redness.

Signs. Scleritis presents with a characteristic violet-bluish hue with scleral edema and dilatation. Other signs vary depending on the location of scleritis and the degree of involvement.

Anterior non-necrotizing diffuse scleritis is characterized by diffuse redness of one or more quadrants of the anterior sclera. The area is swollen and salmon pink to purple in color (fig. 9.2).

Anterior non-necrotizing nodular scleritis is characterized by one or more hard purple elevated scleral nodules, usually located near the limbus. These nodules are immobile and tender on palpation (fig. 9.3).

Anterior necrotizing scleritis with inflammation is an acute localized inflammation associated with areas of infarction due to vasculitis. It causes severe pain and tenderness and often causes permanent damage to the eye. The sclera becomes thin, translucent and ectatic, the underlying pigmented uvea becomes visible through it. There are signs of keratitis and anterior uveitis.

Anterior necrotizing scleritis without inflammation (scleromalacia perforans) typically occurs in elderly females suffering from rheumatoid arthritis. Presents as yellow patches of melting sclera due to obliteration of blood supply, separated from the surrounding normal area. Although the uveal tissue shines through the thin sclera, spontaneous perforation is extremely rare.

Posterior scleritis affects the sclera behind the equator. The condition is frequently asymptomatic. Signs include lid edema, proptosis and limitation of ocular move-

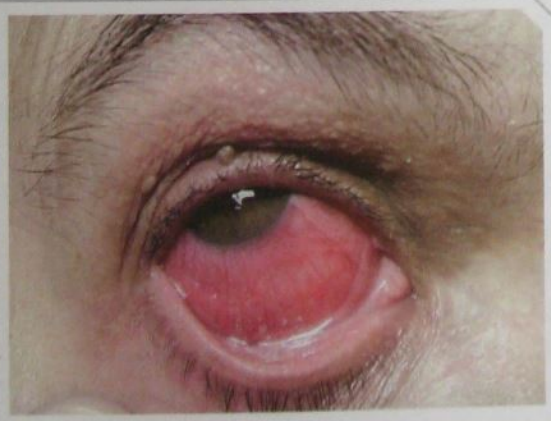


Fig. 9.2. Anterior diffuse scleritis

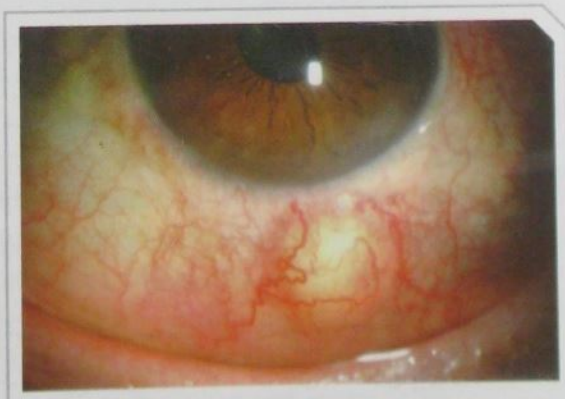


Fig. 9.3. Anterior nodular scleritis (from http://www.eyerounds.org)

ments, macular and optic disc edema, retinal hemorrhages, exudative retinal detachment. There may be loss of vision.

Methods of Examination. VA, slit-lamp examination, ophthalmoscopy, CT scan or B-scan ultrasonography may also help determine the extent of involvement. Except for ophthalmological methods, there are conducted general investigations for rheumatoid factor, syphilis, serum uric acid for gout, the Mantoux test, urine analysis, X-ray of the chest, paranasal sinuses, sacroiliac joint and orbit.

Differential Diagnoses. Episcleritis, allergic conjunctivitis, uveitis.

Scleritis may be differentiated from episcleritis by topical application of Phenylephrine 2.5 % or 10 %, which causes blanching of the superficial episcleral blood vessels in episcleritis but not in scleritis.

Treatment. Treatment of scleritis resulting from an underlying disease usually requires specific therapy for that disease. Topical treatment with eye drops is an adjunct to such systemic treatment. These eye drops will usually be anti-inflammatory, such as topical steroid drops or topical nonsteroidal anti-inflammatory drops (NSAIDs). Topical antibiotics are used if scleritis is caused by infectious.

Anterior non-necrotizing diffuse and nodular scleritis is treated by:

— oral NSAIDs — Indomethacin 75 mg, Ibuprofen 400—600 mg daily until inflammation resolves;

— oral steroids — Prednisolone 1 mg/kg one time a day (if unresponsive to NSAIDs);

— topical steroid eye drops — Dexamethasone, Hydrocortisone, Prednisolone 4—6 times a day;

— topical NSAIDs — Diclofenac, Indometacine, Cyclosporin eye drops 3 times a day.

Anterior necrotizing scleritis with inflammation is treated by:

— oral steroids — prednisolone 1 mg/kg one time a day;

— topical steroids eye drops 4—6 times a day;

— immunosuppressive agents (Methotrexate, Cyclophosphamide or Cyclosporin) in non-responsive cases;

— if there is a threat of perforation, scleral tissue transplantation might be necessary.

Anterior necrotizing scleritis without inflammation does not respond well to medical treatment. Surgical treatment may be needed to preserve globe integrity when the sclera is thinning out to the point of perforation — scleral grafts from donor tissue.

Posterior scleritis is treated by:

— oral NSAIDs — Indomethacin 75 mg, Ibuprofen 400—600 mg daily until inflammation resolves;

— oral steroids — Prednisolone 1 mg/kg one time a day (if unresponsive to NSAIDs);

— topical steroid eye drops — Dexamethasone, Hydrocortisone, Prednisolone 4—6 times a day;

NOTE!

In necrotizing scleritis subconjunctival injections of medications are contraindicated because they may lead to scleral thinning and perforation.

— immunosuppressive agents (Methotrexate, Cyclophosphamide or Cyclosporin) in non-responsive cases.

Prognosis depends on the severity of the disease. In non-necrotizing forms in relation to visual functions prognosis is relatively good. Patients with necrotizing scleritis have a high incidence of significant visual loss.

Complications. Despite of rapid and effective treatment scleritis can have various complications, including keratopathy, scleral thinning with staphyloma formation, scleral rupture/perforations, uveitis, glaucoma, cataract, fundus abnormalities that can lead to significant loss of vision.

Prophylaxis. There are no preventive methods other than the control of underlying conditions and early treatment of scleritis to minimize the risk of complications development.

4.2. Staphyloma

Definition. Staphyloma is a clinical condition characterized by thinning and protrusion of the sclera with the uveal tissue translucent through it. It is why this protrusion is generally dark blue or even black in color.

Etiology. Staphyloma can occur in response to progressive myopia, inflammatory or degenerative conditions of the sclera and increased IOP, or after perforating injury, complicated cataract surgery with wound dehiscence, radiotherapy, scleritis, chronic uncontrolled glaucoma or closed-angle glaucoma.

Clinical Picture. Staphylomas usually progress slowly with mild or no pain.

By anatomical classification staphylomas can be divided into:

— anterior (anteriorly to the limbus);
— intercalary (the limbal area lined by the root of the iris);
— ciliary (the scleral ring over the ciliary body zone — up to 8 mm behind the limbus);
— equatorial;
— posterior (behind the equator).

Complaints. Patients complain of black or dark-blue protrusion in front of the eye (anterior staphyloma), decreased vision, eye fatigue, eye pain.

Signs. Bulging and thinning of the sclera with transillumination of the underlying parts of the tunica uvea and their incarceration with progression, severe axial myopia.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, B-scan ultrasonography.

Differential Diagnoses. Blue sclera, buphthalmos, glaucoma, axial myopia, coloboma.

Treatment must be preventive in the first place — inflammatory or degenerative diseases of the sclera should be treated immediately, increased IOP must be controlled, high myopia corrected. Small progressive areas of thinning can be excised with corneoscleral graft implanting. In case of large staphyloma on incurably blind eyes the eye is enucleated and replaced with an implant.

Prognosis is determined by the underlying disease. Localized staphylomas have a good prognosis if treated properly. Staphylomas caused by severe local or general conditions are more difficult to treat and may have poorer prognosis. Vision is determined by other factors such as macular pathologies or glaucomatous damage.

Complications. Rupture of the staphyloma is the most dangerous complication. Postoperative complications include graft failure and infection.

Prophylaxis. Timely and adequate treatment of the underlying condition.

4.3. Blue Sclera

Definition. Blue sclera is a localized or generalized bluish discoloration of the sclera due to its abnormal thinning, through which the uveal tissue is seen.

Etiology. Blue sclera can be hereditary but it can also be seen in a number of conditions including high myopia, buphthalmos, ciliary staphyloma, osteogenesis imperfecta, rheumatoid arthritis, Marfan syndrome, Ehlers—Danlos syndrome, etc., some medications (like steroids), iron deficiency anemia, aging or trauma.

Clinical Picture. Blue sclera is asymptomatic; the condition is characterized by a blue scleral discoloration.

Complaints. Patients notice painless marked blue discoloration of the sclera.

Signs. Thinness and transparency of the collagen fibres of the sclera that allow visualization of the underlying uvea.

Methods of Examination. Slit-lamp exam, B-scan ultrasonography.

Differential Diagnoses. Ciliary staphyloma, conjunctival melanoma, conjunctival nevus.

Treatment. There is no specific treatment for blue sclera. The diagnosis and treatment of any underlying disease is very important. If a localized area of thinning is about to rupture, sometimes a "scleral patch graft" can be used to strengthen the area.

Prognosis depends on the underlying condition.

Complications. Scleral rapture.

Prophylaxis. Timely and adequate treatment of the underlying condition.

4.4. Tumors of the Sclera

True primary tumors of the sclera are extremely rare. They may be derived from the episclera, from the vessels or nerves which pass through it, from conjunctival intraocular growths or be a manifestation of a systemic neoplasia. Tumors may be sometimes mistaken for inflammatory diseases and require careful examination.

Review:

1. Key Points

Diseases of the sclera according to the origin may be *congenital or acquired;* according to the location — *superficial* or *deep; anterior* or *posterior;* according to the pathogenesis — *inflammatory, degenerative, trauma, tumors;* according to clinical presentation — *localized (nodular)* or *extended (diffuse); non-necrotizing* or *necrotizing*.

The main symptoms of scleral diseases are redness of the eye, mild to severe ocular pain, tenderness on palpation, photophobia, tearing, occasionally decreased vision. The main examination methods are VA, slit-lamp exam, ophthalmoscopy, B-scan ultrasonography, CT scan.

Inflammation of the sclera may be superficial (episcleritis) or deep (scleritis).

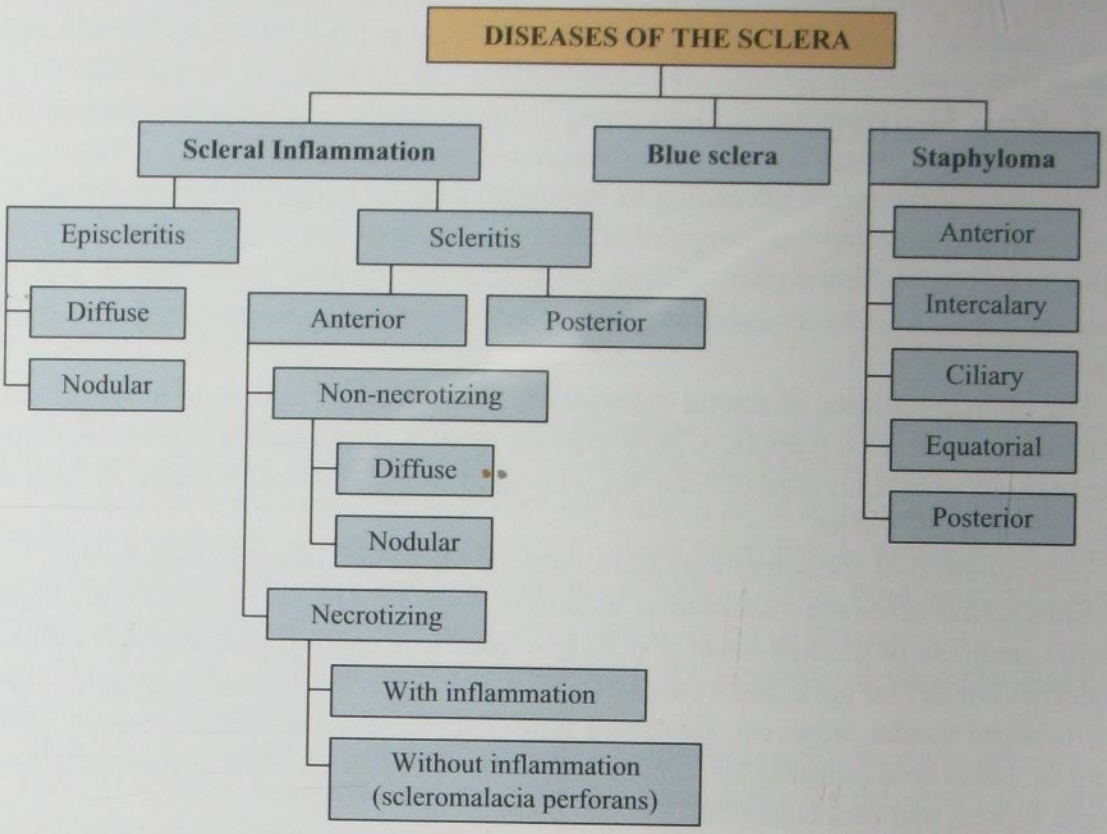
Episcleritis cases are classified as diffuse or nodular. The main symptoms of episcleritis are acute or gradual onset of diffuse or localized eye redness, mild to moderate tenderness or ocular pain. Most cases of episcleritis improve without treatment within two to three weeks, however, treatment may help the disease resolve sooner. Treatment includes cold compresses, topical lubricant eye drops, topical corticosteroid eye drops, topical and oral NSAIDs.

Scleritis is classified as anterior or posterior. Anterior scleritis can be non-necrotizing (diffuse and nodular) or necrotizing (with or without inflammation). The symptoms of scleritis are eye redness, deep eye pain, tenderness, painful eye movements, tearing, photophobia, occasionally decreased vision. Treatment starts from oral NSAIDs or steroids and topical steroid drops or NSAIDs and antibiotics.

Staphylomas can be divided into anterior, intercalary, ciliary, equatorial, posterior. The main complaints are scleral protrusion with underlying uvea transillumination, eye pain, decreased vision. The main treatment is surgical.

Blue sclera is asymptomatic and is characterized by a blue scleral discoloration. It can be congenital or acquired. There is no specific treatment for blue sclera.

2. Diagrams


DISEASES OF THE SCLERA
Scleral Inflammation
Blue sclera
Staphyloma
Episcleritis
Scleritis
Diffuse
Nodular
Anterior
Posterior
Non-necrotizing
Diffuse
Nodular
Necrotizing
With inflammation
Without inflammation
(scleromalacia perforans)
Anterior
Intercalary
Ciliary
Equatorial
Posterior

3. The Review Questions

A. Control Questions

1. What are the inflammatory diseases of the sclera and their symptoms?
2. What are the treatment methods of episcleritis?
3. What clinical types of scleritis do you know?
4. What are the general principles of scleritis treatment?
5. What complications may be caused by scleritis?
6. What diseases may cause scleral staphyloma?
7. Blue sclera, its clinical presentation and treatment methods.

B. Tests

1. **Point the symptoms typical for inflammatory diseases of the sclera:**
   A. Mild to severe ocular pain
   B. Photophobia
   C. Diffuse or sectoral eye redness
   D. Mucopurulent discharge
   E. Tearing
   F. Decrease of vision

2. **The main diagnostic methods of scleral disorders are:**
   A. Pachymetry
   B. Slit-lamp exam
   C. Ophthalmoscopy
   D. CT scan
   E. Tonometry
   F. B-scan ultrasonography

3. **Episcleritis may be of the following types:**
   A. Anterior
   B. Posterior
   C. Diffuse
   D. Nodular
   E. Non-necrotizing
   F. Necrotizing

4. **Scleritis may be of the following types:**
   A. Anterior
   B. Posterior
   C. Diffuse
   D. Nodular
   E. Non-necrotizing
   F. Necrotizing

5. **Scleral staphyloma may be caused by all these diseases except:**
   A. Perforating injury of the eye
   B. Posterior scleritis
   C. Progressive myopia
   D. Glaucoma
   E. Scleral melanosis
   F. Radiotherapy

6. **The main treatment methods of scleritis are:**
   A. Oral NSAIDs
   B. Oral steroids
   C. Topical steroid eye drops
   D. Topical NSAIDs
   E. Topical antibiotic eye drops
   F. Immunosuppressive agents

C. Clinical Cases

Case 1

A 45-year-old woman complains of moderate pain, photophobia, tearing, diffuse redness in the right eye that occurred 7 days ago. According to the patient's anamnesis, this is the second episode of the disease that occurs after overcooling. The patient suffers from polyarteritis. Visual acuity of both eyes is 1.0. Objectively: the right eye conjunctiva is diffuse, purple red in the outer segment, swollen and tender on palpation. There is no discharge from the eye fissure. The optical media are transparent. Examination of the dilated fundus shows no pathology. What is the possible diagnosis, its etiology and treatment?

Case 2

A 20-year-old female patient complains of burning, foreign body sensation, tearing, mild redness in both eyes. The patient denies presence of mucous or purulent discharge or eyelid crusts in the morning. The patient is allergic and suffers from psoriasis. Visual acuity on both eyes is 1.0. Objectively, there is a pink firm nodule near the limbus surrounded with dilated episcleral vessels, with freely moving overlying conjunctiva on both eye. The optical media are transparent. Dilated fundus examination shows no pathology. What is the diagnosis?

Case 3

A 72-year-old female patient complains of gradual moderate visual loss. There's a history of rheumatoid arthritis. Visual acuity OD=0.5; OS=0.3, does not improve with glasses. Objectively: the eyelids and conjunctiva are not affected. On both eyes in the perilimbal area there are focuses of ischemic sclera with thinning and ectasia, with pigmented uveal tissue shining through it. The iris is atrophic. Non-homogenous opacifications in the lens. Fundus examination shows glaucomatous optic disc cupping. IOP is 28 mm Hg OD, 32 mm Hg OS. What is the diagnosis?

Case 4

A 55-year-old male patient is preparing for surgical treatment of cataract on the left eye. B-scan measurement of the anterior-posterior eye globe length must be performed to calculate IOL power. According to the anamnesis, the patient has myopia. The measurement results are 28.6; 29.3; 30.2; 31.2; 30.8 mm. What is the cause of a big difference in measurement?

EYE FACTS

In humans the whole sclera is white, contrasting with the colored iris, but in other mammals the visible part of the sclera matches the color of the iris.

The eyes of all non-human primates are dark with small, barely visible sclera.

EYE FACTS

The relatively small cornea and iris contrasting with the sclera in humans plays a role in social communication as it helps to recognize the direction of individual's gaze, increasing the efficacy of nonverbal communication.

CHAPTER 10

Diseases of the Uvea

OBJECTIVES

Upon completion of the chapter the students should:

— know the classification of uveal diseases;
— know the basic diagnostic methods;
— be able to establish the diagnosis of uveitis, differentiating it from other causes of red eye;
— know the main symptoms and clinical picture of acute and chronic uveitis;
— identify the cause(s) of uveitis; prescribe appropriate therapy;
— know the principles of emergency care of uveal diseases;
— know the specifics of uveal disease complications.

Plan:

1. CLASSIFICATION OF UVEAL DISEASES
2. SYMPTOMS OF UVEAL DISEASES
3. EXAMINATION METHODS
4. DISEASES OF THE UVEA
   - **4.1. Congenital Anomalies**
     - Coloboma of the Iris
   - **4.2. Inflammatory Uveal Diseases**
     - Anterior Uveitis
       - Acute Iridocyclitis
     - Intermediate Uveitis
     - Posterior Uveitis
     - Panuveitis
     - Other Intraocular Inflammations
       - Endophthalmitis
       - Panophthalmitis
       - Sympathetic Ophthalmia
   - **4.3. Specific Types of Uveitis**
     - Ocular Lyme Disease
     - Ocular Toxoplasmosis
     - Siphilitic Uveitis
     - Tuberculosis Uveitis
   - **4.4. Tumors**
     - Nevus
     - Melanoma

1. Classification of Uveal Diseases

Uveal diseases can be classified:
— according to the origin — congenital or acquired;
— according to the etiology — inflammatory, autoimmune, idiopathic, ischemic, degenerative, inherited, toxic, traumatic, tumors;
— according to the onset of the pathological process — sudden or insidious;
— according to the course of the pathological process — acute, chronic, and recurrent;
— according to the clinical presentation — mild, moderate or severe;
— according to the location — anterior, intermediate, posterior, panuveitis;
— according to the nature of exudation — serous, fibrinous, purulent, hemorrhagic, mixed;
— according to the morphologic characteristics of the pathological process — granulomatous, nongranulomatous.

1.1. *Congenital anomalies* — colobomas, polycoria, aniridia, persistent pupillary membrane, corectopia, heterochromia, albinism.
1.2. *Inflammatory diseases (uveitis)* — iritis, iridocyclitis, pars planitis, cyclitis, choroiditis, panuveitis, endophthalmitis, panophthalmitis, sympathetic ophthalmia.
1.3. *Tumors* — benign: cyst, nevus, hemangioma; malignant: epithelioma, melanomas; metastatic tumors.

2. Symptoms of Uveal Diseases

— Decreased visual acuity.
— Blurred vision.
— Eye pain radiating to the forehead and temple.
— Eye redness and irritation.
— Tearing.
— Increased sensitivity to light — photophobia.
— Dark, floating spots before the eyes.
— Narrowing of the pupil.

3. Examination Methods

— Visual acuity.
— Examination by side illumination.
— Slit lamp exam.
— Gonioscopy.
— Ophthalmoscopy.
— Pupillary size and light response.
— Palpation for pain assessment.
— Tonometry.
— Perimetry.

4. Diseases of the Uvea

4.1. Congenital Anomalies

Coloboma is a defect or absence of an eye tissue that arises from improper fusion of the layers in the developing eye. It can affect the eyelid, iris, choroid, retina, optic disc or lens of the eye. A similar defect can also occur as a result of injury or eye surgery — but mostly coloboma is a congenital malformation.

Coloboma of the Iris

Definition. Coloboma of the iris is a full thickness defect in the iris that gives rise to the so-called "keyhole" pupil. A partial coloboma involves only the pupillary margin making the pupil oval. Although isolated iris coloboma is observed, it is often associated with coloboma in other parts of the eye.

Etiology. Most cases of coloboma have no known cause and are not related to other abnormalities. A small number of people with coloboma may be associated with hereditary or developmental conditions, trauma to the eye, or eye surgery (fig. 10.1).

Clinical Picture. There may or may not be any symptoms related to coloboma; it all depends on the size and location of the defect. Persons with an isolated coloboma may have normal vision and no symptoms, or they may have mild to severe vision impairment such as decreased visual acuity, blurry vision, glare, light sensitivity or photophobia.

Complaints. Unusual shape of the pupil or a hole in the iris, cloudy and decreased vision, glare, photophobia.

Signs. A keyhole-shaped or oval pupil, or a notch or hole in the iris.

Methods of Examination. Physical examination, slit-lamp exam; for any associated syndrome or anomaly ophthalmoscopy, CT, MRI are indicated.

Differential Diagnosis. Ectopia lentis et pupillae, iridocorneal endothelial syndrome, traumatic iris tear, aniridia.

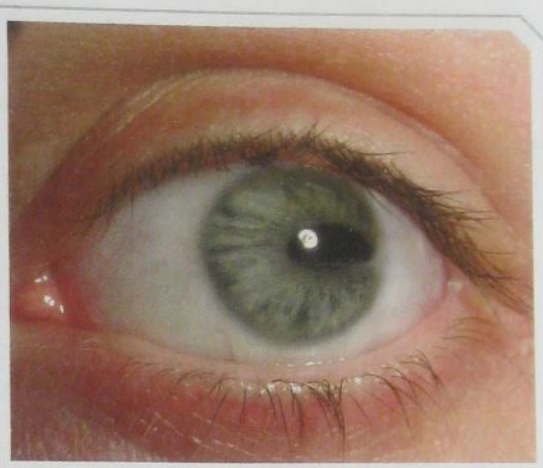


Fig. 10.1. Posttraumatic iris coloboma

Treatment. May not require treatment unless for cosmetics or photophobia. Supportive with sunglasses, tinted contact lenses or contact lenses with artificial pupils. Surgical iridoplasty in patients with severe and intolerable symptoms.

Prognosis is excellent in isolated iris coloboma. Visual prognosis depends on the extent of retinal, optic nerve, and other organ or system involvement.

Complications. Visual discomfort, photophobia, double vision, blurring.

Prophylaxis. No specific steps are identified for coloboma prevention.

Other congenital anomalies of the uvea are:

Albinism — a genetic disorder of melanin production that results in little or no pigment in the eyes, skin and/or hair. It is also associated with vision problems.

Aniridia — an eye disorder characterized by complete or partial absence of the iris.

Anisocoria — a condition where the pupil of one eye differs in size from the pupil of the other eye.

Heterochromia — a medical condition, in which the irides of both eyes are differently colored. It can be complete and/or partial.

Persistent pupillary membrane — presence of fine strands of pigmented tissue, which arises from the iris collarette and attaches to another spot on the iris without touching the pupillary margin. These strands are remnants of the fetal anterior vascular sheath supplying blood to the lens, which normally regresses shortly before birth.

Polycoria — a condition that is characterized by the presence of more than one pupil in the iris.

4.2. Inflammatory Uveal Diseases

Uveitis is a general term for intraocular inflammation affecting the uveal tract. The most widely used classification of uveitis is the one developed in 1987 by the International Uveitis Study Group (IUSG) on the basis of anatomical localization of

the inflammation. In 2004, the Standardization of Uveitis Nomenclature (SUN) workshop analysed these criteria, found them very useful and added criteria for the onset, duration and course of the disease.

EYE FACTS

Prior to the twentieth century, uveitis was typically referred to in English as *ophthalmia*.

Anterior uveitis is an inflammation of the front part of the uveal tract; it includes inflammation of the iris (*iritis*) and inflammation of the iris and ciliary body (*iridocyclitis*).

Intermediate uveitis is an inflammation that involves the posterior part of the ciliary body (*posterior cyclitis* or *pars planitis*).

Posterior uveitis is an inflammation of the part of the uveal tract behind the lens of the eye. It includes inflammation of the choroid (*choroiditis*) and inflammation of the choroid and retina (*chorioretinitis*).

Uveitis that affects the entire uveal tract is called *panuveitis* or *diffuse uveitis*.

Uveitis by onset may be *sudden* or *insidious*; by duration it can be *limited* (< 3 months' duration) or *persistent* (> 3 months' duration). According to the course uveitis may be:

— *acute* — sudden onset of inflammation that may last for less than 3 months with the usual duration of around 6 weeks;
— *chronic* — persistent inflammation that lasts more than 3 months, then recurs within 3 months after finishing the treatment;
— *recurrent* — characterized by relapse and remission of the disease.

According to etiology uveitis can be classified into exogenous and endogenous.

Exogenous uveitis is caused by penetrating injury to the uvea or invasion of micro-organisms from the outside.

Endogenous uveitis results from the factors that originate within the patient and subsequently produce ocular inflammation — systemic infective or non-infective diseases, auto-immune disorders:

— Infective — virus (e.g. herpes, cytomegalovirus), fungus (e.g. candida), parasites (e.g. toxoplasmosis) or bacteria (e.g. tuberculosis, syphilis, Lyme disease).
— Non-infective — lymphoma.
— Auto-immune — sarcoidosis, ankylosing spondylitis, rheumatoid arthritis, psoriasis, multiple sclerosis.

But, in many cases, the cause of uveitis is unknown — it is idiopathic non-specific uveitis.

According to the type of inflammatory response uveitis may be classified as granulomatous or non-granulomatous (table 10.1).

Granulomatous inflammation is associated with large, mutton-fat keratic precipitates (KPs), inflammatory cells (epitheloid cells and macrophages) in the anterior chamber, and presence of iris nodules (Koeppe nodules at the pupillary border and Busacca nodules located on the surface of the iris). Granulomatous uveitis is often associated with systemic conditions such as syphilis, Lyme disease, tuberculosis (TB), sarcoidosis or a local reactivation of herpetic viral infection.

Table 10.1

Differentiation of Granulomatous and Non-granulomatous Uveal Inflammation

	Granulomatous Uveal Inflammation	Non-granulomatous Uveal Inflammation
Onset	Insidious	Acute
Course	Chronic	Acute
Laterality	Bilateral	Unilateral
Pain	None or minimal	Marked
Photophobia	Slight	Marked
Blurred vision	Marked	Moderate
KPs	Mutton-fat, large, greasy	Small, fine
Cells	Macrophages and epitheloid cells	Lymphocytic cells
Pupil	Small and irregular	Small and irregular
Posterior synechiae	Broad-based, thick	Fine, filamentous
Iris nodules	Present (Koeppe and Busacca)	Absent
Site	Anterior, posterior or diffuse	Anterior
Posterior segment involvement	Common	Generally absent
Vitreous haze	Present	Absent
Recurrence	Sometimes	Common

Granulomatous uveitis is characterized by an insidious onset, minimal or no pain, only slight photophobia, blurred vision, a small and irregular pupil, and often a vitreous haze.

Nongranulomatous inflammation usually involves the anterior segment of the eye and is associated with small lymphocytic cells in the anterior chamber. It is most often idiopathic (juvenile idiopathic arthritis, ankylosing spondylitis, Reiter's syndrome, psoriasis, ulcerative colitis, Behcet's syndrome) or caused by human leukocyte antigen B27 (HLA-B27) conditions.

Non-granulomatous uveitis is a more common type and is characterized by acute onset, pain, photophobia, blurred vision, a small and irregular pupil, and a marked circumcorneal flush; there is usually no vitreous haze; it is usually unilateral. Recurrence is common.

Anterior Uveitis

Definition. Anterior uveitis is an inflammation of the anterior uveal tract that involves the iris (iritis), the anterior part of the ciliary body (anterior cyclitis), or both (iridocyclitis). It is the most common form of uveitis, predominantly occurring in young and middle-aged people.

Etiology. Anterior uveitis can result from a blunt or penetrating eye trauma, an intraocular foreign body, eye surgery, infections. It can also be a complication of other eye diseases, or it may be associated with general health problems such as rheumatoid arthritis, juvenile rheumatoid arthritis, ankylosing spondlyitis, lupus, syphilis, gout, Lyme disease, herpes virus infection, ulcerative colitis, psoriasis. In most cases, there is no obvious underlying cause.

Clinical Picture. Anterior uveitis can be classified according to its clinical course into acute or chronic anterior uveitis or according to its clinical appearance into granulomatous or non-granulomatous anterior uveitis.

Complaints. Patients with anterior uveitis usually complain of:

— eye pain;
— redness;
— blurred vision;
— photophobia;
— tearing.

Signs include:

— decreased VA;
— hyperemia of the conjunctiva adjacent to the cornea (circumlimbal or circumcorneal flush, or ciliary injection);
— constricted (myosis) or irregular-shaped pupil;
— keratic precipitates (KPs) (fig. 10.2);
— inflammatory cells and flare within the aqueous humor (the Standardization of Uveitis Nomenclature (SUN) Working Group developed a grading scheme for both anterior chamber cells and flare (table 10.2));
— posterior synechiae (fig. 10.3);
— hypopyon and fibrin within the anterior chamber (in severe cases);
— IOP may be reduced due to ciliary body inflammation that results in impairment of aqueous humor secretion.

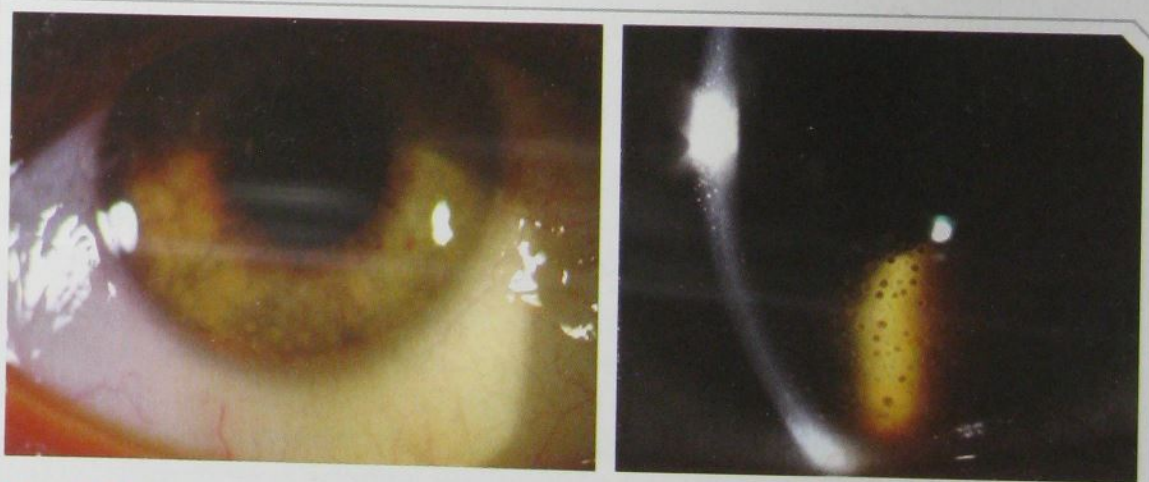


Fig. 10.2. Keratic precipitates on the corneal endothelium front view and at slit-lamp exam

Methods of Examination. Visual acuity test, slit-lamp examination, ophthalmoscopy, tonometry. If an underlying condition is the cause of uveitis, a patient may be referred to another doctor for a general medical examination and laboratory tests.

Differential Diagnosis. Conjunctivitis, keratitis, angle-closure glaucoma, diffuse uveitis, intraocular foreign body.

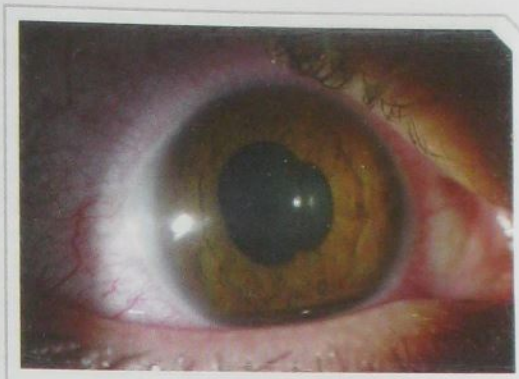


Fig. 10.3. Posterior synechiae

Table 10.2

Grading of Cells and Flare

Grade	Cells	Flare
0	No cells	Complete absence
1+	5 to 10 cells in view	Faint (barely detectable)
2+	10 to 20 cells in view	Moderate (the iris and lens still clear)
3+	20 to 50 cells in view	Marked (the iris and lens hazy)
4+	>50 cells in view	Intense (coagulated aqueous, no circulation, fibrin visible)

Treatment. The aim of the treatment is to relieve pain, to reduce inflammation, to treat any underlying cause (if possible), and to prevent complications. The length of treatment depends on disease severity and how well the eye improves with the treatment.

— *Mydriatic or cycloplegic eye drops*, such as Atropine 1 %, Tropicamide 1 % are used — to dilate the pupil, which helps to relieve the eye pain and to prevent adhesions between the iris and the lens.

— *Topical steroids* are the mainstay of treatment for anterior uveitis to reduce inflammation — Prednisolone acetate 1 %, Dexamethasone 0.1 % every 2 hours while awake.

— *Treatment of an underlying condition* — if there is an underlying cause, appropriate treatment should be given. Infectious uveitis is treated with appropriate antiviral or antibiotic eye drops as well as corticosteroids and cycloplegics.

— *Treatment of complications* — laser peripheral iridotomy for posterior synechiae, cataract or/and glaucoma surgery.

Prognosis. Most cases of anterior uveitis respond favorably to early diagnosis and treatment. Anterior uveitis may recur, especially when there is a systemic etiology.

Complications. Complicated cataract, anterior synechiae, glaucoma, choroiditis, macular edema, retinal detachment, phthisis bulbi.

Prophylaxis. Other than avoiding certain infections, there is no way to prevent anterior uveitis.

Acute Iridocyclitis

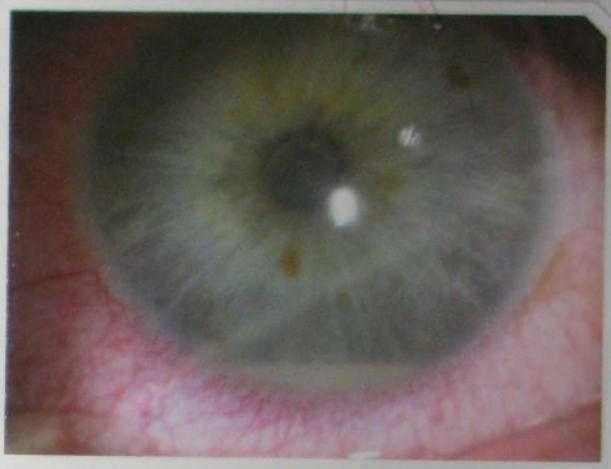


Fig. 10.4. Acute fibrinous iridocyclitis with ciliary injection, pupil occlusion and hypopyon in the anterior chamber

Definition. Acute iridocyclitis is a type of anterior uveitis that is characterized by sudden symptomatic onset. It is mostly unilateral and lasts for about 6 weeks.

Etiology. Acute iridocyclitis may occur as an isolated medical problem without any association with illness or inflammation elsewhere in the body. It might arise as a result of ocular trauma or surgery, local infection, may occur as an adverse reaction to a medication. It may also be associated with ankylosing spondylitis, rheumatic diseases, sarcoidosis, tuberculosis, syphilis, ulcerative colitis, vasculitis. In most cases, there is no obvious underlying cause.

Clinical Picture. Acute iridocyclitis is characterized by a sudden onset and limited duration. The symptoms typically develop rapidly, over a few days.

Complaints. The classical five symptoms of acute iridocyclitis are:

— progressive eye pain that is worse at night;
— red eye;
— photophobia;
— excessive tearing;
— blurred or cloudy vision.

Signs. The typical findings include (fig. 10.4):

— ciliary injection (circumlimbal flush);
— constricted (myosis) and irregularly shaped pupil;
— discoloration of the iris and loss of its normal pattern;
— KPs;
— flare and cells in the anterior chamber;
— posterior synechiae;
— iris nodules (are seen in granulomatous uveitis);
— hypopyon;
— fibrin;
— hyphema in the hemorrhagic type of uveitis;
— anterior vitreous cells.

Methods of Examination. Visual acuity test, slit-lamp examination, ophthalmoscopy.

Differential Diagnosis. Acute conjunctivitis, acute angle-closure glaucoma (table 10.3).

Table 10.3

Differential Diagnosis of Acute Iridocyclitis, Acute Conjunctivitis and Acute Angle-Closure Glaucoma

Clinical Signs	Acute Conjunctivitis	Acute Iridocyclitis	Acute Angle-Closure Glaucoma
Onset	Gradual	Usually gradual	Sudden
Pain	Foreign body sensation, no pain	Localized in the eye	Severe, radiating in the trigeminal nerve distribution
Vision	Normal	Decreased	Loss of vision
Colored halos	No	No	Yes
Discharge	Mucopurulent	Watery	Watery
Conjunctiva	Conjunctival injection	Ciliary injection (circumlimbal flush)	Ciliary injection
Cornea	Normal	Normal, KPs	Hazy (edema)
Pupil	Normal, reacting	Constricted (myosis), fixed	Dilated (mydriasis), fixed
Anterior chamber transparency	Clear	Cells and flares	Hazy due to corneal edema
Anterior chamber depth	Normal	Normal	Shallow
IOP	Normal	Normal or decreased	Markedly elevated
Systemic complication	Normal	Normal	Headache, nausea, vomiting

Treatment. Acute iridocyclitis management aims at suppressing inflammation, relieving pain caused by spasm in the ciliary body, and preventing the development of complications. Treatment involves:

— *mydriatic or cycloplegic eye drops*, such as Atropine 1 %, Tropicamide 1 % to dilate the pupil, which helps to relieve eye pain and prevent adhesions between the iris and the lens;

— *topical steroids* that reduce inflammation — Prednisolone acetate 1 %, Dexamethasone 0.1 % every 2 hours while awake;

— *periocular or oral steroids* are used if the patient is unresponsive to topical therapy;

— *treatment of an underlying condition* — if there is an underlying cause, appropriate treatment should be given. Infectious uveitis is treated with appropriate antiviral or antibiotic eye drops as well as corticosteroids and cycloplegics;

NOTE!

Acute iridocyclitis requires immediate treatment as it can lead to the development of significant complications and irreversible loss of vision.

— *systemic analgesics* such as Paracetamol, Ibuprofen may also help to reduce pain;
— *sunglasses* might help to deal with photophobia;
— *warm compresses* may help relieve the pain.

Prognosis. There is usually a good visual outcome with prompt and effective treatment but the condition has a tendency to recur especially if there are underlying causes, which contribute to inflammation of the eye, or if the patient has autoimmune or inflammatory diseases.

Complications. Corneal opacity, formation of synechiae, secondary glaucoma, complicated cataract, neovascularization, permanent vision damage, and even blindness.

Prophylaxis. Prevention consists in timely treatment of the basic disease and elimination of chronic centers of infection.

Intermediate Uveitis**

Definition. Intermediate uveitis is an inflammation of the uvea localized in the posterior part of the ciliary body — the pars plana. *Pars planitis*, *posterior cyclitis*, *chronic cyclitis* and *vitritis* are other terms used to describe this type of uveitis. Intermediate uveitis is the least common type of uveitis, making up only 7—15 % of cases. It tends to affect patients in their late teens or early adult years. Men are affected more commonly than women.

Etiology. Intermediate uveitis is typically idiopathic though it can be associated with, or be an ocular manifestation of some diseases, e.g. sarcoidosis, multiple sclerosis, tuberculosis, syphilis, and Lyme disease.

Clinical Picture. Intermediate uveitis involves the vitreous and peripheral retina. It usually causes painless blurred vision and does not cause redness and photophobia.

The condition is usually insidious, chronic, relapsing, and affects both eyes with one eye being worse than the other.

Complaints. Most patients experience:
— blurred or cloudy vision;
— decreased vision;
— dark spots that float in the visual field (floaters).

Signs:
— decreased VA;
— the conjunctiva and sclera are white without injection;
— the cornea is clear and appears uninvolved;
— minimal anterior chamber cells;
— no posterior synechiae, but may be present in intermediate uveitis with granulomatous inflammation (sarcoidosis, tuberculosis);
— yellowish-white exudates ("snowbanks") over the pars plana and ora serrata;
— yellowish-white round inflammatory cells in the inferior peripheral vitreous free-floating as snowballs;

— vitreous haze.

Methods of Examination. VA test, slit-lamp exam, ophthalmoscopy, B-scan ultrasonography. To determine the cause of the disease the following investigations may be useful — basic laboratory tests, serologic studies, chest X-ray, brain MRI. Fluorescein angiography is useful to confirm cystoid macular edema, optic nerve involvement, peripheral retinal vasculitis. Consultation with a gastroenterologist, a neurologist, or an infectious disease specialist may be necessary depending on physical findings.

Differential Diagnosis. Chronic iridocyclitis, posterior uveitis.

Treatment of intermediate uveitis includes the following:

- *corticosteroids* to reduce inflammation:
  - *topical instillations* of Prednisolone acetate 1 % or Prednisolone sodium phosphate 1 %;
  - *periocular injections* using a sub-Tenon approach are indicated in uveitis unresponsive to topical treatment;
  - *intravitreal injections* of Triamcinolone or Dexamethasone intravitreal implant;
  - *oral steroids* — if local therapy is not effective;
- *immunosuppressive agents* (Cyclosporine, Azathioprine, and Methotrexate) are added in cases when inflammation is not resolved within 3 months of corticosteroids treatment;
- *surgical options* — cryotherapy, vitrectomy, and laser photocoagulation.

Prognosis is variable since the visual outcome depends on the presence and degree of macular edema. The condition has a chronic course with episodic exacerbations and remissions.

Complications. Cataract, glaucoma, cystoid macular edema, peripheral retinal vasculitis, optic disc edema or neovascularization, vitreous opacification, retinal detachment.

Prophylaxis. Unfortunately, intermediate uveitis be cannot directly prevented since most often the cause is unknown or not modifiable.

Posterior Uveitis

Definition. Posterior uveitis is an inflammation of the posterior segment of the uvea that often involves both the choroid and the retina. It is called choroiditis or chorioretinitis (fig. 10.5). This condition is comparatively rare but can cause severe vision loss.

Etiology. Posterior uveitis may be congenital or acquired at any age as an ocular manifestation of a severe infectious or autoimmune systemic disease or eye injury.

Congenital toxoplasma and cytomegalovirus (CMV) are the most common causes of congenital chorioretinitis and they are often asymptomatic at birth. Tuberculosis, syphilis, AIDS, Lyme disease, fungal infection, parasitic condition, as well as intraocular malignancy may lead to posterior uveitis. In many cases the cause is unknown.

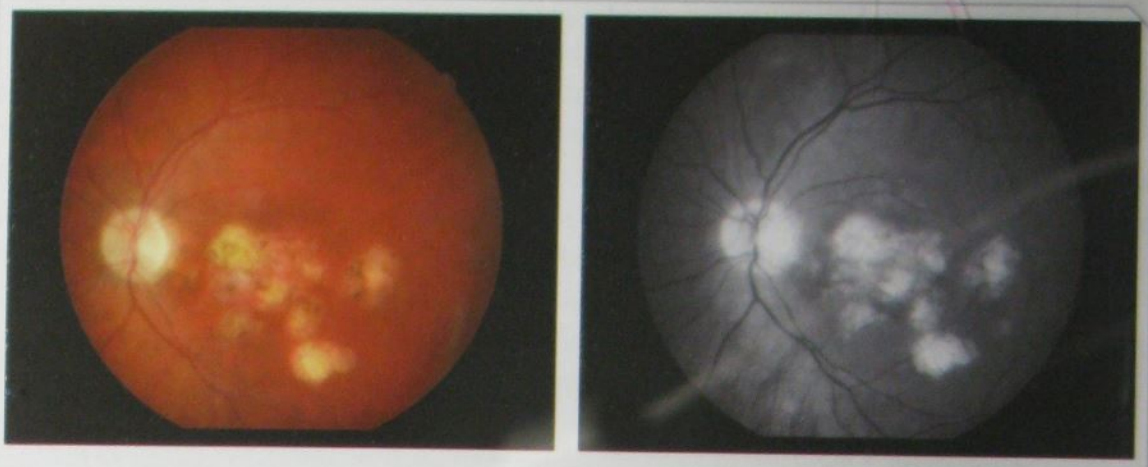


Fig. 10.5. Chorioretinitis (ophthalmoscopic picture and view at fluorescein angiography)

Clinical Picture. Posterior uveitis causes painless blurring of vision and affects both eyes. To some people, it can cause severe vision loss. It is usually chronic and frequently a recurrent condition. It often lasts longer than other types of uveitis — for months, and even years.

Complaints. Patients complain of:

— blurred or cloudy vision;
— dark spots that float in the visual field, or floaters;
— seeing flashing lights, or photopsia;
— distortion of the shape of objects, or metamorphopsia;
— loss of peripheral vision;
— impaired night and color vision.

Signs. Ophthalmologic examination can reveal:

— decreased VA;
— scotomas in the visual field;
— vitreous opacities;
— isolated or multiple choroiditis foci, which in the early stage appear as grey-white or yellow-white cotton balls with indistinct borders due to surrounding edema, and in the healing (scarring) stage become white with defined dark pigmented borders;
— the major choroidal vessels are visible;
— spill-over anterior uveitis (signs of anterior segment inflammation) may be present.

Methods of Examination. VA test, slit-lamp exam, ophthalmoscopy, perimetry, B-scan ultrasonography, OCT. Blood tests and X-rays may be required to diagnose associated conditions.

Differential Diagnosis. Posterior vitreal detachment, retinal detachment, intraocular foreign body, choroidal dystrophy, intraocular tumors, central nervous system lymphoma.

Treatment options may vary depending on the underlying cause and include steroids, antibiotics and various combinations of these medicals:

— *steroids in the form of injections or/and oral* to control inflammation;

— *antibiotics, antiviral, anti-parasitic, or antifungal medicines* to treat causative infection;

— *immunosuppressants* are recommended if the cause is autoimmune disorders;

— *sunglasses* should be worn to protect the eyes from UV rays.

Prognosis. While treatment causes the disease to regress, recurrences are possible and vision damage may happen even with treatment.

Complications. Secondary glaucoma, macular edema, retinal vein occlusion, retinal detachment, vision loss.

Prophylaxis. Because posterior uveitis is often caused by infections or systemic illnesses, treatment of an underlying disorder may help to prevent the condition development.

Panuveitis

Definition. Panuveitis (diffuse uveitis, iridocyclochorioiditis) is an inflammation involving the entire uveal tract — the iris, the ciliary body, and the choroid. The intermediate segment of the eye is not involved.

Etiology. Panuveitis can be caused by topical or systemic infections or chronic inflammatory diseases such as Behɜet's disease, tuberculosis, sarcoidosis, toxoplasmosis and syphilis. Some other possible causes may include bruises, tumors, or its cause may be unknown.

Clinical Picture. Panuveitis is the most severe form of the disease. It is a rare but severe condition that can lead to irreversible vision loss.

Complaints. Panuveitis may have any combination of symptoms. Most often patients complain of eye pain, redness of the eye, blurred vision, gradual vision loss, photophobia, tearing, and seeing dark spots.

Signs. Clinical signs result in a combination of anterior, intermediate, and posterior uveitis.

Methods of Examination. VA test, slit-lamp exam, ophthalmoscopy, perimetry, B-scan ultrasonography, OCT. Other tests may be required to diagnose causative disorders.

Differential Diagnosis. Acute conjunctivitis, acute anterior uveitis, acute angle-closure glaucoma, posterior uveitis, endophthalmitis, scleritis.

Treatment of panuveitis has three main points: to prevent vision loss, to relieve patients' complains, and to manage the underlying disease. Corticosteroids are commonly used to reduce the inflammation in panuveitis. Mydriatics are administered to relieve pain due to ciliary spasm and to prevent posterior synechiae formation. Immunosuppressive drug therapy may also be necessary.

Prognosis for vision is very poor.

Complications. Endophthalmitis, cataract, glaucoma, retinal detachment, neovascularization of the retina, optic nerve disc or iris, cystoid macular edema, irreversible vision loss.

Prophylaxis. Protecting the eyes from trauma, prompt control of infections can prevent some cases of panuveitis, but most cases cannot be prevented.

Other Intraocular Inflammations

Endophthalmitis

Definition. Endophthalmitis (or vitreous abscess) is a severe purulent intraocular inflammation involving the internal structures and humors of the eye (fig. 10.6).

Etiology. Endophthalmitis may be infectious or non-infectious, and the infectious cases may be of endogenous or exogenous origin. Non-infectious, or sterile, endophthalmitis can result from inflammation related to retained lens material or a reaction to intraocular drugs such as steroids.

Endogenous endophthalmitis occurs secondary to a hematogenous spread from a distant infective source in the body. Exogenous endophthalmitis can be a complication following eye surgery or a penetrating eye injury and retained intraocular foreign bodies. The most common infections that may cause endophthalmitis are various bacteria (e.g., Staphylococcus species, Streptococcus species, Gram-negative bacteria), fungi (e.g., Candida Aspergilla) and rarely viruses (e.g., Herpes Simplex or Herpes Zoster).

Clinical Picture. The clinical course of the disease can be acute or chronic. Acute endophthalmitis is characterized by acute loss of VA, deep ocular pain, severe reddening of the eye. Chronic endophthalmitis has a far less severe clinical course with moderate symptoms.

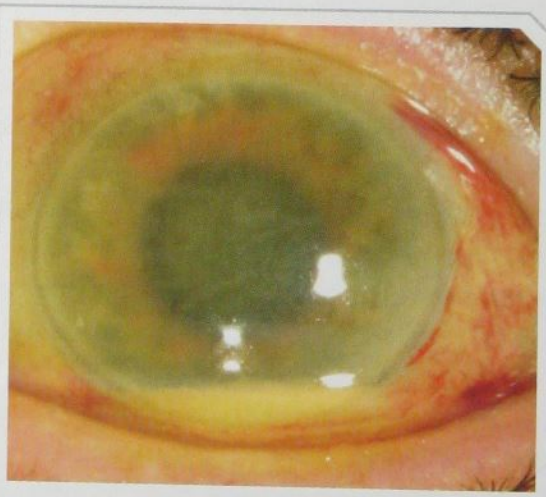


Fig. 10.6. Postoperative endophthalmitis with conjunctival congestion, corneal edema, hypopyon, and fibrin in the pupil (from ophthalmology-management.com)

Complaints. Patients usually complain of severe eye pain and irritation, redness of the eye, extreme sensitivity to bright light, tearing, decreased vision, occasionally swelling of the eyelid, and headache, but chronic condition may be asymptomatic.

Signs vary widely and include decreased VA, lid edema, ciliary injection, and chemosis of the conjunctiva, corneal edema, KPs, cells and flare in the anterior chamber, hypopion, decreased red reflex, vitreous cells, cotton wool spots, proptosis, and decreased ocular mobility, decreased IOP.

Methods of Examination. VA test, slit-lamp examination, ophthalmoscopy,