B-scan ultrasoundgraphy, CT or MRI scan of the orbit may help to rule out other ophthalmic conditions.

Differential Diagnosis. Corneal laceration, corneal ulcer, iritis, uveitis, vitreous hemorrhage, intraocular lymphoma.

NOTE!

Acute bacterial endophthalmitis is a medical emergency, because delay in treatment may result in vision loss.

Treatment is based on the underlying cause. If the infection is caused by bacteria, treatment involves intravitreal and systemic antibiotics (Gentamicin, Ciprofloxacin, Cephazolin), intraocular anti-inflammatory agents, and steroid topical therapy (Prednisolone, Dexamethasone). If infection is severe, vitrectomy may be performed to remove infectious material from the inside of the eye. Mycotic endophthalmitis is usually treated with Amphotericin B, Fluconazole and steroids.

Prognosis. Even with early and appropriate treatment, the prognosis for vision is often poor.

Complications. Panophthalmitis, corneal ulcer, orbital cellulitis, glaucoma, phthisis bulbi, blindness.

Prophylaxis. This condition usually cannot be prevented. But avoiding eye trauma, prompt treatment of endogenous infections and proper sterile preparation of the surgery site may help to avoid endophthalmitis.

Panophthalmitis

Definition. Panophthalmitis is an acute purulent inflammation of all the eye structures and the immediate surrounding tissues.

Etiology. Panophthalmitis may develop after penetrating ocular injury, especially with a retained intraocular foreign body or as a result of postoperative bacterial or fungal infection. The most common infections are pyogenic organisms — Staphylococcus, Streptococcus, or Pneumococcus species, Escherichia coli. Endogenous panophthalmitis is rare and metastatic in origin and develops from an infective embolus in the retinal or choroidal vessels.

Clinical Picture. The disease develops very rapidly. It begins either as purulent anterior or posterior uveitis; and soon a full picture of panophthalmitis develops and the eyeball may eventually perforate.

Complaints. Patients usually complain of severe ocular pain and headache, marked eye redness, rapid swelling of the eyelids, painful and limited ocular movement, complete loss of vision, excessive tearing, purulent discharge, and constitutional symptoms like fever and vomiting.

Signs. Eyelids edema, proptosis, ciliary and conjunctival injection, chemosis of the conjunctiva, corneal edema and haziness, massive pus in the anterior chamber and vitreous, IOP is markedly raised, perception of light is absent.

Methods of Examination. External examination, slit-lamp exam, tonometry, and imaging studies.

Differential Diagnosis. Corneal ulcer, acute interior uveitis, panuvelitis, endophthalmitis, orbital cellulitis.

Treatment includes analgesics to relieve pain, intensive systemic and local antibiotic therapy to prevent further spread of infection in the surrounding structures, and systemic steroids to reduce inflammatory reactions. Evisceration should be performed in case of loss of light perception and/or eyeball rupture to avoid the risk of intracranial dissemination of infection.

Prognosis of panophthalmitis is poor. It usually ends in blindness, loss of the eyeball. In addition, there exists a possibility of infection spreading from the eye to the brain, which could cause even more serious problems, such as meningitis or encephalitis.

Complications. Loss of vision, phthisis bulbi, eye rupture, orbital cellulitis, cavernous sinus thrombosis, meningitis or encephalitis.

Prophylaxis consists in prompt and proper treatment and follow-up of penetrating ocular injury and proper sterile preparation of the surgery site during the eye operation procedure.

NOTE!

Panophthalmitis may be not only a vision-threating, but even a life-threatening condition.

Sympathetic Ophthalmia

Definition. Sympathetic ophthalmia (or sympathetic uveitis) is a bilateral granulomatous uveitis that occurs after penetrating trauma or surgery to one eye. The injured eye is called the exciting eye, and the uninjured one is called the sympathizing eye.

Etiology. Although the exact cause of this condition is unknown, it may be a result of an autoimmune inflammatory response to the uveal pigment. The tissues in the injured eye (the uveal tract, lens, and retina) act as antigens and provoke an autoimmune disorder in the unaffected eye. The injured eye becomes inflamed first, and the other eye follows (i.e., sympathetically).

In approximately 80 % of cases, sympathetic ophthalmia appears in 2—12 weeks after penetrating injuries or intraocular surgery in the fellow eye, and 90 % of cases occur within 1 year after injury. However, isolated cases developing 30 years after initial injury have been reported. Very rarely, sympathetic ophthalmitis can also occur following an intraocular surgery.

Clinical Picture. The onset of the disease is insidious. The injured eye becomes painful and photophobic, and visual acuity is diminished. The uninjured (sympathizing) eye then follows a similar course.

Complaints. The patient's complaints may initially include blurred vision due to a loss of accommodation and floaters in the sympathizing eye. As the disease progresses, pain and redness of the eye, photophobia, metamorphopsia and significant visual loss are observed.

Signs. Mild to moderate ciliary injection, KPs, vitreous opacities, multiple yellowish-white choroidal lesions, optic disc edema.

Methods of Examination. Thorough medical history, VA test, slit-lamp exam, ophthalmoscopy, B-scan ultrasonography, fluorescein angiography, OCT.

Differential Diagnosis. Vogt—Koyanagi—Harada syndrome, iridocyclitis, and choroiditis from other causes.

Treatment involves prompt and aggressive use of corticosteroids delivered by topical, periocular, or systemic routes. However, if inflammation cannot be controlled, immunosuppressives, such as Azathioprine, Methotrexate, Chlorambucil, or Cyclosporine may be necessary. The goal of the treatment is to stop inflammation and autoimmune response. Patients with sympathetic ophthalmia require close monitoring, since this is a recurrent disorder.

When the injured eye is blind and has no potential for any visual recovery, prophylactic enucleation within the first 2 weeks of injury is indicated to prevent the onset of sympathetic ophthalmia in the fellow eye. However, it is not advisable if the exciting eye retains useful vision, since this traumatized eye may ultimately retain better vision than the sympathizing eye.

Prognosis. When sympathetic ophthalmia is diagnosed early and immediate treatment with steroids is started, vision may be retained. However, in advanced cases, it can progress to complete bilateral blindness.

Complications include cataract, glaucoma, chorioretinal scarring, macular edema or scars, retinal detachment, optic nerve atrophy, and phthisis bulbi, which may affect either or both the exciting and sympathizing eyes.

Prophylaxis of sympathetic ophthalmia development is prompt and careful ocular trauma treatment and appropriate immunomodulatory therapy.

4.3. Specific Types of Uveitis

Ocular Lyme Disease

Lyme disease is a multisystem infectious disease caused by spirochete Borrelia burgdorferi, transmitted by the bite of an infected Ixodes tick, also known as deer tick. In the last decades it became the most common vector-borne illness in the USA, Scandinavia, Eastern and Middle Europe, and Asia. Most cases are seasonal with peak incidences occurring in late spring and autumn, but in areas with mild climates activity may be year-round.

Lyme is called "The Great Imitator", because its symptoms mimic many other diseases. It can affect any organ of the body, including the brain and nervous system, muscles and joints, and the heart. Ocular manifestations in Lyme disease have been considered comparatively rare (up to 5 % of cases). The most common forms of ocular Lyme disease are posterior uveitis and panuveitis, but anterior uveitis may be noted too.

Definition. Ocular Lyme disease is ocular manifestation of the systemic Lyme disease.

Etiology. Lyme disease is caused by a spirochete — a corkscrew-shaped bacterium called Borrelia burgdorferi. It is most commonly transmitted to humans through the bite of an infected tick. After an incubation period of several days, the spirochetes disseminate hematogenously to multiple organs. The disease may also occur due to the body's immune response to the infection.

Clinical Picture. The disease follows a three-stage pattern. Primary systemic symptoms include circular red, spreading rash on the skin around the tick bite (erythema migrans, or bull's-eye rash), headache, malaise, fatigue, fever, and lymphadenopathy. They may be followed weeks to months later by neurologic, cardiac, or joint abnormalities.

Ocular manifestations may occur at any stage but more common in the last two stages. The most common presentation of the first stage is follicular conjunctivitis. At the second stage the disease manifests as anterior uveitis, intermediate uveitis, posterior uveitis or panuveitis. The most common ocular manifestation in the third stage is keratitis, and much less common episcleritis. These may present months to years after the primary infection.

Complaints. The most often patients complain of blurred vision, periodic ocular pain, eye redness, photophobia, floaters, partial loss of the field of vision, and diplopia.

Signs. Decreased VA, scotomas in the visual field, posterior synechiae, vitreous opacities, optic disc swelling, retinal hemorrhages.

Methods of Examination. Medical history, systemic clinical findings, VA test, slit-lamp exam, ophthalmoscopy, B-scan ultrasonography, fluorescein angiography, blood tests, PCR.

Differential Diagnosis. Uveitis associated with syphilis, tuberculosis, sarcoidosis, rheumatoid arthritis, retinal vasculitis, multifocal choroidal inflammation.

Treatment. Early infection or nonspecific symptoms with positive Lyme titers may be treated with oral Doxycycline (100 mg twice daily for 4 to 6 weeks) or Tetracycline (500 mg four times a day for 4 to 6 weeks).

In late stages intraocular Lyme disease is usually treated with intravenous Ceftriaxone (2 g/day in two divided doses) or Cefotaxime (2 g every 8 hours) for 21 days. Topical steroids and cycloplegics in conjunction with antibacterial agents is recommended for ocular inflammation.

Quercetin, Bromelain, and antioxidant vitamins E and C may be helpful to strengthen the immune system in order to lower systemic inflammation and reduce damage to the eye.

Prognosis. Prompt systemic antibiotic treatment is effective and leads to recovery of VA.

Complications. Loss of vision due to long-standing intraocular inflammation if left untreated.

Prophylaxis. People in endemic areas should take precautions against tick bite. Transmission of Borrelia burgdorferi does not usually occur until the infected tick has been in place for more than 36 hours. Thus, searching for ticks after potential exposure and removing them promptly can help prevent infection. A single dose of

200 mg doxycycline can be used to prevent Lyme disease if given promptly after ticks are removed.

Ocular Toxoplasmosis

Ocular toxoplasmosis is a major cause of infectious posterior uveitis owing to the widespread distribution of the causative organism Toxoplasma gondi throughout the world. It has been estimated that Toxoplasma gondii infects up to a third of the world's population. It predominantly affects children and young people (25—45 years of age) and is characterized by recurrences that can ultimately lead to significant visual loss.

Definition. Ocular toxoplasmosis, or toxoplasmic retinochoroiditis, is an inflammation of the posterior part of the eye caused by the parasite Toxoplasma gondii. The most common part of the eye to become affected is the retina and choroid (retinochoroiditis).

Etiology. Ocular toxoplasmosis is caused by an obligate intracellular parasitic protozoan Toxoplasma gondii. Members of the cat family are its definitive hosts; other species, including mammals, birds, and reptiles, may serve as intermediate hosts. The parasite can be found in the host's tissues, such as muscles and body fluids (saliva, milk, and urine).

Humans can acquire toxoplasmosis through food (e.g., raw or undercooked meat, unpasteurized milk, contaminated water, unwashed fruit and vegetables), by coming into contact with infected cat litterbox or sandboxes, or soil contaminated with cats' feces, and rarely dust in the air. The parasite could be transmitted from an infected person to another during an organ transplant or blood transfusion. Congenital toxoplasmosis is transmitted from the mother to the fetus across the placenta during pregnancy.

Clinical Picture. Ocular toxoplasmosis can affect one or both eyes.

In most individuals, a Toxoplasma infection does not lead to immediate clinical symptoms because a healthy person's immune system usually keeps the parasite in an inactive state. Around 10—15 % of people may develop symptoms similar to mild flu or glandular fever, such as a temperature, sore throat, and muscle aches.

In people with weakened immune systems, such as those who have had an organ transplant, those with HIV and AIDS, and those receiving certain types of chemotherapy treatment Toxoplasma infection could cause serious complications including irreversible vision loss and neurological problems.

Congenital toxoplasmosis is also more serious and occurs when a woman becomes infected during pregnancy and passes the infection on to her unborn baby. This can result in the baby developing serious health problems such as blindness and brain damage.

Complaints. Typical symptoms of active disease are reduced vision, blurred vision, and floaters. With secondary iritis patients complain of pain, redness, photophobia and sometimes tearing.

Signs. The hallmark of ocular toxoplasmosis is focal necrotizing retinochoroiditis with overlying vitritis ("headlight in the fog" appearance). The active retinal lesion is fluffy yellowish-white in color, varies in size and is usually circular or oval in shape located mainly in the macular region. The size varies from one-tenth to five disc diameters. Recurrent lesions tend to occur at the margins of old scars (satellite lesion), but they also can occur elsewhere in the fundus (fig. 10.7, 10.8).

Granulomatous anterior uveitis is seen with mutton-fat KPs, cells and flare, and posterior synechiae. Raised IOP is often encountered in ocular toxoplasmosis. Optic nerve head involvement in the form of neuroretinitis, papillitis can be seen.

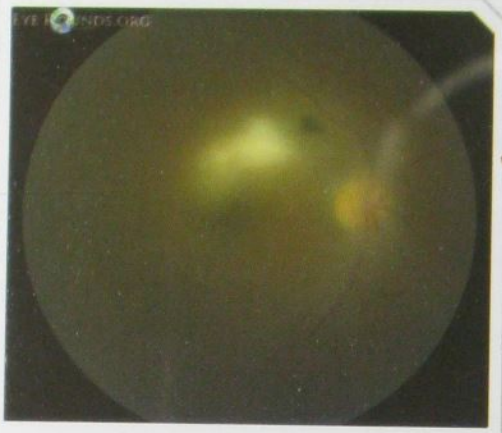


Fig. 10.7. Toxoplasmic posterior uveitis with "headlight in fog" appearance, white active lesions adjacent to an old scar (from http:// http://www.eyerounds.org)

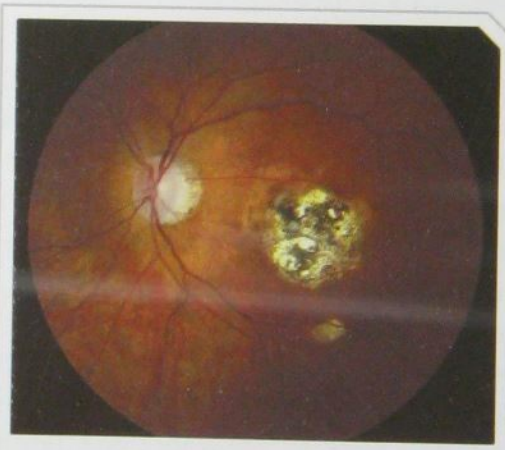


Fig. 10.8. Toxoplasmic posterior uveitis with a chorioretinal scar within the macula (from http:// http://www.eyerounds.org)

When lesion heals after treatment, it is replaced with a sharply demarcated atrophic scar with pigmented borders.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, fluorescein angiography, OCT, blood tests, serological studies, polymerase chain reaction (PCR).

Differential Diagnosis. Acute retinal necrosis, intraocular foreign body, uveitic glaucoma, uveitis associated with tuberculosis, sarcoidosis, or syphilis.

Treatment. The goal of medical therapy is to control the infection and prevent damage to the retina and optic nerve, thereby preventing vision loss.

The most common treatment of ocular toxoplasmosis includes a four-drug regimen consisting of Pyrimethamine (75—100 mg/day), Sulfadiazine (2—4 g/day), Folinic acid (5.0 mg) and systemic corticosteroids (Prednisone). In severe cases intravitreous injections of Dexamethasone are prescribed.

In cases of anterior uveitis topical corticosteroids and cycloplegics are used.

Prognosis for acquired toxoplasmosis in adults with strong immune systems is excellent if the macula is not involved. However, the prognosis for immunodeficient patients is not as positive and can be fatal, particularly if not treated.

When congenital toxoplasmosis is acquired during the first three months of pregnancy, the prognosis is poor. Afflicted chil-

dren die in infancy or suffer damage to their central nervous systems that can result in physical and mental retardation.

Complications of ocular toxoplasmosis include cataract, secondary glaucoma, vascular occlusions, scleritis, retinal gliosis, tractional retinal detachment, cystoid macular edema, macular scarring, optic atrophy, and choroidal neovascular membrane that can lead to severe loss of vision and blindness.

Prophylaxis. Toxoplasmosis can be prevented by cooking meat thoroughly; washing or peeling fruits and vegetables before eating; not drinking untreated water or milk; washing hands properly; cleaning contaminated surfaces with hot, soapy water; wearing gloves when gardening and washing hands thoroughly after contact with soil. Pregnant women and immunosuppressed persons should avoid changing cat litter if possible.

Syphilitic Uveitis

Syphilis is a sexually transmitted chronic systemic infection caused by the spirochete Treponema pallidum. According to the World Health Organization, the current incidence of syphilis is estimated as 12 million new cases per year. If left untreated, the disease progresses through four stages and may cause significant pathology in any major organ of the body.

Involvement of the eye is uncommon, being more frequent at the second or third stage of the disease and is often associated with delayed diagnosis and delayed treatment, which may result in irreversible visual loss and structural damage. The eyes are affected in approximately 10 % cases and the most common presentation of syphilis in the eye is uveitis. Other ocular manifestations include interstitial keratitis, scleritis, retinal vasculitis, neuroretinitis, and optic neuropathies.

Definition. Syphilitic uveitis is an inflammation of the uvea that occurs in patients with syphilis.

Etiology. Syphilis is caused by the bacteria Treponema pallidum, a small, highly motile, spiral gram-negative bacterium. Transmission is almost always through sexual contact, but it can also be passed on to a fetus from an infected mother during pregnancy or delivery, causing congenital syphilis.

Clinical Picture. Syphilis begins with a painless ulcer in the genital region, rectum or mouth. 4 to 10 weeks later, syphilis can lead to a number of other symptoms in various organs, including eyes. The course can be acute or chronic, involvement — unilateral or bilateral.

Acute anterior uveitis usually causes ocular pain, redness, photophobia, and vision loss due to the accumulation of inflammatory debris in the anterior segment of the eye. Posterior uveitis is often painless, although vitreal opacification and inflammatory involvement of the posterior segment structures can cause severe and often permanent vision loss, even when the eye is externally "quiet".

Complaints. Due to the varying degrees of presentation patients may complain of blurred vision, floating spots, light sensitivity, double vision, eye pain, redness and foreign body sensation.

Signs. The fundus is hazy due to vitritis (ground-glass appearance), on the periphery there are multifocal white pale-yellowish or grey chorioretinal infiltrates with atrophic centers and pigment proliferation (the fundus appears as if it was sprinkled with salt and pepper), swelling of the optic disc and adjacent retina, retinal hemorrhages, mild exudative retinal detachment, macular edema.

Methods of Examination. Thorough medical history, VA test, slit-lamp exam, ophthalmoscopy, perimetry, B-scan ultrasonography, fluorescein angiography, serologic tests.

Differential Diagnosis. Uveitis associated with tuberculosis, Lyme disease, herpetic disease, toxoplasmosis, sarcoidosis, Wegner's granulomatosis, cytomegalovirus retinitis, or intraocular lymphoma.

Treatment usually involves 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.

Topical, or periocular, and systemic corticosteroids (depends on the type and severity of uveitis) are adjunctive therapy to reduce ocular inflammation related to syphilis.

Prognosis. Syphilitic uveitis is a curable form of uveitis. Early detection and prompt treatment usually result in full visual recovery. If left untreated, uveitis will worsen and can lead to permanent and profound loss of vision.

Complications. Posterior synechiae, secondary glaucoma, cataract, retinal scarring, macular edema, and retinal detachment.

Prophylaxis. Practicing safe sex is the best way to prevent syphilis infection. Prompt treatment can prevent disease progression.

Tuberculous Uveitis

Tuberculosis (TB) is a chronic granulomatous infection with an increasing number of new cases every year that represents a major health problem worldwide. The World Health Organization currently estimates that nearly two billion people, or one-third of the world's population, are infected with tuberculosis, and that roughly 10 % of the infected people are symptomatic. TB affects the lungs in 80 % of patients, while in the remaining 20 % the disease may affect other organs, including the eye.

Ocular TB is usually not associated with clinical evidence of pulmonary tuberculosis, as up to 60 % of extrapulmonary tuberculosis patients may not have pulmonary disease. Primary ocular infection in the form of keratitis, conjunctivitis, and/or scleritis can occur, but is rare. Most commonly, TB presents as uveitis.

Definition. Tuberculous uveitis is a type of uveal tract inflammation that occurs in patients with tuberculosis.

Etiology. TB is a disease caused by airborne transmission and infection with the bacillus Mycobacterium tuberculosis. Ocular TB generally develops following hematogenous spread from a primary focus or immune-mediated hypersensitivity response to mycobacterial antigens; in rare cases, it can also occur as a result of a primary infection following epithelial injury.

Clinical Picture. TB uveitis has a variety of clinical manifestations affecting nearly every ocular tissue. These include granulomatous anterior uveitis, intermediate uveitis, chorioiditis, choroidal granulomas, retinal vasculitis, subretinal abscess, panuveitis, endophthalmitis and panophthalmitis (fig. 10.9, 10.10). The majority of cases are bilateral, and both anterior and posterior segnments are involved.

Complaints. Blurry vision and light sensitivity may be the only reported symptoms. Other complaints may include headache, redness of the eye, floaters, or flashes. Patients may also be asymptomatic.

Signs. Ophthalmologic sings depend on the part of the uvea involved in the inflammation and may include decreased VA, episcleral injection, KPs, epitheliod cells and giant cells within the aqueous humor, angle granulomas, iris nodules, posterior synechiae, snow banking, vitreous opacities, intraretinal hemorrhages, cotton-wool spots on the fundus, choroidal tubercles, macular edema, optic dis edema.

Methods of Examination. Thorough medical history, VA test, slit-lamp exam, ophthalmoscopy, tonometry, B-scan ultrasonography, fluorescein angiography, OCT, complete blood laboratory test, chest X-ray, tuberculin skin test.

Differential diagnosis includes other etiologies for uveitis including histoplasmosis, sarcoidosis, syphilis, toxoplasmosis, and acute retinal necrosis.

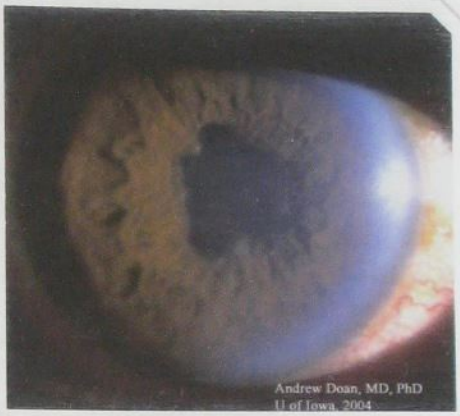


Fig. 10.9. Tubercular granulomatous anterior uveitis with keratic precipitates on the corneal endothelium, posterior synechiae, irregular pupil and Koeppe nodules on pupil margin (from http://www.eyerounds.org)

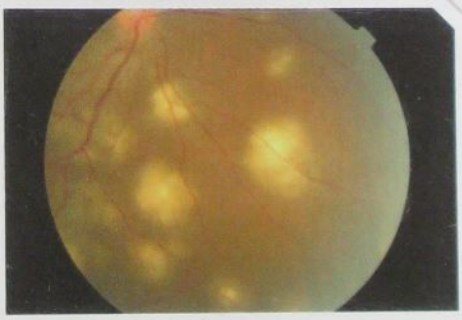


Fig. 10.10. Tubercular chorioiditis with multiple choroidal granulomas (from http://www.eyerounds.org)

Treatment. Tuberculous uveitis is a vision-threatening disease that inevitably leads to blindness if not properly diagnosed and treated.

The treatment of tuberculosis uveitis involves treatment of the underlying infection, whether it manifests as pulmonary or disseminated systemic disease. Ocular therapy alone is not curative and must be used only in conjunction with systemic therapy.

Anti-tuberculosis 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 two-drug (Rifampicin and Isoniazid) regimen for four to seven months.

Depending on the severity and site of inflammation, the ocular treatment includes topical cycloplegics, topical and/or oral corticosteroids (initially to control the inflam-

mation, and then tapered and stopped over 6—12 weeks). Immunosuppressive agents may be added when required.

Prognosis. Ocular TB and its complications cause moderate to severe visual impairment in the majority of affected eyes, especially in the eyes with posterior uveitis or panuveitis. Appropriate management of this condition can significantly recover visual loss.

Complications. Complicated cataract, secondary glaucoma, pupillary membrane, vitreous hemorrhage, cystoid macular edema, retinal scarring, retinal detachment, optic nerve atrophy, visual loss.

. *Prophylaxis.* Primary prevention of tuberculous uveitis involves prevention of exposures to actively infected individuals in order to prevent systemic infection.

4.4. Tumors

The commonest primary tumors of the uveal tract are nevi and malignant melanomas, which arise from melanocytes. Other types of tumor, which are rarer, can arise from blood vessels (hemangioma), nerves (neurilemmoma), smooth muscle (leiomyoma), as well as the nonpigmented and pigmented epithelia of the iris and ciliary body (cysts, adenoma, adenocarcinoma, medulloepithelioma). Other tumors of the uveal tract include metastases and choristomas (i.e. normal tissue in an abnormal site, for example, osseous choristomas).

Nevus

Definition. The uveal nevus is a benign tumor that arises from melanocytic cells. Uveal nevi occur most commonly in the choroid, but similar lesions also occur in the iris, ciliary body, and optic disc.

Etiology. A nevus is caused by cells called melanocytes, which produce the pigment that colors the skin, hair, and eyes. Most of the time, melanocytes are distributed evenly, but occasionally they form clusters, which leads to the development of nevi.

Clinical Picture. A nevus almost rarely causes symptoms. Choroidal nevi are often first detected during routine fundus examinations. Sometimes they can cause scotomas due to subretinal fluid, cystoid retinal edema or, rarely, choroidal neovascularization.

Complaints. Patients notice a hyperpigmented area on the iris that does not affect vision. Choroidal nevi are asymptomatic; however, they can cause blurry vision or blind spots in the vision field.

EYE FACTS

Nevus is a Latin word meaning birthmark or mole.

Sometimes it is called a *freckle in the eye*.

Uveal nevi are more commonly found in patients with blue eyes.

Signs. Iris nevi — hyperpigmented regions of the iris with minimal disruption of the normal iris architecture (fig. 10.11).

Choroidal nevi — "dark spots" that can be grey, blue-grey, greenish-grey or brownish-grey, sometimes they can be whitish ("amelanotic"). These spots are flat, they can be slightly raised, with indistinct margins, surface drusen are common. They usually measure between 1.5 to 5 mm in diameter and less than 2 mm in thickness.

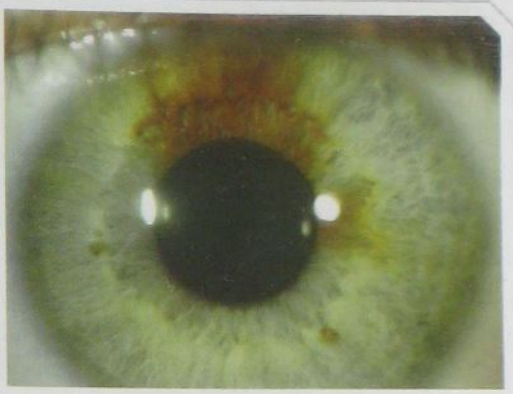


Fig. 10.11. Nevus of the iris

Methods of Examination. Slit-lamp exam, ophthalmoscopy, gonioscopy, B-scan ultrasonography, fluorescein angiography, OCT, color fundus photography.

Differential Diagnosis. Choroidal detachment, melanocytoma, melanoma.

NOTE!

If the choroidal nevus has orange pigmentation, if the nevus is leaking fluid, or has a thickness of 2 mm or more, it may be/or become a malignant choroidal melanoma.

Treatment. Uveal nevi do not require treatment, and it is generally not advised to surgically remove a nevus. In rare cases when an iris nevus has become a significant cosmetic problem, surgery may be possible. Annual ophthalmic examination and photographic documentation for nevi to check for growth or malignant progression are recommended for nevi in all locations.

Prognosis for all forms of nevi is mostly good as the condition is benign.

Complications. Occasionally nevi give rise to malignant melanoma.

Prophylaxis. There are no known preventable methods for uveal nevi. Wearing sunglasses with UV protection while outdoors is thought to be helpful but not proved.

Melanoma

Definition. Uveal melanoma is the most common primary intraocular malignant tumor arising in the uveal tract of the eye. The majority of these tumors arise in the choroid.

Etiology. Melanomas arise from pre-existing nevi, which are benign in appearance, and then transform into true cancers. The exact cause of this transformation is unknown. The predisposing factors include family history of choroidal melanoma, dysplastic nevus syndrome, xeroderma pigmentosum, and congenital ocular melanocytosis. Exposure to sunlight has been identified as a potential cause of the disease but there is no convincing evidence of this.

Clinical Picture. Choroidal melanomas remain asymptomatic for prolonged periods of time; they may be found incidentally during ophthalmoscopy.

It is of vital importance to differentiate choroidal nevus and melanoma. It is based on symptoms and ophthalmoscopic findings.

Uveal melanomas are characterized by rapid enlargement that influences visual functions. Patients notice a progressive decrease of vision, blind spots in the visual field, flashing lights, metamorphopsias, pain, bulging of the eye.

Ophthalmoscopic examination reveals lesions of irregular shape and color with orange pigment; its thickness is greater than 2.0 mm and diameter exceeds 7.0 mm; its margin touches the disc; subretinal fluid, which is associated with neovascularization.

NOTE!

The acronym **SPOTS** for suspicious choroidal melanoma:
S — symptoms;
P — position in the fundus;
O — orange color;
T — thickness more than 2 mm;
S — subretinal fluid.

One of these signs prompts suspicion of melanoma.

Complaints. Patients with melanomas may be asymptomatic at the initial stage or may present with a variety of symptoms, depending on the size, location, and extent of the lesion. Blurry vision, floaters, photopsias and visual field loss corresponding to the location of the lesion may occur. The patient may experience pain, bulging of the eyeball, change in the position or movement of the eyeball.

Signs. A small choroidal melanoma occurs as a black or dark grey choroidal mass with fairly well-defined borders. A typical early feature is the orange pigment on the surface of the tumor. A larger melanoma usually assumes a dome shape (fig. 10.12). An amelanotic melanoma is characterized by large, visible blood vessels in the substance of the tumor. A choroidal melanoma can eventually break through Bruch's membrane assuming a mushroom-shaped configuration.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, gonioscopy, B-scan ultrasonography, color fundus photography, fluorescein angiography, OCT, CT, MRI.

Differential Diagnosis. Uveal nevi, metastatic tumors, melanocytoma, disciform macular degeneration, hemangioma, intraocular foreign bodies.

Treatment of melanomas varies depending on the location and size of the melanoma. Generally, it involves local radiation therapy (plaque brachytherapy), transpupillary thermotherapy, trans-scleral local resection, enucleation of the eyeball.

Prognosis. Iris melanoma is relatively non-aggressive and has the best prognosis regarding mortality, especially for younger patients. On the contrary, choroidal melanomas have the worst prognosis with a five-year mortality rate around 40 %, and an eventual 50 % 10-year mortality rate, primarily due to aggressive metastasis.

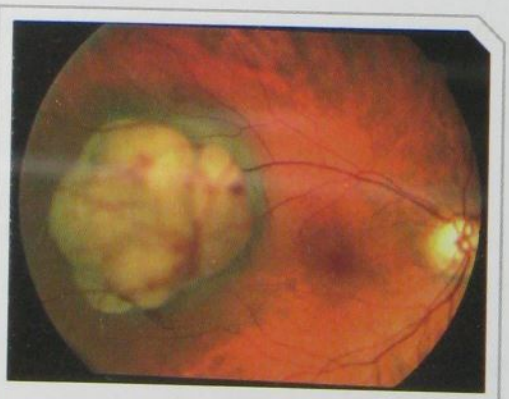


Fig. 10.12. Choroidal melanoma (from http://imagebank.asrs.org/)

Complications. Glaucoma, vision loss, loss of the eyeball, metastasis/spread that most often results in death.

Prophylaxis. Annual eye examination and avoiding excessive exposure to sunlight are helpful.

Review:

1. Key Points

Diseases of the uvea according to the origin can be *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*.

The main symptoms of uveal diseases are decreased VA, blurred vision, eye pain and redness, photophobia, floaters, myosis. The main examination methods are VA, slit-lamp exam, ophthalmoscopy, gonioscopy, tonometry, perimetry.

Congenital anomalies of the uvea — coloboma, albinism, aniridia, anisocoria, heterochromia, persistent pupillary membrane. It may not require treatment unless it influences the vision, then surgery should be performed.

Inflammatory diseases of the uvea (uveitis) according to anatomical classification (developed by IUSG) can be *anterior, intermediate, posterior, panuveitis*; according to onset (based on SUN) — *sudden* or *insidious*; accoring to the duration — *limited* (< 3 months' duration) or *persistent* (> 3 months' duration); according to the course — *acute* (sudden onset and limited duration), *recurrent* (repeated episode separated by inactivity > 3 months) or *chronic* (persistent uveitis with relapse in < 3 months). Anterior uveitis according to the morphologic classification can be *nongranulomatous* or *granulomatous*.

Other intraocular inflammatory disorders include *endophthalmitis, panophthalmitis, sympathetic ophthalmia.* Uveitis may be ocular manifestation of systemic diseases such as Lyme disease, toxoplasmosis, syphilis, tuberculosis, etc.

Symptoms of uveitis depend on the site of inflammation. For anterior uveitis the most common are ocular pain and redness, myosis, photophobia; whereas intermediate and posterior types of uveitis present with complains of painless blurred vision and floaters. *Treatment methods* depend on the type of uveitis and the underlying condition and usually include steroids (topical, intravitreal or systemic), mydriatics, analgesics, topical or systemic antibiotics, or antiviral, anti-parasitic, or antifungal medicines. The most severe cases may require immunosuppressive drugs. *Complications of uveitis* may include glaucoma, cataract, neovascularization, macular edema, retinal detachment, phthisis bulbi, vision loss.

Tumors of the uveal tract are benign, malignant or metastatic. The most common are nevi and melanomas that must be differentiated in order to save the patient's vision or even life. Whereas uveal nevi require only regular observation, treatment of melanomas involves local radiation therapy (plaque brachytherapy), transpupillary thermotherapy, transscleral local resection, enucleation of the eyeball.

2. Diagrams

UVEAL DISEASES
Congenital-disorders
Coloboma
Albinism
Aniridia
Anisocoria
Polycoria
Heterochromia
Persistent pupillary membrane
Uveitis
According to the location
Anterior
Granulomatous
Non-granulomatous
Intermediate
Posterior
Panuveitis
According to the onset
Sudden
Insidious
According to the duration
Limited
Persistent
According to the course
Acute
Chronic
Recurrent
According to the etiology
Exogenous
Endogenous
Intraocular inflammation
Endophthalmitis
Panophthalmitis
Sympathetic ophthalmia
Tumors
Benign
Cyst
Nevus
Hemangioma
Leiomyoma
Malignant
Epithelioma
Melanoma
Metastatic

3. The Review Questions

A. Control Questions

1. What is the classification of uveal tract diseases?
2. What are the main symptoms of uveal disorders?
3. What are the congenital anomalies of the uvea and their symptoms?
4. What is uveitis and its classification?
5. What is the difference between granulomatous and non-granulonatous uveitis?
6. What is acute iridocyclitis, its differentiation from acute conjunctivitis and angle-closure glaucomatous attack?
7. What are the treatments methods of acute iridocyclitis?
8. Chorioiditis, its symptoms, signs, and treatment methods.
9. Endophthalmitis, its etiology, clinical picture, and treatment methods.
10. Ocular manifestations of Lyme disease, clinical pictures, treatment methods, and preventive measures.
11. Ocular toxoplasmosis, clinical pictures, treatment methods, and preventive measures.
12. Syphilitic uveitis, its clinical picture, treatment methods, and preventive measures.
13. Tuberculous uveitis, its clinical picture, treatment methods, and preventive measures.

B. Tests

1. The main examination methods of the uveal tract diseases are:

A. VA test
B. Slit-lamp exam
C. Ophthalmoscopy
D. Tonometry
E. Perimetry
F. Gonioscopy

2. What are the main clinical signs of granulomatous uveitis?

A. Insidious onset
B. Acute onset
C. Mutton-fat KPs, large, greasy
D. Myosis
E. Vitreous haze
F. Iris nodules

3. What are the main clinical signs of nongranulomatous uveitis?

A. Insidious onset
B. Acute onset
C. KPs are small, fine
D. Myosis
E. Vitreous haze
F. Iris nodules

4. What are the main clinical signs of acute iridocyclitis?

A. Acute pain
B. Floaters
C. Myosis
D. KPs
E. Circumlimbal flush
F. Elevated IOP

5. **What are the main treatment methods of acute iridocyclitis?**
   A. Mydriatic eye drops
   B. Myotic eye drops
   C. Topical steroids
   D. Systemic analgesics
   E. Topical antibiotics
   F. Warm compresses

6. **What are the main complications of anterior acute uveitis?**
   A. Cataract
   B. Posterior synechiae
   C. Macular edema
   D. Secondary glaucoma
   E. Retinal detachment
   F. Corneal opacity

7. **What are the main clinical signs of posterior uveitis?**
   A. Acute pain
   B. Vitreous opacities
   C. Myosis
   D. KPs
   E. Scotomas
   F. "Cotton balls" on the fundus

8. **What are the main clinical signs of endophthalmitis?**
   A. Corneal edema
   B. KPs, cells, and flare in the anterior chamber
   C. Hypopion
   D. Low IOP
   E. High IOP
   F. Ciliary injection

9. **The treatment of endophthalmitis includes:**
   A. Mydriatic eye drops
   B. Topical steroids
   C. Topical antibiotics
   D. Systemic antibiotics
   E. Systemic analgesics
   F. Vitrectomy

10. **What signs are suspicious of choroidal melanoma?**
    A. Blurred vision
    B. Irregular shape of the lesion
    C. Orange color of the lesion
    D. Thickness is greater than 2 mm
    E. Margin touching the optic disc
    F. Subretinal fluid

C. Clinical Cases

Case 1

A 38-year-old man complains of right eye pain with redness for 5 days' duration with associated photophobia, excessive tearing, and reduced vision. On examination there was no significant findings in his left eye. On the right eye his best-corrected visual acuity (BCVA) was 0.5. He had circumcorneal injection, diffuse KPs, cells grade 4+ and flare grade 3+ in the anterior chamber with posterior synechiae and myosis. On palpation — severe ciliary pain. IOP OD 13 mm Hg. The fundus appeared normal. The patient didn't have any clinical features suggestive of systemic disorder. Define the diagnosis.

Case 2

An 11-year-old boy presented with bilateral blurred vision, ocular pain that worsens at night, photofobia, and red eyes. One month before he had an upper respiratory tract infection, including cough, sore throat, and rhinitis. On ocular examination, his

BCVA was 0.9, IOP — 14 mm Hg bilaterally. On palpation — severe ciliary pain. Slit-lamp biomicroscopy showed ciliary injection, 1+ cells in the anterior chamber, fine KPs, narrowed and irregular pupils, posterior synechiae. Make a diagnosis.

Case 3

A 40-year-old woman has been suffering from rheumatism for 5 years. A week ago, after an attack of rheumatism she noticed pain in the left eye that worsened at night, redenning of the eye, blurry vision, photophobia, and tearing. On examination, VA OS — 0.6, ciliary pain on palpation, ciliary injection, flare and cells in the anterior chamber, KPs, discoloration of the iris, and smoothing of its pattern, pupillary miosis, an irregularly shaped pupil, synechiae, hypotension, anterior vitreous inflammatory cells. Make a diagnosis.

Case 4

What are the emergency care options you must take to treat a patient with acute iridocyclitis?

Case 5

2 weeks ago a 48-year-old man noticed a decrease and blurring of vision, scotoma in the field of vision and floaters in the left eye. Symptoms were not associated with ocular pain or eye redness. On examination OS, VA — 0.4, anterior segment is without changes, inflammatory cells in the vitreous, on the fundus — yellow-white "cotton balls" with indistinct borders, retinal hemorrhages, optic disc edema. What is the diagnosis?

Case 6

A 53-year-old man with no known systemic illness presented with a 2-day-old penetrating injury with a tree branch to the right eye. He complained of loss of vision, severe deep ocular pain, redness of the eye, extreme sensitivity to bright light, tearing, swelling of the eyelid and headache. On examination, VA — counting fingers at 2 m, lid edema, proptosis, decreased ocular motility. Slit-lamp biomicroscopy of the right eye revealed conjunctival injection, 3 mm from the limbus at 9:00, which was probably the entry wound, chemosis of the conjunctiva, corneal edema. The anterior chamber had grade 4 cells and flare with an exudative membrane and 1 mm hypopyon. The lens had nuclear sclerosis grade 1. On ophthalmoscopy, red reflex was decreased, severe vitritis. B-scan ultrasonography did not reveal any intraocular foreign body. What is the diagnosis?

Case 7

A 37-year-old man presented with 2 weeks of conjunctival redness and burning discomfort of the left eye. Roughly 7 days after noticing these initial symptoms, he developed blurry vision of the left eye with floaters and the appearance of spider webs. Slit-lamp examination of the left eye showed marked conjunctival injection with fine KPs and posterior synechiae. The fundus appeared like "ground glass" due to vitreous opacity, multiple yellow infiltrates of the retina with atrophic centers and pigment

proliferation (the fundus appears as if it was sprinkled with salt and pepper), exudates along the retinal vessels, and swelling of the optic disc. Make a diagnosis.

Case 8

A 34-year-old patient presented with a decrease of vision, blurry vision, and mild pain in his left eye of 3-week duration. On examination, BCVA OD — 1.0, OS — 0.05, IOP OU — normal. Slit-lamp biomicroscopy of the anterior segment of the left eye showed ciliary congestion of the conjunctiva, transparent cornea, a deep anterior chamber without cells in the aqueous humor, regular iris structures, and a normal pupil with regular direct and indirect light reaction. Both lenses were clear and ophthalmoscopy of the posterior segment was unremarkable in the right eye. Ophthalmoscopy of the left eye showed optic nerve head hemorrhage and edema, yellowish-white, subretinal exudations with indistinct margins of approximately 7 disc diameters in its greatest dimension in the macular region. Chest X-ray showed infiltration in the top of the left lung with coexisting fibrotic and calcified lesions. The tuberculin test was positive at 48 hours and the erythrocyte sedimentation rate (ESR) was 50 mm/h. What is the diagnosis?

Case 9

A 42-year-old woman complains of blurred vision and floaters in the left eye, gradually worsening over the past two weeks. The changes have been painless, and the eye has not become red or irritated. No past ocular surgery or trauma, no contact lens use. Ocular symptoms were preceded by mild flu-like symptoms. OS — BCVA — 0.1, slight conjunctival injection, fine KPs and 1+ cell in the anterior chamber, the pupils were equally round and reactive to light. Dilated fundus examination revealed an elevated yellow-white central macular lesion with indistinct margins and pigmentation. The vitreous is hazy due to inflammatory cells. The fundus had the "headlight in the fog" appearance. Laboratory evaluation showed elevated toxoplasma IgG levels. Define the diagnosis.

Case 10

A 52-year-old man noticed a brownish spot on his right iris. He had no pain, no visual disturbance or any other symptoms. Slit-lamp examination showed a dark brown well-circumscribed flat nodule of the right eye iris, which was sized as a small seed. It was located in the inferior temporal quarter without distortion of the surrounding iris architecture or pupil. There was no prominent vascularity to the lesion or extension into the surrounding trabecular meshwork. Ultrasonography and MRI didn't reveal any signs of tumor spread. What is the diagnosis and treatment measures?

Case 11

A 58-year-old woman complains of progressive decrease of vision, flashes, floaters and pain in her right eye. On examination, vision of OD was counting fingers. Ophthalmoscopic and ultrasound examination revealed an orange-pigmented, dome-shaped elevation centered at the inferior arcade and measuring 3 mm in thickness and 4 disc diameters in the maximal diameter. The lesion margins are within 3 mm of the optic disc. There was subretinal fluid present. What is the diagnosis?

CHAPTER 11

Lens Diseases

OBJECTIVES

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

— know the main anomalies of the lens;
— know the basic diagnostic methods;
— distinguish types of congenital and acquired cataracts;
— describe association of cataracts with aging, trauma, medications, systemic and ocular diseases;
— evaluate and manage patients with lens abnormalities and cataracts;
— explain the principles of cataract surgery techniques;
— know the basic ways for postoperative aphakia correction and types of IOL.

Plan:

1. Lens Pathology Classification

Lens pathology according to the time of onset may be congenital or acquired. Diseases of the lens may affect its position, shape, and structure.

1.1. Anomalies of Lens Position: ectopia lentis — displacement (subluxation) or dislocation (luxation) of the lens.

1.2. Anomalies of Lens Size and Shape: congenital aphakia, lenticonus, lentiglobus, lens coloboma, microspherophakia, remnants of the fetal vasculature system of the lens.

1.3. Pathology of Lens Structure — Cataracts:
1.3.1. Congenital and Juvenile Cataract
1.3.2. Acquired Cataract
1.3.3. Secondary Cataract

2. Symptoms of Lens Diseases

— Decrease of vision to total blindness.
— Increase of refractive errors.
— Increase of optical aberrations.

3. Examination Methods

— Visual acuity testing.
— Examination with transillumination.
— Focal illumination with a slit lamp.
— Ophthalmoscopy.
— Ultrasound studies.

4. Lens Diseases

The lens is a biconvex and transparent organ, held in its position behind the iris by the suspensory ligaments (zonules). The zonules attach the equator of the lens to the ciliary body. Diseases may affect the position, size, shape, and structure of the lens. Abnormalities of lens position, size, and shape may lead to refractive errors of the eye, impairing patient's vision and increasing optical aberrations. Beside this, such kinds of anomalies are often associated with lens opacities.

4.1. Anomalies of Lens Position

Ectopia Lentis

Definition. Ectopia lentis is a displacement of the lens out of its normal anatomic position which may be partial (subluxation, displacement) or total (luxation, dislocation). Subluxated lens, although being displaced, still remains in the pupillary area due to rupture or extension of the part of Zinn ligaments. Complete dislocation of the lens, which is a result of total ligament rupture, is quite a rare condition and appears in two variants — dislocation into the anterior chamber and into the vitreous. In the latter case the lens cannot be seen in the pupillary area (this condition is called *aphakia*).

Etiology of ectopia lentis includes two groups of causes: hereditary and acquired.

Hereditary ectopia may be seen in cases of:

— familial ectopia lentis (simple) — isolated and monosymptomatic, inherited as an autosomal dominant trait,
— Marfan's syndrome,
— Weill—Marchesani syndrome,
— homocystinuria (metabolic disease).

Acquired causes include:

— trauma (the most frequent cause),
— pseudoexfoliation (in aged patients),
— cilliary body tumor,
— buphthalmos.

Signs and Symptoms. Monocular diplopia, impairment of vision due to refraction changes (severe astigmatism, high hyperopia).

General ophthalmic findings:

— *for subluxation:*
  • iridodonesis (tremulous iris);

- phacodonesis (tremulous lens on eye movement);
- the anterior chamber becomes deep or irregular;
- irregular pupil (sometimes);
- marked displacement of the lens equator can be seen in the pupillary area (fig. 11.1);

— *for anterior displacement:*

- pupillary block with angle-closure glaucoma.

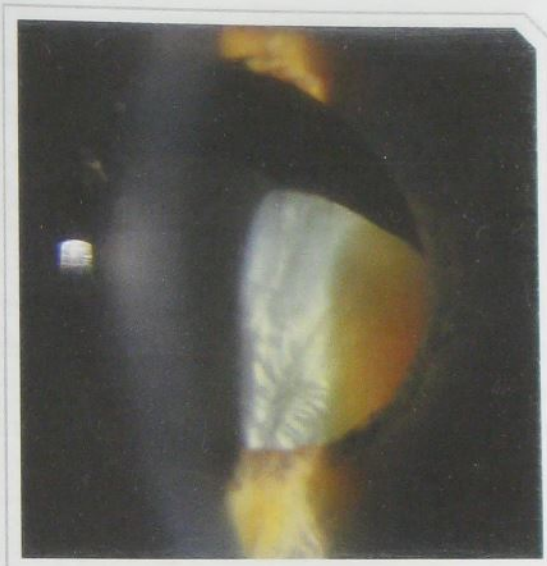


Fig. 11.1. Inferior displacement of opaque (cataractous) lens due to contusion. The superior part of the equator is seen in the pupil

Marfan's syndrome is an autosomal dominant disorder of collagen synthesis characterized by skeletal, cardiac, and ocular findings. *Systemic findings* include tall, thin stature, mitral valve prolapse, dilated aortic root and aortic dissection, arachnodactyly, hyperextensible joints. *Ophthalmic findings* are bilateral ectopia lentis (mainly superotemporal), myopia, increased incidence of retinal detachment (fig. 11.2).

Weill—Marchesani syndrome is an autosomal recessive condition characterized by short stature, small hands and feet (brachydactylia). *Ophthalmic findings*: the lens is abnormally round and often very small (microspherophakia); it is usually eccentric and displaced inferiorly.

Homocystinuria is an autosomal recessive systemic disorder of amino acid metabolism resulting in lenticular zonular fragility, seizures, and an increased risk for thromboembolic events. *Systemic findings* are characterized by oligophrenia, osteoporosis, and skeletal deformities. *Ophthalmic findings:* lens displacement is usually medial and inferior; torn zonule fibres appear as a "permanent wave" on the lens.

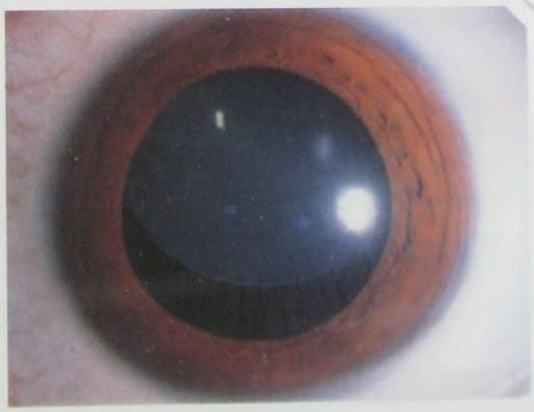


Fig. 11.2. Superior lens subluxation in a Marfan's syndrome patient. The capsular lens zonules remain stretched but intact for the most part (from *Ophthalmology. A short Textbook* / G. K. Lang, M.D. — Thieme, Stuttgart—New York, 2000)

The treatment of lens position anomalies includes:

— therapeutic options — optic correction of refractive errors with spectacles or contact lenses;

— surgical options — lensectomy with intraocular lens (IOL) implantation in cases of optical correction inability or presence of attendant cataracts.

4.2. Anomalies of Lens Size and Shape

Anomalies of lens size and shape are very rare congenital conditions connected mostly with eye malformation. To this group of diseases we refer the following disorders:

— congenital aphakia,
— lenticonus,
— lentiglobus,
— lens coloboma,
— microspherophakia,
— remnants of the fetal vasculature system of the lens.

Congenital aphakia is a very rare anomaly characterized by total absence of the lens.

Lenticonus is a localized cone-shaped deformation of the anterior (anterior lenticonus) or posterior (posterior lenticonus) lens surface. Posterior lenticonus is more common than the anterior one and is usually unilateral. Anterior lenticonus, which is often bilateral, may be associated with Alport syndrome (kidney disease accompanied by sensorineural hearing loss and lens shape anomalies) (fig. 11.3).

Lentiglobus is rare, usually unilateral, generalized hemispherical deformation of the lens, which may be associated with posterior polar lens opacity. *The symptoms* of lenticonus and lentiglobus include reduced visual acuity due to myopic refraction.

Fig. 11.3. Anterior and posterior lenticonus (from *Rapid Diagnoses in Ophthalmology: Lens and Glaucoma* / J. Schuman, V. Christopoulos, D. Dhaliwal et al. — MOSBY, 2008)

Lens coloboma is a wedge-shaped defect or indentation of the lens periphery that occurs as an isolated anomaly typically located inferiorly and may be associated with colobomas of the uveal tissue.

Microspherophakia is a developmental abnormality in which the lens is small in diameter and spherical. The entire lens equator can be seen with the slit lamp when the pupil is dilated. The spherical shape of the lens results in increased refractive power and appearance of high myopia. Microspherophakia is most often seen as a part of Weill—Marchesani syndrome as well as an isolated hereditary abnor-

mality or, occasionally, in association with Peters anomaly, Marfan's syndrome, Alport syndrome, Lowe syndrome, or congenital rubella.

Mittendorf dot is a remnant of the posterior pupillary membrane of the tunica vasculosa lentis. It is a common anomaly observed in many healthy eyes as a small, dense white spot generally located inferonasally to the posterior pole of the lens. Sometimes a Mittendorf dot is associated with a fibrous remnant of the hyaloid artery projecting into the vitreous. This anomaly usually does not require any treatment.

Treatment options in case of these anomalies depend on the degree of vision impairment and abilities of optic correction of refractive errors. Surgical treatment is indicated in cases of ineffective optical correction and developed opacification of the media. However, making the choice, the doctor should always consider the risks of further surgical complications.

4.3. Cataracts

Cataract is the presence of any persistent opacity in the lens. This disease is the most common cause of treatable blindness in the world.

Classification

Despite numerous classifications of cataracts, there is still no one that would satisfy ophthalmologists completely. Usually cataracts are classified according to following criteria: time of onset; morphology (opacity localization); etiology and stages of maturation.

By the *time of onset* all cataracts are divided into:

— congenital and juvenile,

— acquired.

Congenital cataracts are detected at birth; juvenile cataracts appear during the first 12 years of life. Acquired cataracts may be detected in adulthood.

By *morphology* acquired and congenital cataracts are divided in different ways.

The following morphological types of *age-related cataracts* are distinguished:

— cortical,

— nuclear,

— posterior subcapsular,

— mixed.

Morphological changes in *congenital and juvenile cataracts* are classified as follows:

I. Total (complete) — the lens is completely opaque or hazy at birth.

II. Partial (incomplete) — opacities localized in a definite part of the lens:

A. Anterior and posterior polar cataracts.

B. Zonular cataracts:
1. Lamellar.
2. Sutural (stellate).
3. Nuclear.
4. Coronary.
5. Blue dot (cerulean).

C. Membranous cataracts.

Etiological classification of acquired cataracts:

— age-related (senile) cataract,
— traumatic cataracts (contusion, penetrating injury, infrared radiation, alcali burn),
— metabolic cataracts (diabetes mellitus, Wilson disease),
— complicated cataracts (chronic anterior uveitis, retinal vasculitis, retinitis pigmentosa),
— toxic cataracts:
• corticosteroid-induced,
• miotic-induced,
• copper- and iron-induced,
— radiation cataract,
— electrical cataract,

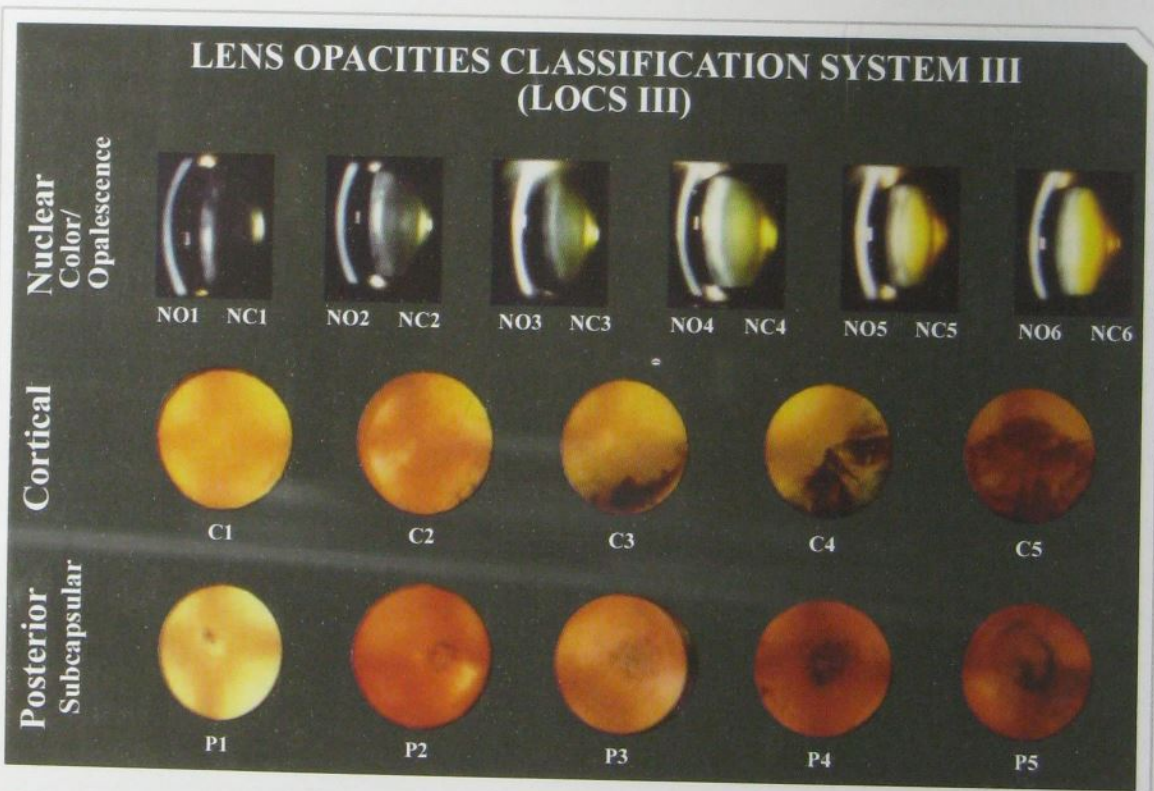


Fig. 11.4. Lens Opacities Classification System III (from *Arch Ophthalmol* — Vol. 111 — June, 1993)

— postoperative cataracts (following vitrectomy or glaucoma penetrating surgery),
— cataracts associated with systemic diseases (atopic dermatitis, myotonic dystrophy, tetany, Fabry's disease),
— cataracts associated with miscellaneous syndromes (Down's syndrome, Lowe's syndrome).

By the stage of maturation senile cataracts are usually grading into:
— incipient cataract,
— immature cataract,
— mature cataract,
— hypermature cataract.

Cataract grading systems have developed significantly during the last decades. They transformed from simple assessment by direct ophthalmoscopy to more up-to-date methods such as the Lens Opacities Classification System III (LOCS III) (1993) or Age-Related Eye Diseases Study (AREDS) (2008), where slit-lamp examination is compared to a standard set of photographs (special sets for nuclear, cortical, and posterior subcapsular) (fig. 11.4, 11.5).

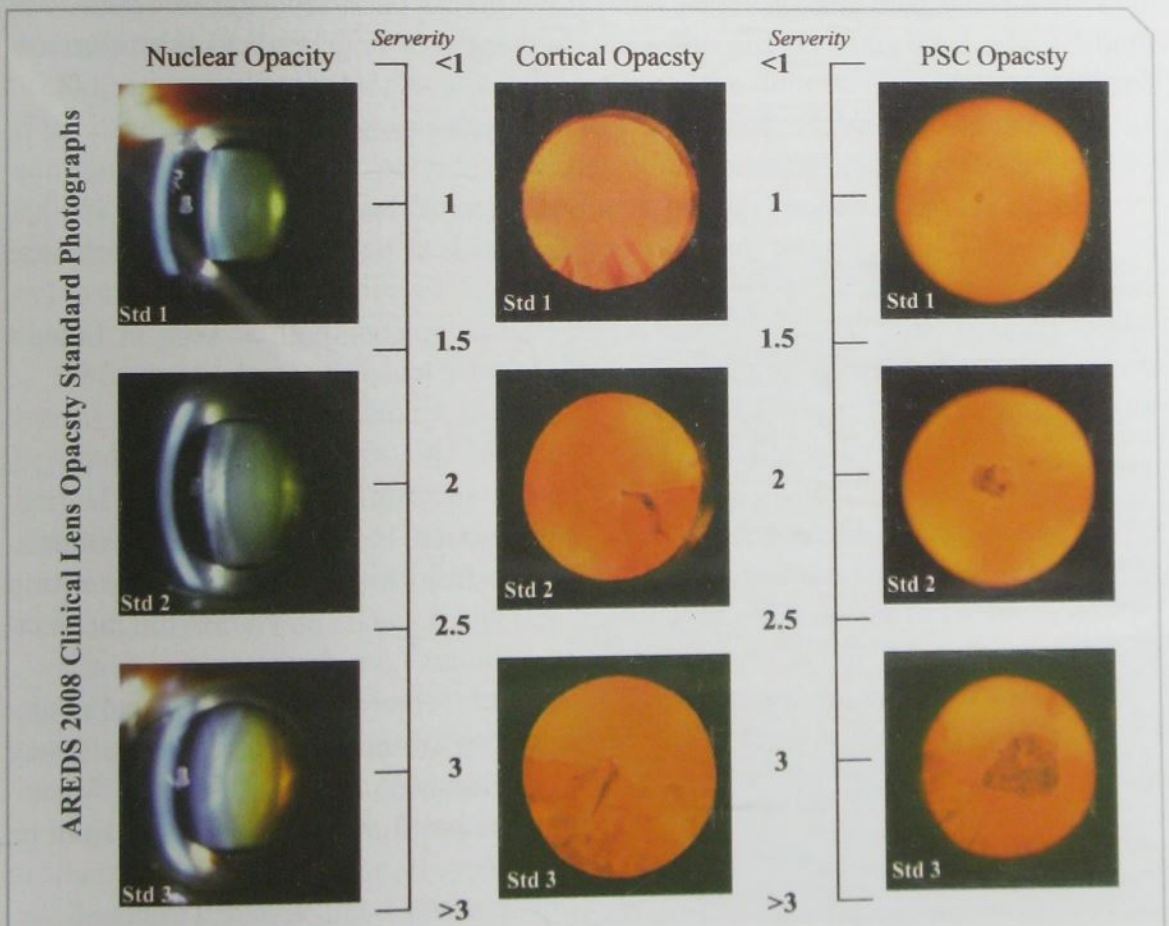


Fig. 11.5. The cataract grading system AREDS (2008)

Congenital Cataracts

Congenital cataracts occur due to any disturbance in the process of normal growth of the lens. Congenital and developmental opacities assume very variable appearance and minimal opacities (without visual disturbance); they are common in normal population. Otherwise, significant opacification of the central areas of the lens or total cataracts may lead to great vision problems.

Etiology. Among numerous predisposing factors in congenital cataract development, the following play the main role:

I. Heredity.

Genetically-determined cataract is due to chromosomal aberrations. About one-third of all congenital cataracts are hereditary — inherited as an autosomal dominant trait: cataract pulverulenta (nuclear), zonular cataract (also occurs as non-familial), coronary cataract and total soft cataract (may also occur due to rubella).

II. Maternal factors.

1. Malnutrition during pregnancy (non-familial zonular cataract).

2. Infections. Maternal infections like rubella are associated with cataract in 50 % cases as well as mumps, toxoplasmosis, hepatitis, CMV-infection.

3. Drug ingestion. Congenital cataracts have also been reported in children of mothers who have taken certain drugs during pregnancy (e.g., thalidomide, corticosteroids).

4. Radiation. Maternal exposure to radiation during pregnancy.

III. Fetal or infantile factors.

1. Deficient oxygenation (anoxia) owing to placental hemorrhage.

2. Metabolic disorders of the fetus or infant such as galactosemia, galactokinase deficiency, and neonatal hypoglycemia.

3. Cataracts associated with other congenital anomalies, e.g., as seen in Lowe's syndrome, dystrophic myotony, etc.

4. Birth trauma.

5. Malnutrition in early infancy.

IV. Idiopathic. About 50 % cases are sporadic and of unknown etiology.

Symptoms and signs. As has been mentioned above, the degree of vision impairment in congenital cataracts varies considerably from total absence of any vision problems to loss of vision up to light perception and depends mostly on the localization and density of opacities in the lens.

In case of total and nuclear cataracts, especially if they are bilateral, visual acuity is decreased significantly. But even if lens opacities are not so severe, mothers may mention changes in their baby's behavior and appearance:

— an opaque dot or a disk can be seen in the pupillary area; the pupil may be totally white (the latter is called leukocoria);
— strabismus and nystagmus may develop;
— at two months of age the baby doesn't fix his/her gaze at relatives' faces or bright toys, doesn't watch or follow objects;

— in case of monocular vision loss the baby constantly turns to the same side while watching a toy.

In *total* cataracts all portions of the lens are completely opaque.

Anterior and posterior polar cataracts involve the lens capsule in the anterior or posterior pole of the lens. They are sometimes associated with a localized anatomical abnormality in the region (i.e. posterior polar cataracts are commonly associated with posterior lenticonus). They may cause severe visual symptoms, however, they are usually stable, and patients may do well with conservative measures. The familial type is bilateral and inherited as an autosomal-dominant trait.

In *zonular cataracts* only one region or zone of the lens is opaque. They may be stationary or progressive. Subtypes of zonular cataracts are: lamellar, sutural (stellate), nuclear, coronary, blue dot (cerulean) cataract.

Lamellar cataract is the most common type of congenital cataract. Such cases are usually bilateral and symmetric, and the density of opacification may vary considerably. Less opaque lamellar cataracts may be compatible with good vision and require minimal medical intervention (e.g., optical correction, therapeutic mydriasis).

These cataracts may be inherited as an autosomal-dominant trait, but in some cases they may be a result of some intrauterine toxic agent, affecting only the layer of cells developing at the time of fetal exposure (fig. 11.6).

Sutural or stellate cataracts affect the region of the sutures. They may be Y-shaped if the cataract occurs at the intrauterine stage of development as the sutures have this configuration during this period. Anterior sutural cataracts are Y-shaped; posterior sutural cataracts are shaped like an inverted Y. Sutural cataracts that develop later on have a more stellate shape.

Nuclear cataracts are usually bilateral and involve the fetal or embryonal nucleus. They may be inherited as an autosomal-dominant, autosomal-recessive or X-linked trait and are associated with infections (e.g., rubella).

Coronary cataracts are radial, club-shaped discrete opacities located in the cortex. The term "coronary" means that their appearance is like the top of the crown. Because of their peripheral location, they do not decrease visual acuity. Coronary cataracts are dominantly inherited and have been described in cases

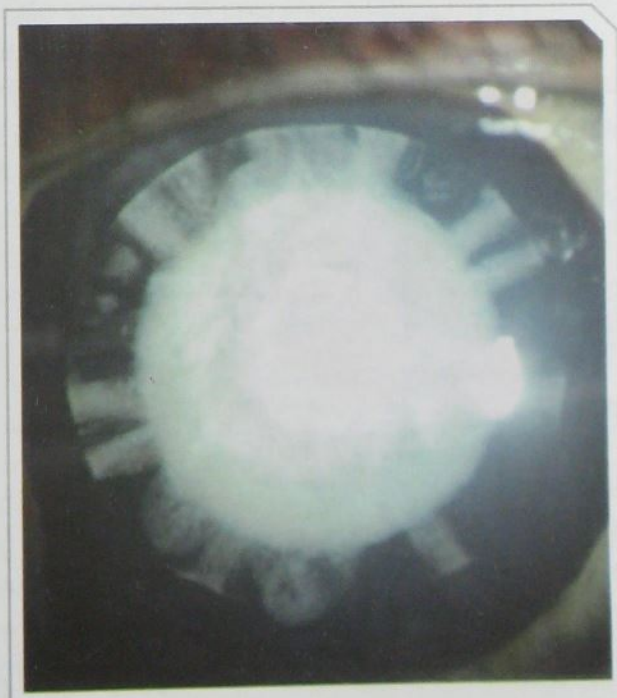


Fig. 11.6. Lamellar cataract

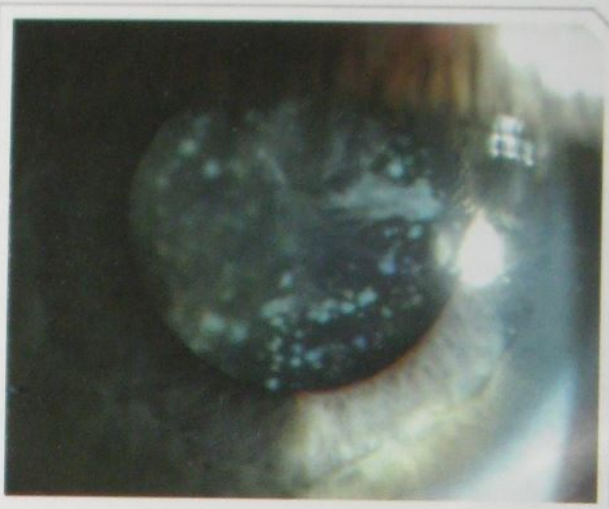


Fig. 11.7. Blue dot cataract associated with cortical cataract

of Down's syndrome and myotonic dystrophy.

Blue dot (cerulean) cataracts consist of small, discrete opacities that have a distinct bluish hue. These opacities are located in the cortex, are non-progressive, and do not cause visual symptoms. They may be present together with other congenital cataracts (fig. 11.7).

Membranous cataracts are thin but dense and contain fibrous tissue. They may occur when lens proteins are reabsorbed (e.g., traumatized lens), such that the anterior and posterior lens capsules fuse producing a dense membrane.

Examination methods in babies and children include visual acuity testing, focal and slit-lamp ocular examination, and ophthalmoscopy. In some cases methods like fixation reflex, visually evoked response, and optic-kinetic nystagmus (OKN) may be amended. The aims of thorough examination are working out prognostic factors, indications, and timing of surgery.

Differential diagnosis. Congenital cataracts with leukocoria are to be differentiated from various conditions presenting with this sign such as retinoblastoma, retinopathy of prematurity, persistent hyperplastic primary vitreous (PHPV), etc.

Treatment. Surgical treatment is the only effective method for cataract treatment. In pediatric cataracts the decision about surgery should be developed basing on thorough analysis of many factors such as: current visual functions, density of the cataract, unilateral or bilateral cataract, time of onset, associated ocular and systemic defects, etc. General approach to pediatric cataract surgery should be as follows:

Partial cataracts and small central cataracts, which are visually insignificant and non-progressive, can safely be observed or treated non-surgically with optical correction (if required).

Bilateral dense cataracts should be removed early (within 6 weeks of birth) to prevent amblyopia development.

Unilateral dense cataracts should be removed as early as possible after birth. However, it must be remembered that visual prognosis in these cases is poor even after successful surgery because proper correction of aphakia and prevention of amblyopia in infants is an extremely hard task considering further growth of the eyeball.

Surgical procedures include phacoemulsification or extracapsular cataract extraction (details of these are described further in the chapter). Post-operative aphakia should be corrected with intraocular lenses (IOL), in some cases (elder children) it may also be corrected with contact lenses or spectacles.

Acquired Cataracts

In acquired cataracts opacification of the lens fibres occurs under the influence of various causative factors like aging, trauma, metabolic disorders, different physical (radiation, electricity) and toxic agents, etc. The most common type of acquired cataract, which takes about 90 % of all cases, is age-related (senile) cataract.

Etiology of senile cataract still remains unknown, but several predisposing factors for lens opacity development shown in multiple studies are the following: heredity, ultraviolet irradiation from sunlight, dietary factors (deficiency of certain proteins, amino acids, vitamins), dehydrational crisis (due to diarrhea, cholera), smoking.

Symptoms and signs. The clinical picture of senile cataract is determined by its morphological type (cortical, nuclear or subcapsular) and degree of maturation but the main complaint of such patients is gradual decrease of vision which they have observed during several months to several years and even decades.

Nuclear cataracts tend to progress slowly. Usually they develop bilaterally but asymmetrically. Impairment of distant vision is more marked than near vision, at early stages refraction changes are known as myopic shift (lenticular myopia), so presbyopic patients can read without spectacles because of some improvement of near vision. Other symptoms of nuclear cataract may be monocular diplopia, disorders in color perception (especially for blue color).

Cortical cataracts in contrast to nuclear cataracts progress relatively rapidly, are usually bilateral, and often asymmetric. Their effect on the visual function varies greatly depending on the location of opacification in relation to the visual axis. At the daytime, when the pupil is constricted, patients report better vision. A common symptom of cortical cataracts is glare from intense focal light sources, such as car headlights. Monocular diplopia may also be present. At early stages of cataract the patients may perceive colored halos around light sources.

Posterior subcapsular cataracts are located in the posterior cortical layer and are usually axial. The patient often complains of glare and poor vision under bright lighting conditions because opacity obscures more of the pupillary aperture when miosis is induced by bright lights, accommodation, or miotics. Near visual acuity tends to be reduced more than distance visual acuity. Some patients experience monocular diplopia.

Objective signs in cataracts vary according to cataract maturation.

1. Incipient cataracts. The first signs of cortical cataract formation visible with the slit-lamp biomicroscope are vacuoles in the anterior or posterior cortex (fig. 11.8).

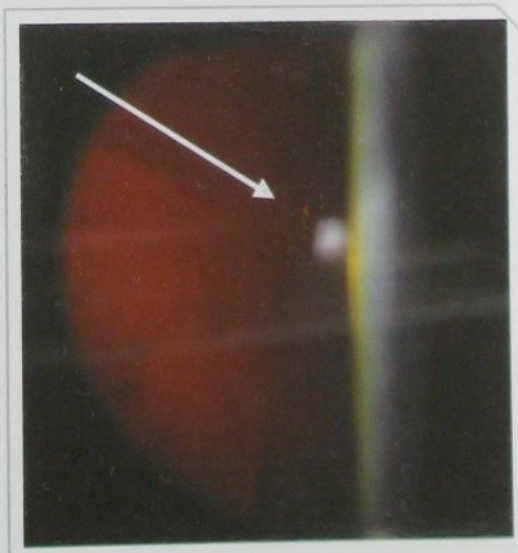


Fig. 11.8. Vacuoles in the posterior cortex (shown with an arrow)

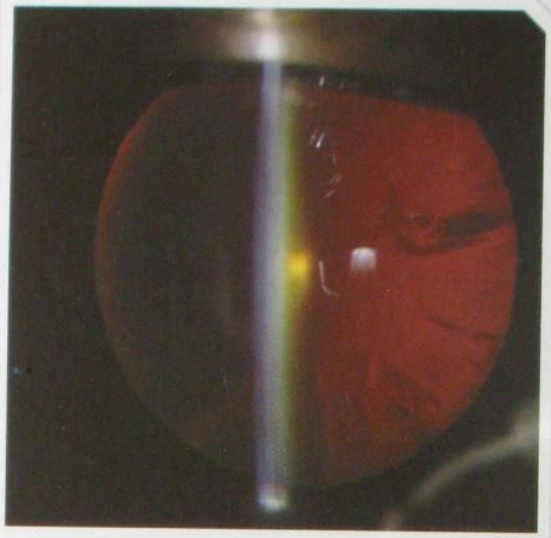
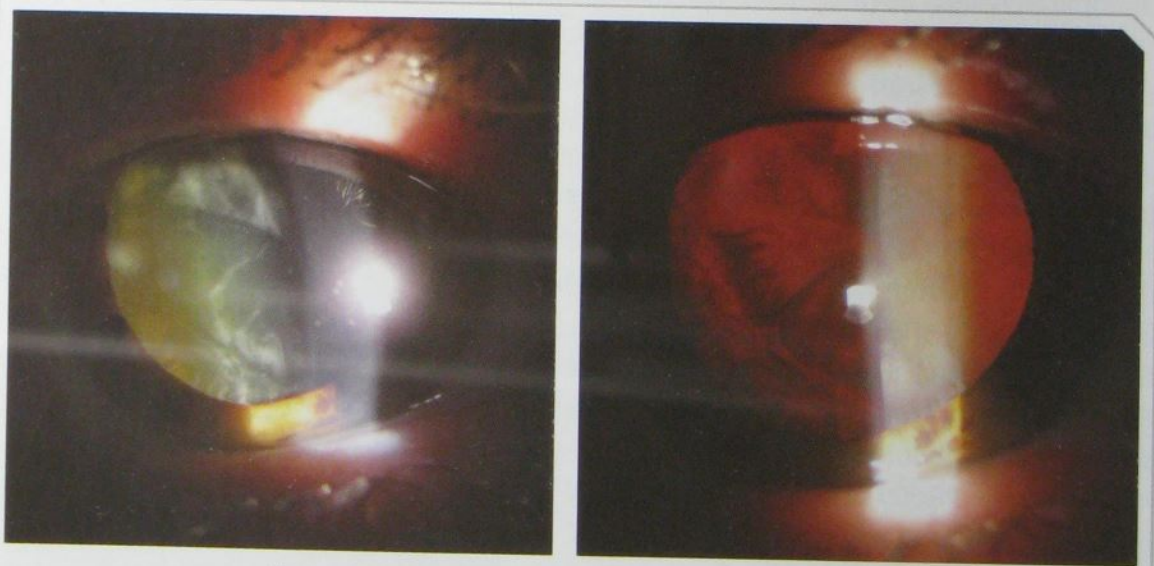


Fig. 11.9. Cuneiform cortical opacities (cortical spokes)

Wedge-shaped opacities (often called cortical spokes or cuneiform opacities) form near the periphery of the lens, with the pointed end of the opacities oriented toward the center. They are first seen in the lower nasal quadrant. These opacities are present both in the anterior and posterior cortex. On oblique illumination these present a typical radial spoke-like pattern of grayish white opacities. On distant direct ophthalmoscopy, these opacities appear as dark lines against the red fundal reflex (fig. 11.9). The nucleus at this stage shows signs of sclerosis and initial opacities. VA at this stage is normal or mildly decreased (1.0—0.8).

2. Immature cataract. At this stage, opacification progresses further. The lens appears grayish-white but clear cortex is still present and iris shadow is visible. Scattered opacities of the lens are separated by clear zones (fig. 11.10, 11.11). In some patients, at this stage, the lens may become swollen due to continued hydration. This condition is called *intumescent cataract*. In this case due to swollen lens the anterior chamber becomes shallow. Visual acuity at this stage keeps decreasing to 0.7—0.01.



A B

Fig. 11.10. Immature cortical cataract in focal (A) and direct (B) illumination. Also yellowish opacification of the nucleus (A) can be seen

3. Mature cataract. At this stage opacification becomes complete, the whole cortex is totally opaque, the lens structure cannot be differentiated. The lens becomes pearly white or brownish in color, no iris shadow is formed. Vision at this stage decreases from 0.01 to light perception (fig. 11.12).

Fig. 11.11. Immature nuclear cataract

4. Hypermature cataract. When mature cataract is left untreated, the next stage is hypermaturity. Hypermature cataract may appear in any of the two forms:

(a) *Sclerotic cataract* occurs when degenerated cortical material leaks through the lens capsule leaving the capsule wrinkled and shrunken (fig. 11.13).

(b) *Morgagnian cataract* occurs when further liquefaction of the cortex allows free movement of the nucleus within the capsular bag. The small brown nucleus usually depresses downwards so that its upper equatorial part is visible.

Methods of examination. Examination methods for cataract should include: visual acuity testing, evaluation of intraocular pressure, distant direct ophthalmoscopic examination. Slit-lamp examination with maximally dilated pupil is the main method in cataract patients; it allows evaluating complete morphology of opacity (site, size, shape, color pattern, and hardness of the nucleus). Where the fundus is not visible in mature lens opacity, ultrasound studies (one-dimensional A-scan and two-dimensional B-scan) are indicated to exclude involvement of the deeper structures of the eye.

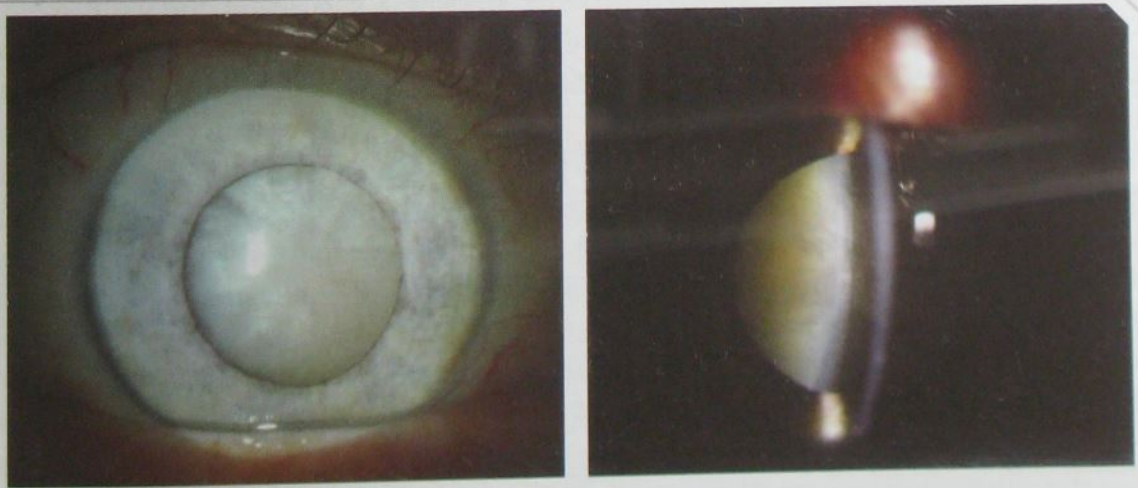


Fig. 11.12. Mature cataract

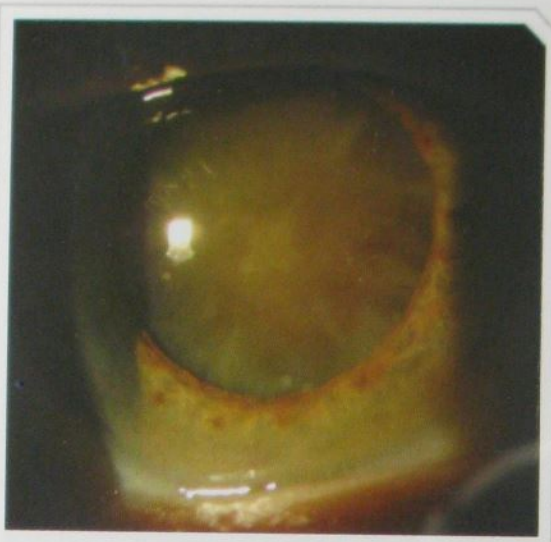


Fig. 11.13. Hypermature sclerotic cataract

Treatment. The only curative treatment of cataracts is their surgical removal. However, in some conditions while surgery cannot be carried out for any reason (psychological — the patient is not ready mentally; medical — the risk of severe complications due to systemic diseases; social or any other), optical correction or therapeutic treatment may be prescribed. Lots of topical preparations delaying cataract progression and consisting mostly of vitamins, antioxidants, and minerals are available at the market nowadays. But none of them have shown satisfying and confirmed effect in delaying the progression of cataract.

The main surgical techniques of cataract removal are as follows:

— intracapsular cataract extraction,
— extracapsular cataract extraction,
— phacoemulsification.

Intracapsular cataract extraction (ICCE) is an obsolete technique at present time and is mentioned here just in historical aspect. In this method, a big (150—180°) corneoscleral incision is made, through which the entire lens is removed within its capsule with the aid of a cryoprobe or a special lens spatula by tearing up all Zinn ligaments. Additionally iridectomy is performed and the incision is closed with sutures. Numerous complications of this method (e.g., retinal detachment, secondary glaucoma, intraocular bleeding, etc.) made ophthalmologists decline it and search for something safer and more effective.

Extracapsular cataract extraction (ECCE) was a big step forward in cataract surgery. In this technique, after corneoscleral incision, the major part of the anterior capsule is cut in "can opener" technique with a bent needle or a special instrument — cystotome — and removed together with the cortex and nucleus of the lens. Therefore, the lens' capsular bag remains intact being held with Zinn ligaments, which creates perfect background for further IOL implantation into the capsular bag or posterior chamber. Advantages of the method are smaller incision, less sutures, less postoperative astigmatism, safe natural barrier between the posterior and anterior segments of the eye, and as a result — absence of vitreous herniation into the anterior chamber, secondary glaucoma, etc. Relative simplicity of this technique and no need for expensive equipment led to appearance of various modifications of the method, and also made it quite popular among ophthalmologists so that even now in some small clinics it can still be seen, however, more and more rarely.

Phacoemulsification of cataract is the most widespread method of cataract surgery in the world. Phacoemulsification uses a special phaco machine with an ultrasonically driven tip moving along its longitudinal axis at a speed of 40,000 times a second and thus fragmenting and emulsifying the nucleus. The technique also uses a surgeon-controlled automated irrigation-aspiration system to remove the fragmented nucleus and cortical material through a small needle introduced through a very small incision (1.2—2.75 mm). Incisions are usually clear corneal (they allow to avoid bleeding) and self-sealing (no need in sutures); besides that, such small incisions completely prevent post-operative astigmatism. Protection of the corneal endothelium and other intraocular structures from instrument manipulations in the anterior chamber is ensured by the use of protective viscoelastic materials previously injected into the anterior chamber. Partial removal of the anterior capsule of the lens (4—6 mm in diameter) is usually carried out by means of continuous curvilinear capsulorhexis (CCC) technique.

Development of phacoemulsification: laser phacoemulsification. The latest modification of conventional phacoemulsification technique is femtosecond laser-assisted phacoemulsification. The principle of femtosecond laser work is ocular tissue disruption due to concentration of a great energy in a very small point during an extremely short period of time — 10^{-9}—10^{-12} sec. The main advantage of the method is that all the cutting procedures like corneal incisions, capsulorhexis, as well as nucleus emulsification can be done very safely and precisely. However, the high cost of the equipment is a serious disadvantage restricting worldwide spread of the technique.

Correction of postoperative aphakia. For full rehabilitation of cataract patients it's not enough to restore optical media transparency; proper correction of refractive abnormality, which appears after opaque lens removal, is also very important. Presently, correction of postoperative aphakia with intraocular lenses (IOL) is considered to be the most physiological. The state of the eye after cataract surgery with implanted IOL is called *pseudophakia*.

IOL consists of two main parts: optical and haptical fixating and centering the lens in the right position.

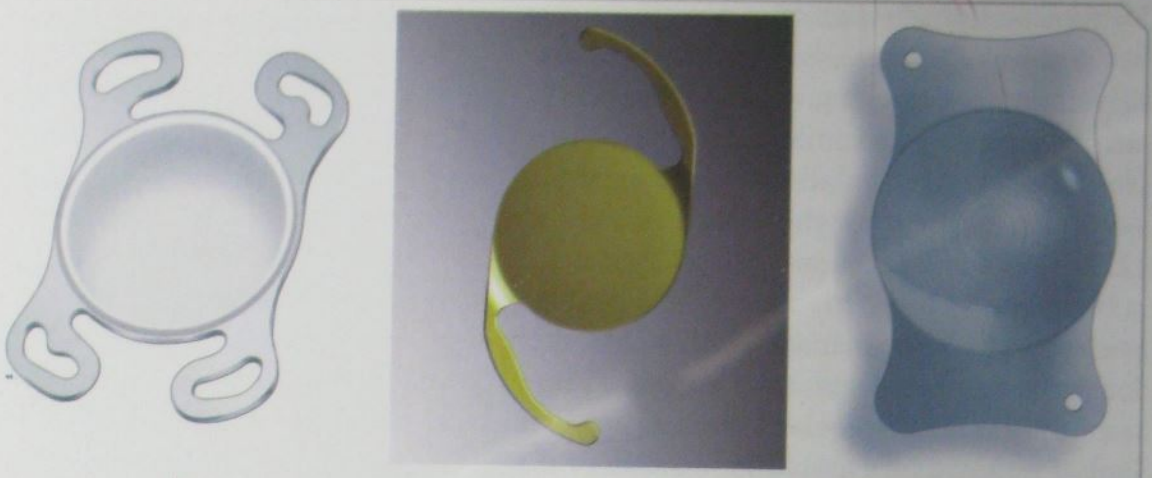
By anatomic position IOLs can be divided into: anterior chamber IOL, posterior chamber IOL, IOL with iris fixation, and capsular fixation IOLs. The latter are obviously the most preferable in case of non-complicated phacoemulsification or extracapsular extraction.

IOL design can be various and keeps improving and progressing (fig. 11.14).

1. Rigid IOLs require an incision larger than the diameter of its optical part. They are made entirely from polymethylmethacrylate (PMMA) and are now generally used only in developing countries.

2. Flexible IOLs are now in general use and allow implantation through a very small incision. For insertion they may be folded in half with special forceps or loaded into an injector delivery system, then unfolded or unrolled inside the eye. By the material they are made of IOLs are classified as follows:

— *silicone* IOLs are available in both loop haptic (1- or 3-piece) and plate haptic (1-piece) conformations,



A B C

Fig. 11.14. Examples of IOL design: A — four-point aspheric IOL *Akreos™ AO*, Bausch+Lomb; B — Toric C-loop IOL *AcrySof® TORIC SN60T5*, Alcon; C — Trifocal IOL *AT LISA® tri 839MP*, Zeiss

— *acrylic* IOLs may be hydrophobic (water content < 1 %) or hydrophilic, with much higher water content. Hydrophobic acrylic materials have a greater refractive index than hydrophilic lenses and are consequently thinner. Hydrophilic acrylic (hydrogel), in theory, offers better biocompatibility,

— *collamer* is composed of collagen, a poly-HEMA based copolymer, and a UV-absorbing chromophore.

3. Aspheric IOLs neutralize spherical aberration and improve contrast, particularly in mesopic conditions.

4. Multifocal IOLs aim to provide clear vision at different focal distances. Recently, the so-called *pseudoaccommodative* IOLs achieving their purpose by refractive or diffractive means have become the most widespread.

5. Toric IOLs have a cylindrical refractive component to compensate for pre-existing corneal astigmatism.

6. Blue light filters. Although essentially all IOLs contain ultraviolet light filters, some of them also include filters for blue wavelengths in order to reduce the probable damage to the retina induced by light of this spectrum.

IOL Optic Power Calculation

Regression formulas (formulas generated by averaging large numbers of postoperative clinical results) are used to choose the appropriate IOL power for achieving emmetropia in the eye. The following formula developed by Sanders, Retzlaff, and Kraff in the 1980s (SRK formula) was the basic one for future development of IOL calculations:

$P = A - (2.5L) - 0.9K$,

where P = lens implant power for emmetropia (diopters); L = axial length of the eye (mm); K = average keratometric reading (diopters); A = a constant, specific to the lens implant to be used, which depends on the lens material, design, positioning, etc.

Newer versions of regression formulas (e.g., SRK/T, Holladay 2, Hoffer Q, Haigis) have been developed for IOL power calculation in the eyes outside the range of 22—25 mm length (patients with initial severe refractive abnormalities).

Secondary Cataract

Secondary cataract is an opacity which persists or develops after cataract surgery carried out with lens capsule retention (ECCE or phacoemulsification).

Etiology. Residual lens epithelial cells are inevitably left behind at the time of surgery, attached to the capsule despite thorough removal of soft lens material. They proliferate, migrate, and undergo metaplasia, resulting in opacification of the posterior capsule. Two patterns of posterior capsular opacification are seen: cellular proliferation, producing drop-like deposits in the visual axis (Elschnig's pearls), or fibrosis and shrinkage of the capsule, producing striae.

Symptoms and signs. Patients report gradual decrease in vision during some months of years after cataract surgery. They describe hazy or misty vision as well as glare. Elschnig's pearls are large deposits on the posterior capsule in the form of droplet-like soap bubbles (fig. 11.15). There also may be striae and a fibrous sheet across the visual axis.

Treatment. There is now considerable evidence that square-profile IOLs reduce the risk of secondary cataracts. YAG-laser capsulotomy is a very effective and safe procedure for the treatment of visually significant opacities. It is performed as an outpatient procedure and does not require any anesthesia.

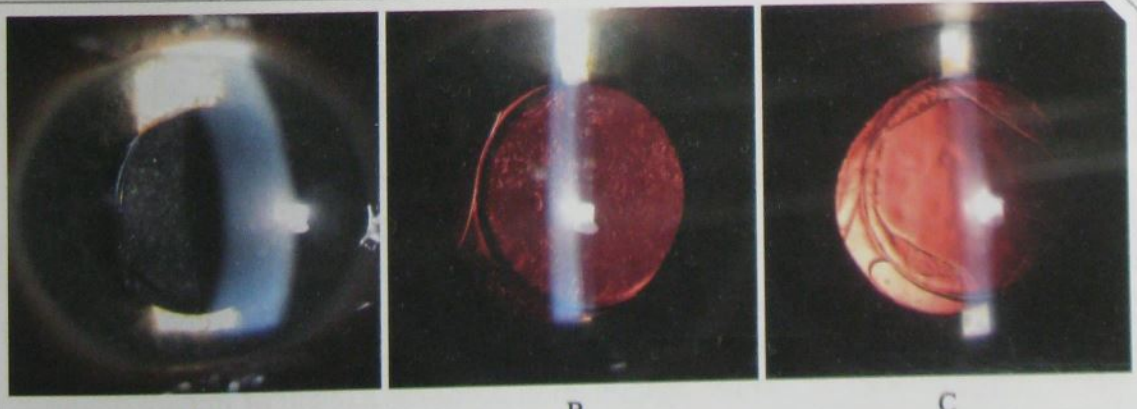


Fig. 11.15. Secondary cataract, the same eye: A — in oblique illumination; direct illumination B — before and C — after YAG-laser capsulotomy

Review:

1. Key Points

Diseases of the lens: abnormalities of lens position, size, and shape; cataracts.

Abnormalities of lens position: subluxation (displacement) or luxation (dislocation) may be hereditary (familial ectopia lentis, Marfan's syndrome, Weill—Marchesani syndrome, homocystinuria) and acquired (due to trauma, pseudoexfoliation, ciliary body tumor, buphthalmos). Treatment of lens position anomalies includes: optic correction of refractive errors or surgical lensectomy.

Anomalies of lens size and shape: congenital aphakia, lenticonus, lentiglobus, lens coloboma, microspherophakia, remnants of the fetal vasculature system of the lens. Treatment of these anomalies depends on the degree of vision impairment and the possibility of optic correction of refractive errors and may be optical or surgical.

Cataract is the presence of any persistent opacity in the lens.

By the time of onset cataracts are classified into: congenital, juvenile, and acquired. By morphology congenital cataracts are divided into: total and partial. Partial cataracts may be anterior and posterior; polar, zonular, and membranous. Zonular cataracts are subdivided into: lamellar, sutural (stellate), nuclear, coronary, and blue dot (cerulean). Age-related cataracts by morphology are classified into: cortical, nuclear, posterior subcapsular, and mixed.

Etiological classification of acquired cataracts includes: age-related (senile), traumatic, metabolic, complicated, toxic, electrical, postoperative, cataracts associated with systemic diseases, and cataracts associated with miscellaneous syndromes.

Senile cataracts are usually graded by the stage of maturation: incipient, immature, mature, and hypermature.

Etiology of congenital cataract includes the following factors: heredity, maternal factors (malnutrition, infections, drugs ingestion, radiation), fetal or infantile factors (anoxia, metabolic disorders, other congenital anomalies (Lowe's syndrome, dystrophic myotony, etc.)), birth trauma, malnutrition in early infancy. Also, about half of congenital cataracts are idiopathic.

Symptoms and signs and the degree of vision loss in congenital cataracts vary significantly and mostly depend on their morphologic type. Treatment strategies consider the degree of vision impairment and may include observation or optic correction as well as cataract surgery at early age in cases when opacities impair vision, which may result in amblyopia development in the future.

In acquired cataracts lens fibre opacification occurs as a result of impact of various causative factors like aging, trauma, metabolic disorders, different physical (radiation, electricity) and toxic agents. Symptoms and signs in these cataracts mostly are: gradual painless decrease of vision, changes in refraction (myopic shift), colored ha-

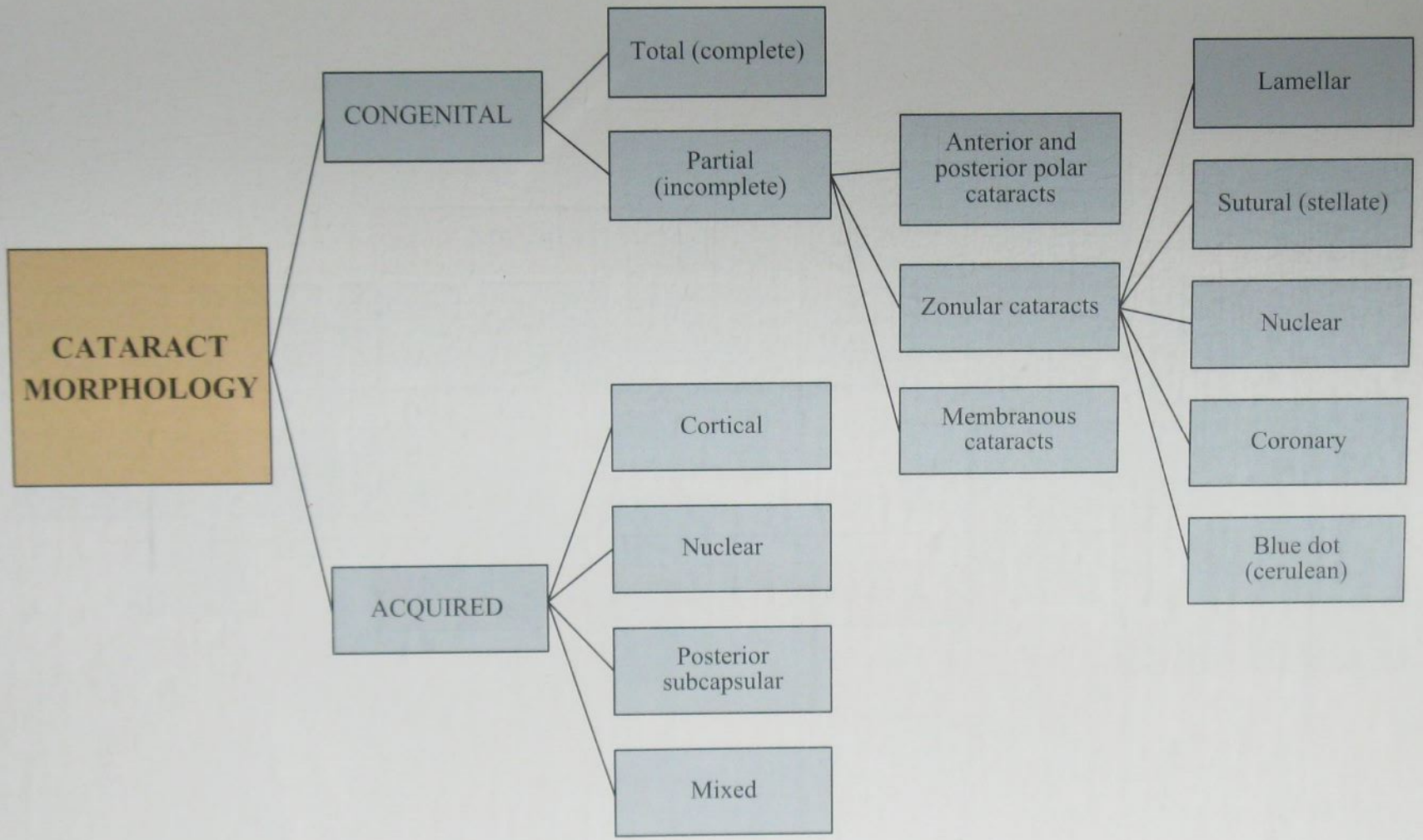
los around the light sources, and monocular diplopia. Acquired cataracts usually are bilateral, but asymmetric. Objective signs vary depending on the stage of maturation and morphologic type: vacuoles in the anterior or posterior cortex, cuneiform cortical opacities, scattered opacities of the lens separated by clear zones, total opacification of the cortex with inability to distinguish zones of the lens at the mature stage, liquification of the cortex with nucleus depression or total sclerosis of the lens in hypermature cataract.

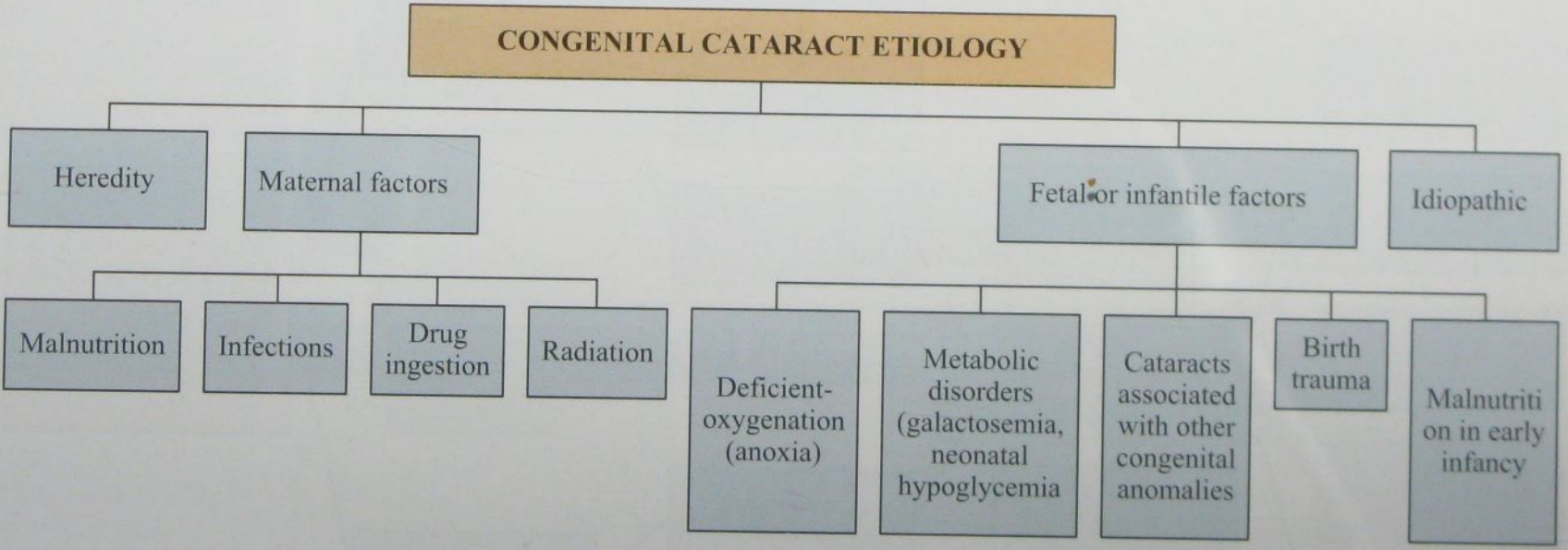
The main treatment of cataracts is surgical lens removal with intraocular artificial lens implantation for the optic correction of ammetropia. Surgical techniques include: intracapsular cataract extraction; extracapsular cataract extraction; phacoemulsification. IOLs may be rigid and flexible, made of different materials (polymethylmethacrylate, silicon, acryl), may have different design of their haptic part and different optic properties (monofocal, multifocal, toric, aspheric, etc.). The optic power of IOLs should be calculated preoperatively for each eye with the aid of special formulas.

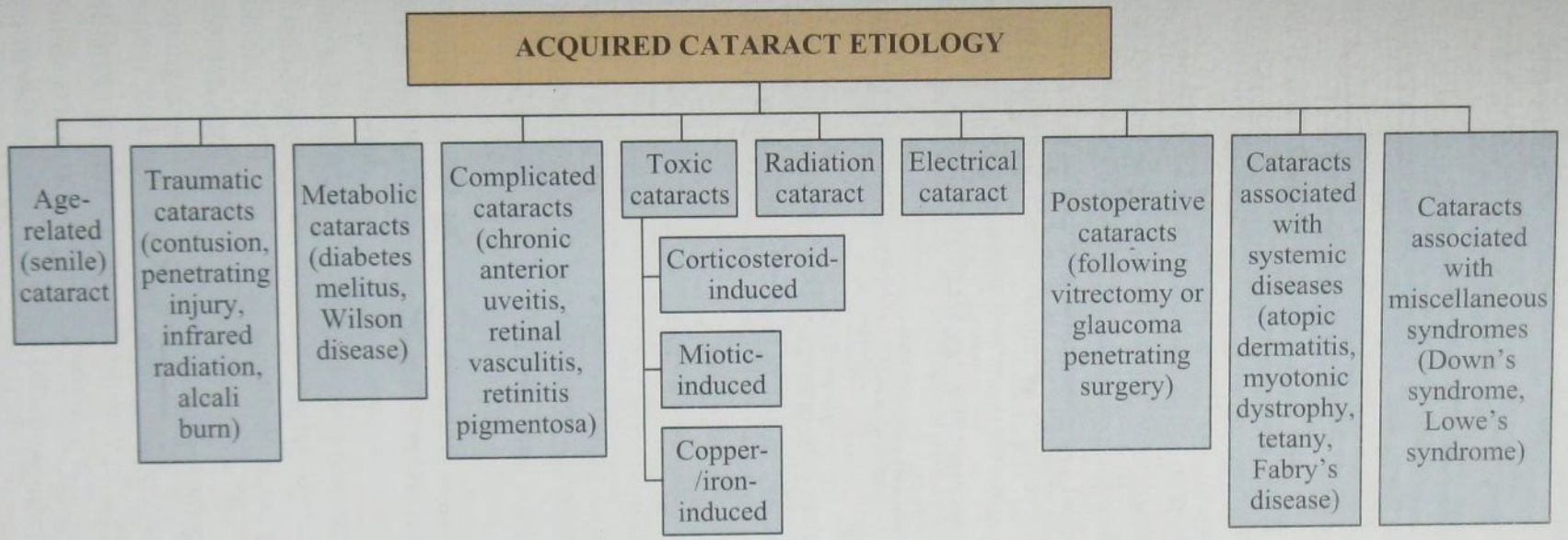
Secondary cataract is opacity in the posterior lens capsule after cataract surgery, which decreases its optical results. Secondary cataract treatment includes laser or surgical capsulotomy.

2. Diagrams

DISEASES OF THE LENS
Anomalies of lens position
Displacement
Dislocation
Anomalies of lens size and shape
Congenital aphakia
Lenticonus
Lentiglobus
Lens coloboma
Micro-spherophakia
Remnants of the fetal vasculature system of the lens
Cataracts


CATARACT MORPHOLOGY
CONGENITAL
Total (complete)
Partial (incomplete)
Anterior and posterior polar cataracts
Zonular cataracts
Membranous cataracts
Lamellar
Sutural (stellate)
Nuclear
Coronary
Blue dot (cerulean)
ACQUIRED
Cortical
Nuclear
Posterior subcapsular
Mixed


CONGENITAL CATARACT ETIOLOGY
Heredity
Maternal factors
Fetal or infantile factors
Idiopathic
Malnutrition
Infections
Drug ingestion
Radiation
Deficient-oxygenation (anoxia)
Metabolic disorders (galactosemia, neonatal hypoglycemia
Cataracts associated with other congenital anomalies
Birth trauma
Malnutrition in early infancy


ACQUIRED CATARACT ETIOLOGY
Age-related (senile) cataract
Traumatic cataracts (contusion, penetrating injury, infrared radiation, alcali burn)
Metabolic cataracts (diabetes melitus, Wilson disease)
Complicated cataracts (chronic anterior uveitis, retinal vasculitis, retinitis pigmentosa)
Toxic cataracts
Corticosteroid-induced
Miotic-induced
Copper-/iron-induced
Radiation cataract
Electrical cataract
Postoperative cataracts (following vitrectomy or glaucoma penetrating surgery)
Cataracts associated with systemic diseases (atopic dermatitis, myotonic dystrophy, tetany, Fabry's disease)
Cataracts associated with miscellaneous syndromes (Down's syndrome, Lowe's syndrome)

3. The Review Questions

A. Control Questions

1. What diseases of the lens do you know?
2. Name anomalies of lens position. In which chromosomal diseases may they occur?
3. Name anomalies of the size and shape of the lens.
4. Give definition to the term *cataract*; classification of cataracts.
5. What is the etiology of congenital cataracts? Describe their main symptoms and signs.
6. Name the differences in clinical picture of different morphological types of congenital cataracts. What are the approaches to congenital cataract treatment?
7. Name the main etiological factors of acquired cataracts.
8. Describe clinical symptoms of senile cataracts depending on their morphology.
9. Specify objective signs of senile cataract depending on its maturation stage.
10. What examination methods will you use in cataract patient examination?
11. What is the treatment strategy for acquired cataract? Name the main techniques of cataract surgery.
12. How to correct postoperative aphakia after cataract surgery? Specify the main types of IOLs.
13. How to calculate IOL optic power in cataract patient?
14. What is secondary cataract? Name the main approaches to its treatment.

B. Tests

1. **Which of the listed objective signs mainly refer to *lens subluxation*?**
   A. Dislocation of the lens into the vitreous
   B. Total opacification of the lens
   C. Opacities of the posterior capsule of the lens
   D. Presence of irido- and phacodonesis
   E. Absence of the lens in the pupillary area

2. **All of these conditions are *anomalies of lens size and shape* except:**
   A. Anterior lenticonus
   B. Microspherophakia
   C. Sutural cataract
   D. Lens coloboma
   E. Remnants of the fetal vasculature system of the lens

3. **Which of the following is not characteristic for *congenital cataracts*?**
   A. Lamellar cataract
   B. Sutural cataract
   C. Polar cataract
   D. Blue dot cataract
   E. Morgagnian cataract

4. **Which of the following congenital cataracts should be treated surgically at early age?**
   A. Bilateral blue dot cataract

B. Bilateral total cataract
C. Unilateral anterior polar cataract
D. Bilateral anterior polar cataract
E. Coronary cataract

5. What is the most frequent etiological factor for acquired cataracts?
A. Trauma
B. Infections of the anterior segment of the eye
C. Toxic effects of medications
D. Metabolic disorders of the body
E. Age

6. Which of following signs describe the *incipient stage* of senile cataract?
A. Dense yellow nucleus of the lens, visual acuity = 0.1
B. Scattered opacities of the lens separated by clear zones, VA = 0.6
C. The cortex is milky-white, a small brown nucleus is dislocated downwards
D. Vacuoles in the posterior cortex, cuneiform opacities in the lower nasal quadrant
E. The cortex is opaque, the lens layer cannot be differentiated

7. In *immature senile cataract* during slit-lamp examination we can see:
A. Grayish-white lens, scattered opacities of the lens, but clear cortex is still present and iris shadow is visible
B. Cuneiform opacities in the lower nasal quadrant
C. The cortex is opaque, the lens layer cannot be differentiated, no iris shadow
D. The lens is brown, its capsule is wrinkled and shrunken
E. Small discrete bluish opacities in the cortex

8. A patient has immature senile nuclear cataract with visual acuity decreased to 0.5. What is the most preferable treatment strategy in this case?
A. Therapeutic treatment with vitamins and antioxidant eye-drops
B. Intracapsular cataract extraction
C. Phacoemulsification
D. Optical correction of refractive abnormalities
E. No treatment required because the patient has normal visual acuity

9. Which of the following methods are considered most physiological for postoperative aphakia correction?
A. Contact lenses
B. Intraocular lenses
C. Glasses
D. Excimer-laser correction
E. No need for any optical correction

10. The term *secondary cataract* means:
A. Cataract developed during the intrauterine period due to maternal infection
B. Cataract developed after a penetrating injury of the eyeball
C. Cataract developed in a patient with diabetes mellitus
D. Opacity of the posterior lens capsule following vitreoretinal surgery for retinal detachment
E. Opacity of the posterior lens capsule following cataract phacoemulsification with IOL implantation

C. Clinical Cases

Case 1

A patient, 20 years old, complains of pure vision in both eyes since early age. General examination reveals tall and thin stature, arachnodactyly, hypermobility of joints. Ophthalmic examination revealed in both eyes 7-diopter myopia, the lenses of both eyes are displaced superiorly and temporally. Which diagnosis should be supposed in this case?

A. Weill—Marchesani syndrome
B. Homocystinuria
C. Marfan's syndrome
D. Bilateral congenital cataract
E. Congenital myopia

Case 2

An 18-year-old patient complains of blurred vision since early childhood, when high myopia was diagnosed. He wears concave (–10.0 diopters) spectacles for both eyes. Objectively the patient has short stature, thick and short fingers, short hands. Ophthalmological signs: bilaterally the lenses have small diameter, their equator is seen in the dilated pupil, they have a spherical shape. Beside this, the lenses are displaced inferiorly and nasally. Which diagnosis is more probable?

A. Weill—Marchesani syndrome, microspherophakia
B. Homocystinuria
C. Marfan's syndrome
D. Bilateral congenital cataract
E. Congenital myopia

Case 3

A woman noted that the pupils of both eyes of her 1-week baby have unusual white color. What pathology may be suspected in this case?

A. Congenital aphakia
B. Congenital cataract
C. Lenticonus
D. Microspherophakia
E. Conjunctivitis

Case 4

A prophylactic slit lamp examination of a 25-year-old woman with visual acuity of both eyes = 1.0 (20/20) and without any complains of vision showed small, discrete bluish opacities of the lens cortex. What is the diagnosis?

A. Congenital nuclear cataract
B. Congenital coronary cataract
C. Senile cortical cataract

D. Blue dot cataract
E. Congenital sutural cataract

Case 5

A woman referred to an ophthalmologist because she noted in her 2-month daughter no reaction to bright toys and relatives' faces, nystagmus, and whitish color of the pupils in both eyes. Ophthalmologic examination revealed dense opacities of the entire nucleus in both baby's lenses. What type of congenital cataract is most probable in this case?

A. Nuclear cataract
B. Total cataract
C. Coronary cataract
D. Posterior polar cataract
E. Sutural cataract

Case 6

A 65-year-old patient complains of gradual decrease of far vision in both eyes during the last 2—3 years, worse vision in conditions of bright illumination or sunshine. At the same time he said he cannot use his reading glasses anymore and can see at near distance without spectacles. Refractometry revealed 4-diopter myopia. Slit-lamp examination showed a dense brownish nucleus of the lens. Which is the most probable diagnosis?

A. Senile cortical cataract
B. Senile nuclear cataract
C. Secondary cataract
D. Congenital total cataract
E. Congenital aphakia

Case 7

A 55-year-old patient complains of decreased vision in the right eye. Visual acuity of the right eye = 0.8. Slit-lamp examination revealed vacuoles in the anterior or posterior cortex, cuneiform opacities in the lens' cortex. Examination in direct illumination showed radial spoke-like opacities in the lens. What is the diagnosis?

A. Immature senile cataract
B. Mature senile cataract
C. Incipient senile cataract
D. Hypermature senile cataract
E. Morgagnian cataract

Case 8

A patient, 67 years old, complains of gradual decrease of vision during the last 3 years. Visual acuity in both eyes = 0.2, intraocular pressure is normal. Objective examination shows no pathological changes in the conjunctiva, cornea, and anterior

chamber. The lens appears grayish-white but clear cortex is still present and iris shadow is visible. Scattered opacities of the lens are separated by clear zones. What is the diagnosis?

A. Immature senile cataract
B. Mature senile cataract
C. Incipient senile cataract
D. Hypermature senile cataract
E. Morgagnian cataract

Case 9

An 82-year-old patient came to an ophthalmologist with complaints of gradual painless vision loss in the right eye. Vision has been getting worse progressively during 10 or 12 years. 2 years ago vision disappeared completely, but the patient didn't visit a doctor. VA in the right eye = 1/∞ pr. l. certa (correct light projection). During slit-lamp examination the lens is white, the cortex seems milky-white and liquefied. A small brown nucleus can be seen depressed downwards within the capsular bag. What is the diagnosis?

A. Immature senile cataract
B. Mature senile cataract
C. Incipient senile cataract
D. Secondary cataract
E. Morgagnian cataract

Case 10

A 64-year-old female patient complains of gradual decrease of vision in her left eye which she noted 3 month ago. 2 years ago she was treated surgically for immature cataract (cataract phacoemulsification with IOL implantation) in this eye. VA in the left eye = 0.6. Objective examination reveals no changes in the anterior segment of the left eye. Pseudophakia, IOL is centered inside the capsular bag. The posterior capsule of the lens is a bit opaque, large droplet-like deposits reminding soap bubbles are seen on the posterior capsule. How is such condition called?

A. Morgagnian cataract
B. Incipient senile cataract
C. Secondary cataract
D. Mature cataract
E. Immature cataract

CHAPTER 12

Glaucoma

OBJECTIVES

Upon completion of the chapter the students should:

- know the ways of aqueous humor production and its drainage pathways;
- know the basic methods and principles of glaucoma diagnosis and its results interpretation;
- know the main risk factors of glaucoma development and its prevention;
- know the classification of glaucoma;
- know the clinical picture of different types of glaucoma;
- know the main principles of glaucoma treatment;
- be able to render first aid in acute angle-closure glaucoma.

Plan:

1. CLASSIFICATION OF GLAUCOMA
2. AQUEOUS HUMOR PRODUCTION AND DRAINAGE
3. RISK FACTORS FOR GLAUCOMA DEVELOPMENT
4. SIGNS AND SYMPTOMS OF GLAUCOMA
5. EXAMINATION METHODS
6. GLAUCOMA
   - **6.1. Open-Angle Glaucoma**
     - Primary Open-Angle Glaucoma
     - Normal Tension Glaucoma
     - Ocular Hypertension
   - **6.2. Angle-Closure Glaucoma**
     - Acute Angle-Closure Glaucoma
   - **6.3. Congenital Glaucoma**
   - **6.4. Secondary Glaucoma**

Content:

Worldwide, glaucoma is the second most frequent cause of blindness in developing countries after diabetes mellitus. Fifteen to twenty per cent of all blind persons lost their eyesight as a result of glaucoma. According to the data of the World Glaucoma Society there are 71 million people with glaucoma in the world and 8 million of them are blind on both eyes. Glaucoma affects one in two hundred people aged fifty and younger and one in ten over the age of eighty. It is why glaucoma is an increasingly important public health concern due to the aging population demographics. It has been nicknamed "the silent sight thief" as it usually causes no symptoms early in its course. Untreated glaucoma leads to permanent damage of the optic nerve and as result to visual field loss, which can progress to blindness. Early detection of glaucoma and its appropriate treatment are the highest priorities for the public health system and are keys to preserving sight in people with glaucoma. The knowledge of clinical presentation, diagnostics, and treatment principles of glaucoma is necessary for any practicing doctor.

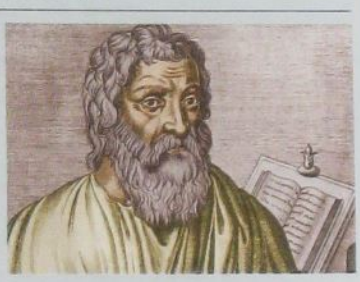
1. Classification of Glaucoma

Glaucoma is a multifactorial progressive optic neuropathy with a characteristic acquired loss of the retinal ganglion cells and atrophy of the optic nerve that leads to irreversible loss of visual functions and blindness.

Glaucoma is a diverse group of eye conditions (at least 70 types), which can be divided into diagnostic groups in the following way:

- — according to the presence or absence of causative factors — *primary* or *secondary*;
- — according to the state of the anterior chamber angle — *open-angle*, *angle-closure* or *mixed*;
- — according to the onset — *acute* or *chronic*;
- — according to the age of onset — *congenital*, *infantile*, *juvenile* or *adult*;
- — according to the IOP level — *normal tension glaucoma, high tension glaucoma, ocular hypertension;*
- — according to the stage of the disease — *mild*, *moderate*, *advanced*, *far advanced*, *end stage*;
- — according to the dynamics of vision — *stabilized* or *unstabilized.*

EYE FACTS



The disorder, now defined as glaucoma, was first documented by the ancient Greeks in 400 BC. Glaucosis (means "blue-green water" or "color of the sea") was first mentioned in Hippocratic writings as a blinding disease occurring most commonly in the elderly. There was described a typical picture of a blind eye caused by glaucoma — a dilated non-reactive pupil with a greenish-blue hue. The description stated "that once the pupil has the color of the sea, eyesight is destroyed".

2. Aqueous Humor Production and Drainage

The anterior chamber of the eye is filled with a clear fluid called the aqueous humor, which is constantly produced by the ciliary body and provides nourishment to the structures in the front of the eye.

EYE FACTS

- the rate of aqueous humor production is about 1.5—3 μl/min;
- the volume of aqueous humor is about 0.25 ml (anterior chamber) + 0.06 ml (posterior chamber);
- the amount of aqueous humor replaced per minute is 1—2 %;
- aqueous humor turnover in the eye is 1.5—2 hours.

After being produced the aqueous humor enters the posterior chamber, flows through the pupil into the anterior chamber and then to the anterior chamber angle formed by the inner cornea and the root of the iris (irido-corneal angle), which contains the trabecular meshwork (fig. 12.1).

The aqueous humor drains out of the eye via one of the two outflow pathways, through the trabecular or uveoscleral outflows (fig. 12.2).

Trabecular outflow is aqueous humor drainage via the trabecular meshwork, that is loose sponge-like avascular tissue, into Schlemm's canal, collector channels, then it passes into the episcleral veins, anterior ciliary veins and enters the systemic blood circulation. This pathway is responsible for approximately 85—90 % of aqueous outflow.

Uveoscleral outflow is aqueous humor drainage from the anterior chamber angle back into the tissue spaces of the ciliary muscle, then into the supraciliary and supra-

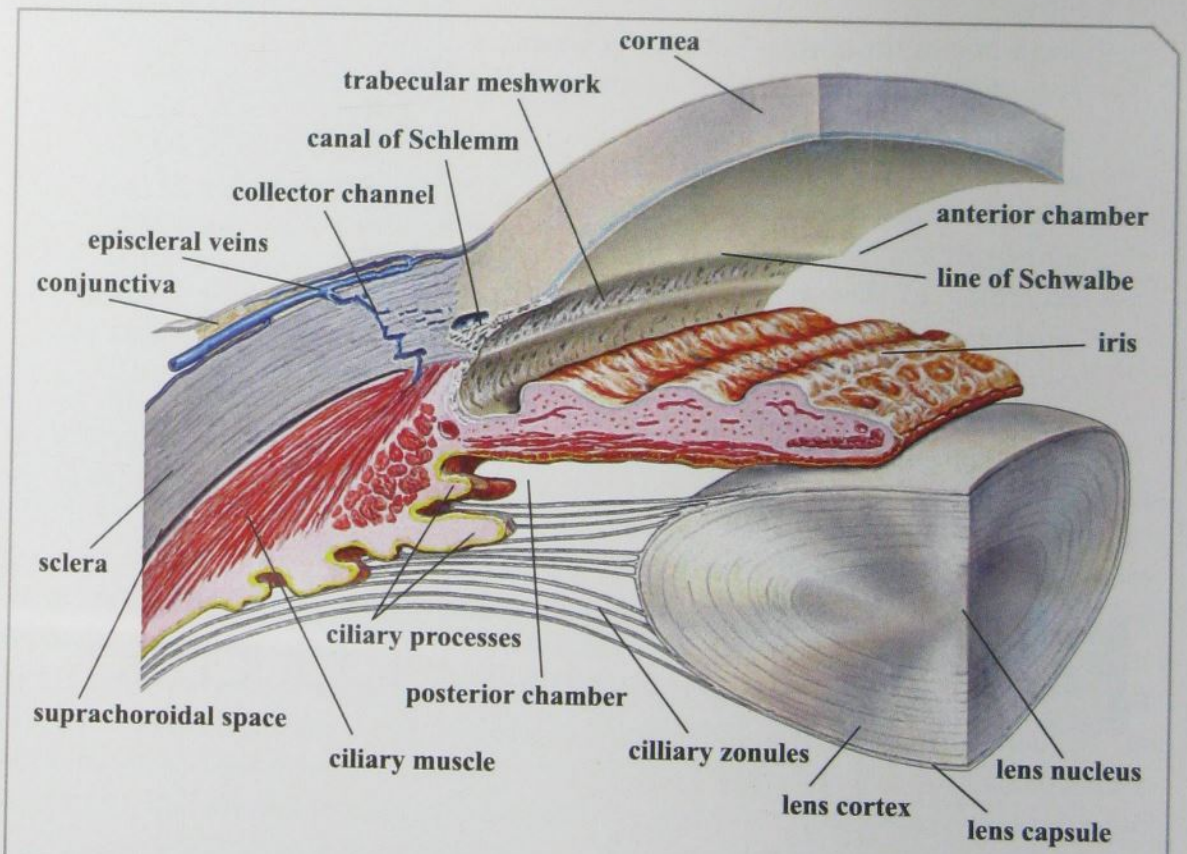


Fig. 12.1. Structures of the anterior chamber angle (from Анатомія людини: підручник: у 3 т. Т. 2 / А. С. Головацький, В. Г. Черкасов, М. Р. Сапін та ін. — Вид. 4. — Вінниця: Нова Книга, 2009. — С. 371—397)

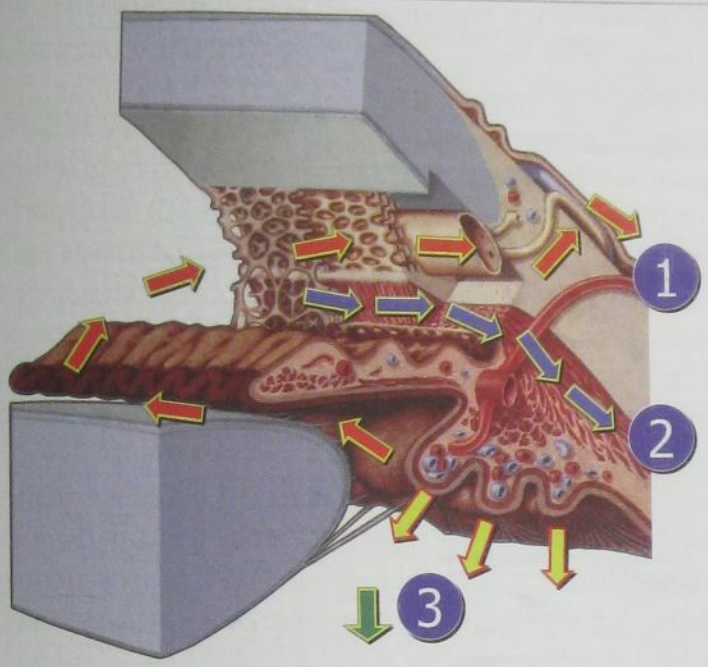


Fig. 12.2. Pathways of the aqueous humor outflow: trabecular (1), uveoscleral (2), across the vitreous (3)

choroidal spaces. The fluid then exits the eye through the intact sclera or along the nerves and the vessels that penetrate it. Uveoscleral outflow has been estimated to account for 10—15 % of total aqueous outflow.

Although the bulk of the aqueous humor exits the eye through the anterior chamber outflow pathways, some of it drains across the vitreous into the retina and retinal pigment epithelium.

The relationship between aqueous humor production and its outflow determines the intraocular pressure (IOP). IOP increases when either too much fluid is produced in the eye or the drainage or outflow channels (trabecular or uveoscleral) of the eye become blocked.

3. Risk Factors for Glaucoma Development

— *Elevated intraocular pressure* — persistent elevated IOP is the most significant risk factor for the development of glaucoma. The higher the level of IOP, the greater the risk of developing the disease;

— *Age over 40 years* — the prevalence of glaucoma increases with age. Persons of age 80 have up to 10 % prevalence rates, while persons of age 35 have 1—2 % prevalence rates of glaucoma;
— *Family history of glaucoma*, particularly in first-degree relatives, is an important risk factor for the development of glaucoma. People with a family history of glaucoma have an about six percent chance of its development;
— *Race* — compared to Caucasians, open angle glaucoma is up to 5 times more prevalent, develops at an earlier age, and is more severe in African-Americans. Blindness caused by glaucoma is up to 8 times more common in African-Americans. Asians are susceptible to angle-closure glaucoma, and Inuits have a twenty to forty times higher risk than Caucasians of developing primary angle closure glaucoma;
— *Cardiovascular diseases* — there is increasing evidence of ocular blood flow to be involved in the pathogenesis of glaucoma. Current data indicate that fluctuations in blood flow are harmful in glaucomatous optic neuropathy. Unstable blood pressure and dips are linked to optic nerve head damage and correlate with visual field deterioration;
— *Myopia* — myopic individuals have a two- to threefold increased risk of developing glaucoma comparing with that of non-myopic subjects;
— *Diabetics* are three times more susceptible to develop primary open angle glaucoma;
— *Use of steroids* can also cause glaucoma;
— *History of injury* to the eye.

Persons with any one or combination of these risk factors should be checked regularly for the development of glaucoma.

4. Signs and Symptoms of Glaucoma

The *clinical triad typical for different types of glaucoma* is:
— permanent or periodical increasing of intraocular pressure;
— specific defects of the visual field (fig. 12.3, 12.4);
— pathologic excavation of the optic nerve head (fig. 12.5, 12.6).

Patients with open-angle glaucoma and chronic angle-closure glaucoma in general have no symptoms early in the course of the disease. So it is important to have regular eye tests to check for glaucoma.

Symptoms of open-angle glaucoma and chronic angle-closure glaucoma are:
— gradual loss of central and peripheral vision, usually in both eyes;
— haziness of vision;

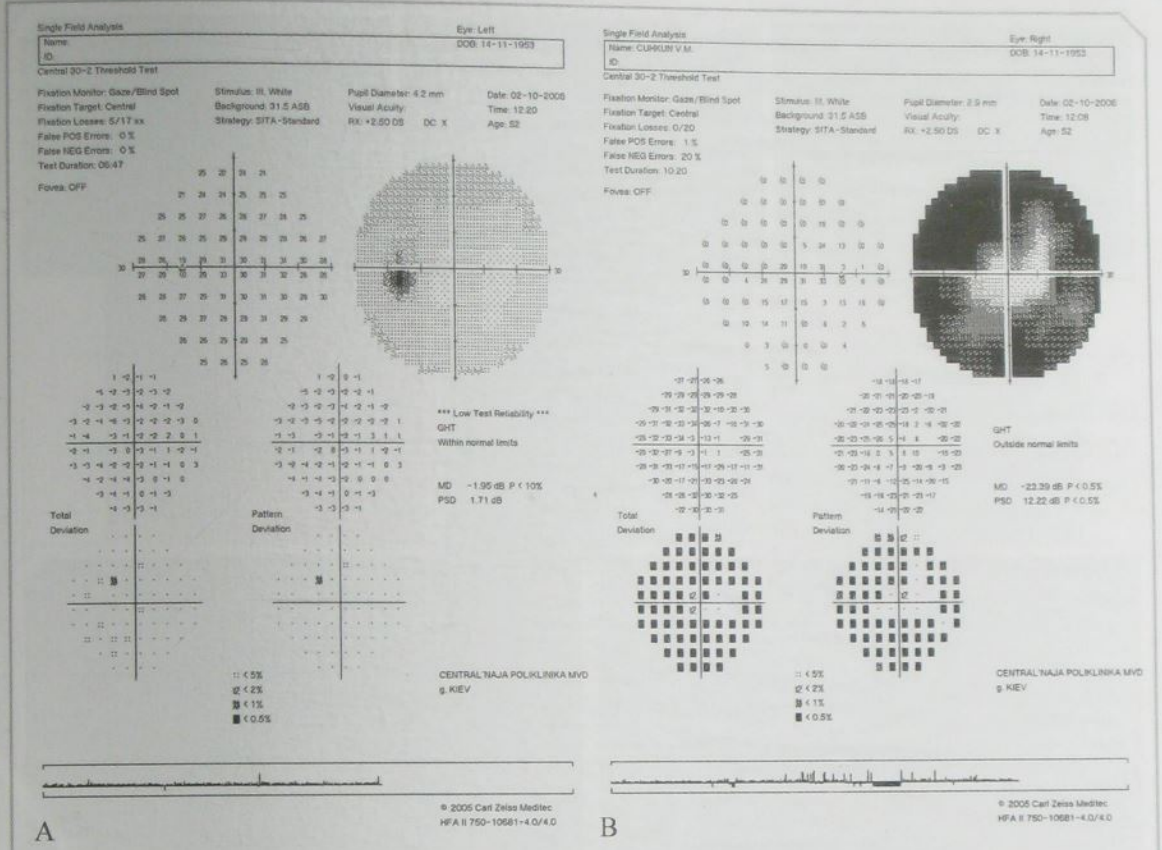
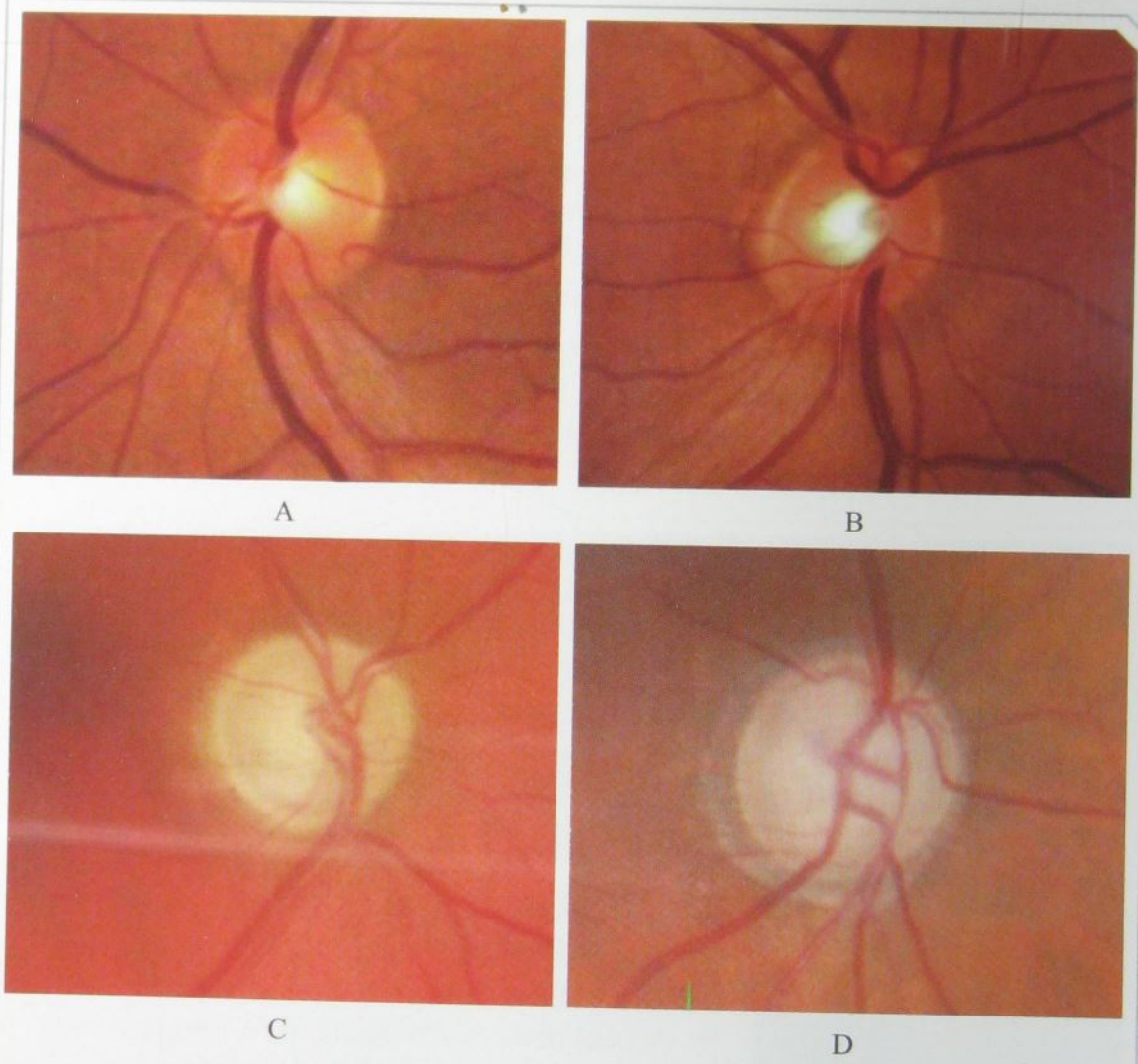


Fig. 12.3. Results of Humphrey static perimetry demonstrating normal full visual field with physiological blind spot (A) and visual field defects with sparing the very central portion of vision at far advanced glaucoma stage (B)

Fig. 12.4. Progressive loss of visual field in glaucoma (normal vision (A); mild generalized constriction of the peripheral visual field (B); arcuate defect with nasal step (C); enlargement of the blind spot, wing-shaped paracentral scotomas isolated or associated with nasal step (D)

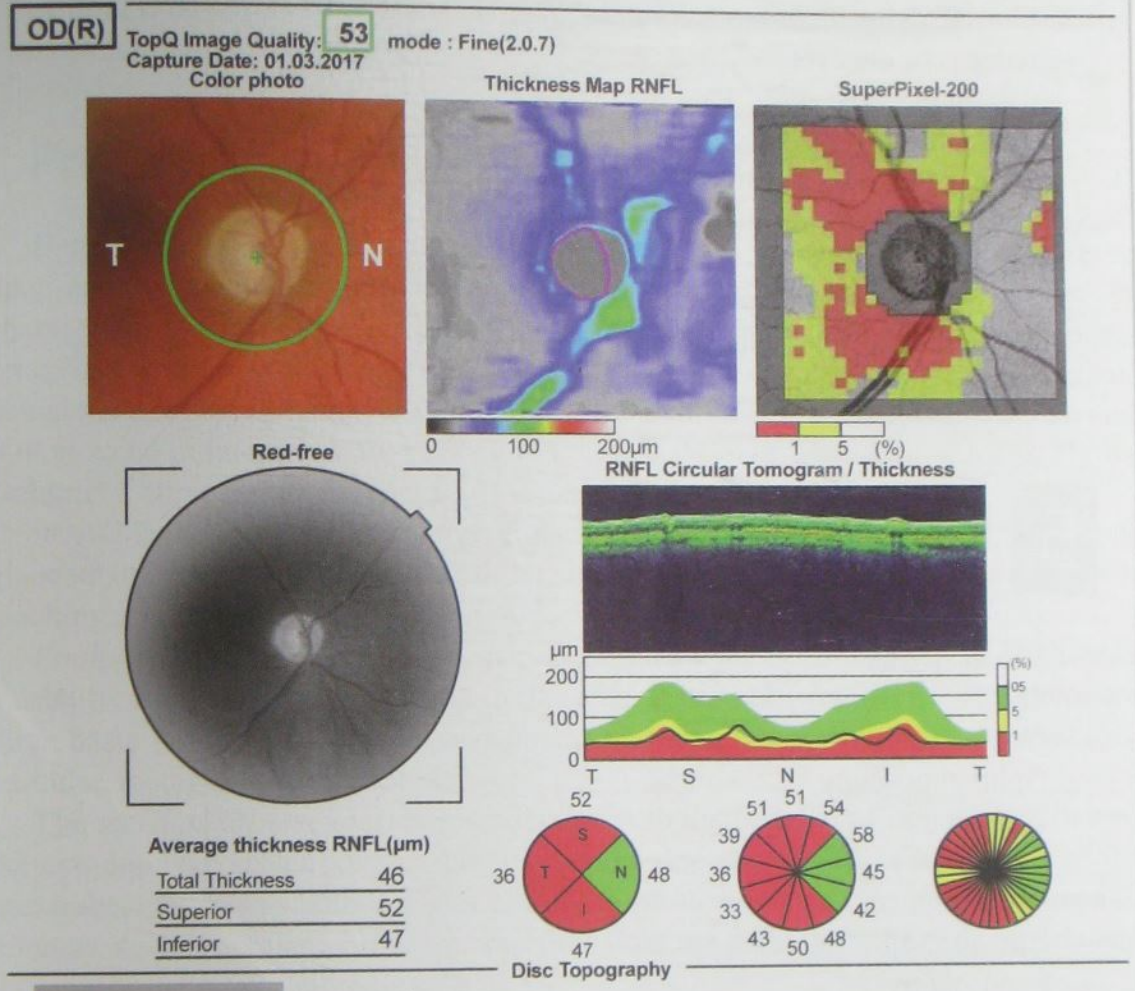
E F G

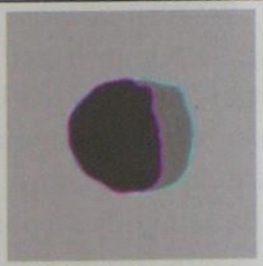
Fig. 12.4. (Continue): arcuate or Bjerrum scotoma with further constriction of peripheral visual field (E); general constriction with sparing of the central vision accompanied by temporal island (tunnel vision) (F); complete visual field defect (temporal island is the most resistant one) (G)



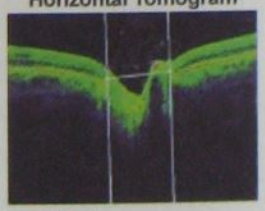
A B C D

Fig. 12.5 Ophthalmoscopic view of optic nerve head cupping progression (normal optic nerve head with physiological excavation (A); optic nerve head cupping at early glaucoma stage (B); cupping of the optic nerve head at moderate (C) and advanced glaucoma (D) stage)



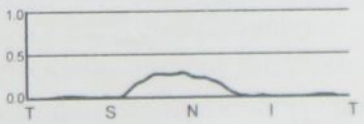


Horizontal Tomogram



Disc Area	(mm^2)	1.67
Cup Area	(mm^2)	1.67
Rim Area	(mm^2)	1.67
C/D Area Ratio		1.67
Linear CDR		1.67
Vertical CDR		1.67
Cup Volume	(mm^3)	1.67
Rim Volume	(mm^3)	1.67
Horizontal D.D	(mm)	1.67
Vertical D.D	(mm)	1.67

R/D Ratio



Disc margin
Cup margin

Disc parameters are determined at the reference plane height of 120 microns from the RPE plane in this version

Comments :

Signature :

Date :

Fig. 12.6. OCT image of optic nerve head examination in a glaucoma patient

— haloes around lights;
— tunnel vision in the advanced stages.

Symptoms of acute glaucoma may include:
— blurred vision and haloes around lights;
— redness of the eye;
— pain in the eye;
— headache;
— feeling sick or vomiting.

Symptoms of congenital glaucoma include:
— watering eyes,
— sensitivity to light, or photophobia;
— cloudy and unusually large eyes.

5. Examination Methods

— Visual Acuity
— Slit-Lamp Examination
— Gonioscopy
— Ophthalmoscopy
— Tonometry
— Pachymetry
— Perimetry
— Ultrasound Biomicroscopy
— Fundoscopy
— OCT
— HRT
— GDx

6. Glaucoma

6.1. Open-Angle Glaucoma

Open-angle glaucoma is a type of glaucoma in which there is no detectable mechanical narrowing or closing of the anterior chamber but the aqueous humor outflow is obstructed by a partial blockage within the trabecular meshwork or a structural

defect of the eye's drainage system resulting in IOP increase, optic nerve damage and corresponding visual field loss.

There are typically no early warning signs or painful symptoms of open-angle glaucoma. It develops slowly and sometimes without noticeable sight loss for many years so it gradually steals vision.

Primary Open-Angle Glaucoma

Definition. Primary open-angle glaucoma (POAG), also referred to as chronic open-angle glaucoma or chronic simple glaucoma, is a type of glaucoma, which is characterized by opened iridocorneal angle, normal anterior chamber depth and absence of a known underlying cause. It is a progressive chronic condition with glaucomatous optic neuropathy, visual field loss compatible with nerve fibre damage and IOP at some point greater than 21 mm Hg (normal range: about 10—21 mm Hg by Goldmann applanation tonometry). It is usually bilateral.

POAG is the most common type of glaucoma and accounts for over 90 % of adult glaucomas. The incidence of the disorder significantly increases beyond the age of 40, reaching a peak between the ages of 60 and 70.

Etiology. The cause of primary open-angle glaucoma is not known. Thus, there is a need for continuous investigation in this area. It has been determined that there are three basic pathophysiological mechanisms in glaucoma development: mechanical, vascular, metabolic, and apoptotic.

The *mechanical mechanism* is associated with disturbance of normal function of the eye drainage system due to clogging of the drainage canals or narrowing of the intratrabecular spaces with aging or as a result of other eye or systemic conditions or traumas. As a consequence, the aqueous fluid does not drain from the eye properly and this causes the steady increase of IOP.

The disorders of the *vascular system* have also been implicated in the development of POAG. There may be a deficit in the rate of blood flow in the intraocular circulation, including the retinal, central retinal artery, optic nerve head circulation, etc.

Among the other *metabolic mechanisms* in glaucoma development the dysregulation of nitric oxide (NO) has a significant role. According to modern concepts, NO plays a unique role as a multifunctional modulator of many physiological processes in the body, and its discovery is considered one of the major achievements of medicine in the last decade. The main biological effect of NO in the eye is the regulation of the microcirculatory blood flow to the inner layers and the optic nerve disc, coordination of transport through the aqueous humor drainage system, and participation in the mechanism of neuronal apoptosis.

The *apoptotic theory* states that genetically programmed destruction of the retinal ganglion cells may play a role in the glaucoma pathogenesis.

Clinical Picture. Most people who have open-angle glaucoma feel fine and do not notice a change in their vision at first because initially side or peripheral vision is lost,

and the visual acuity or sharpness of vision is maintained until late in the disease. Occasionally, patients become aware of earlier visual field defects when performing monocular tasks (such as using the viewfinder of a camera), noticing portions of words missing when reading, or having difficulty with driving, etc.

In cases of high IOP patients may notice haziness of vision or haloes around lights. Late in the disease, however, extensive loss of peripheral and central vision loss will cause "tunnel vision", blind spot, and complete blindness.

Complaints. The majority of patients with POAG do not experience any subjective symptoms for years. However, a small number of patients experience occasional unspecific symptoms such as headache, a burning sensation in the eyes, or blurred or decreased vision that the patient may attribute to lack of eyeglasses or insufficient correction.

Signs. The anterior chamber is of normal depth, the anterior chamber angle is wide, degeneration of the iris, sluggish pupil. The most valuable diagnostic signs are increased IOP (higher than 21 mm Hg), a pathological increase in the excavation of the optic nerve head, asymmetry of excavation in both eyes, visual field defects — enlargement of the blind spot, nasal step defects, arcuate (arc-shaped) scotoma, temporal wedge defects, paracentral scotoma.

Methods of Examination. Visual acuity and slit-lamp examination, gonioscopy, ophthalmoscopy, tonometry, pachymetry, perimetry, OCT, HRT, GDx.

Differential Diagnosis. Ocular hypertension, normal tension glaucoma, primary angle-closure glaucoma, secondary glaucoma, ischemic optic neuropathy, retinal vascular occlusions, compressive nonglaucomatous optic neuropathy, traumatic optic neuropathy, optic nerve swelling, meningiomas or other intraocular tumors.

Treatment. There is no cure for glaucoma at present, but the course of the disease can be slowed or controlled. The problem of glaucoma treatment is very complex and requires a comprehensive approach. First of all, it is aimed at stabilization or preventing of loss of visual functions by reducing the risk factors impact.

The European Glaucoma Society has developed and proposed the following algorithm of glaucoma treatment.

General Principles of Glaucoma Treatment

1. *Medical Treatment:*
   - — hypotensive (decreasing and stabilization of IOP):
     i. improve of aqueous humor outflow through the trabecular and uveoscleral pathways;
     ii. suppress aqueous humor production;
   - — neuroprotection (protection of optic nerve fibres from damage caused by the disease — correction of trophic and circulatory disorders).
2. *Laser Treatment*
3. *Surgical Treatment*

Medical treatment is the treatment of first choice in POAG. Laser and surgical treatments are indicated only in cases of ineffective medical treatment, although they may be accompanied by simultaneous instillation of eye drops.

A number of medications are currently available to treat glaucoma. Typically, medications are used to reduce elevated IOP either by increasing aqueous outflow through the trabecular and uveoscleral pathways or decreasing aqueous humor production. Combination products are also available.

Glaucoma eye drops are divided into medications of the first and second choice.

The *medications of the first choice* are:

— nonselective beta-blockers (Timolol) — administered 2 times a day;
— prostaglandin analogs (Latanoprost, Travoprost, Tafluprost, Bimatoprost) — 1 time a day.

In cases of the lack of effectiveness of the chosen therapy *medications of the second choice* are prescribed. They are:

— selective beta-blockers (Betaxolol) — 2 times a day;
— carbonic anhydrase inhibitors (Dorzolamide, Brinzolamide) — 2—3 times a day;
— selective alpha-adrenergic agonists (Brimonidine) — 2 times a day;
— myotic or cholinergic agents (Pilocarpine) — 4 times a day.

Combined preparations for glaucoma treatment are:

— Dorzolamide + Timolol;
— Brinzolamide + Timolol;
— Latanoprost+ Timolol;
— Travoprost + Timolol;
— Bimatoprost + Timolol;
— Pilocarpine + Timolol;
— Brimonidine + Timolol.

Neuroprotection is an important trend in the treatment of glaucoma. For this purpose, the following systemic preparations are prescribed:

— antioxidants (Superoxide dismutase, Mexidol, Histochrome, Emoxipine);
— neuropeptides (Retinalamin, Semax, Cerebrolysin);
— calcium channel blockers (Nifedipine, Verapamil);
— vasoprotectives (Prodectin, Dicynone, Doxium);
— nootropics (Sermion, Piracetam);
— anti-sclerotic preparations, vitamins and minerals;
— cell therapy is rapidly growing in recent decades.

If medical treatment is ineffective, the next option is *laser treatment*. Nowadays a whole system of different types of laser surgery for glaucoma is introduced into clinical practice, which allows selecting an adequate method for every patient:

— *argon laser trabeculoplasty (ALT)* — argon ("hot") laser is used to create tiny burns on the trabecular meshwork, which causes scarring of the tissue resulting in stretching and then opening of the adjacent trabecular pores and thus improving aqueous humor outflow from the eye;
— *selective laser trabeculoplasty (SLT)* — Nd:YAG ("cold") laser with relatively low power is used to selectively target specific cells (melanocytes) within the trabecular meshwork;

— *cyclophotocoagulation (CPC)* involves application of laser burns to the ciliary body, which causes destruction of the processes tissue, reducing aqueous humor production. This type of laser is typically performed after other more traditional therapies have failed.

In cases when medical and laser treatments are not sufficient *surgery* is indicated with the purpose to create alternative pathways for aqueous humor outflow. Nowadays there are many methods of glaucoma surgery:

— *trabeculectomy* is the most common conventional microsurgical procedure used to treat glaucoma. It involves partial surgical removal of the eye's drainage system and adjacent structures (iris root and sclera), as a result the aqueous humor outflows through the new opening into a reservoir (bleb) underneath the conjunctiva where it is dissolved;

— *non-penetrating deep sclerectomy (NPDS)* is an alternative surgical procedure for glaucoma treatment that does not involve entering into the anterior chamber and the internal trabecular meshwork remains preserved. During the surgery a flap in the outer sclera is made, then a deep piece of the sclera underneath is removed, and a small collagen implant is introduced there. This helps to maintain the intrascleral slit opened, through which the aqueous humor can drain more easily;

— *viscocanalostomy* — a high density viscoelastic substance is injected into Schlemm's canal, in 4—7 days it dissolves and that restores opening of the canals and helps to provide enough space for adequate drainage and eye pressure relief;

— *drainage surgery* involves placing of an artificial drainage device into the eye. It is a microscopic tube attached to a reservoir (or plate). The tube is inserted into the anterior chamber of the eye and the plate is implanted underneath the conjunctiva to allow the aqueous humor to flow out of the eye into a chamber called a bleb. There are several different glaucoma drainage implants. These include the Molteno implant, the Ahmed glaucoma valve implant and Crabic drainage implants (fig. 12.7, 12.8). These are indicated for glaucoma patients not responding to maximal medical therapy, and for those with failed trabeculectomy.

Prognosis. Most people treated for glaucoma will not go on to develop total loss of vision (severe sight impairment). Untreated POAG can lead to irreversible blindness.

Complications. Total blindness.

Prophylaxis. Regular examinations, screening and early diagnostics may help to identify the disease in early stages before irreversible damage occurs.

Normal Tension Glaucoma

Definition. Normal tension (pressure) glaucoma (or low-tension glaucoma) is a form of open-angle glaucoma characterized by progressive optic neuropathy and corresponding loss of peripheral vision even though IOP is within normal ranges or even below.

Fig. 12.7. Crabic glaucoma microdrainage implant and its position on the eyeball (model)

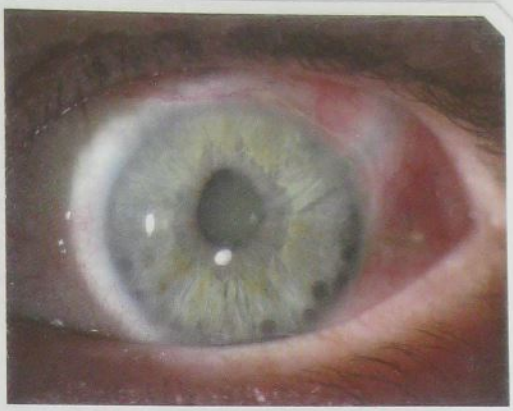


Fig. 12.8. The image of the eye after glaucoma drainage implantation with the presence of the tube in the anterior chamber

Etiology. The causes of optic nerve damage in normal tension glaucoma are mostly unknown, although vascular and mechanical factors have been implicated. It may occur either because of an unusually fragile optic nerve which can be damaged despite normal IOP, or reduced blood flow to the optic nerve. While a fragile optic nerve may be inherited, reduced blood flow to the optic nerve can be due to vascular diseases, including vasospasms and ischemia. It may be associated with migraines.

Complaints. Gradual loss of peripheral and central vision. People do not usually have any visual complaints until late in the course of the disease.

Signs. Clinical signs for normal tension glaucoma are hemorrhages on the optic nerve disc, its cupping with pale neuroretinal rim, visual field loss, sometimes changes in the blood vessels of the conjunctiva.

Methods of Examination. Slit-lamp exam, gonioscopy, tonometry, pachymetry, ophthalmoscopy, funduscopy, perimetry, OCT, HRT.

Differential Diagnosis. Other types of open-angle glaucoma, congenital disc anomaly, optic nerve coloboma, systemic vascular or neurologic pathology.

Treatment. The general principles of medication choice are the same as when managing other types of POAG. The aim is to lower IOP by at least 30 %. Medical treatment options include the drugs, which increase aqueous humor outflow (prostaglandin analogs, alpha-agonists and cholinomimetics), and those, which decrease aqueous production (beta-blockers and carbonic anhydrase inhibitors). Combination products are also available.

Neuroprotection is a therapeutic strategy directed at keeping the retinal ganglion cells alive and functional. Calcium channel blockers may be useful as adjunctive medical treatment in patients with evidence of vasospasm and concurrent hypertension.

If medications are ineffective, another option is laser trabeculoplasty. Patients that continue to progress despite maximum tolerated medical therapy may require trabeculectomy or filtration surgery.

Prognosis. With early diagnosis and medical treatment, further optic nerve damage and/or vision loss may be prevented. If this condition is not detected early, permanent loss of vision can occur.

Complications. Loss of vision is the most common complication.

Prophylaxis. Regular eye examinations (ophthalmoscopy, perimetry, OCT, HRT) are important to screen for optic nerve damage and vision loss despite a normal eye pressure as of its silent nature.

Ocular Hypertension

Definition. Ocular hypertension is a term referring to an elevated IOP without signs of glaucomatous optic neuropathy or visual field defects.

Etiology. High IOP level is caused by an imbalance in the production and drainage of the aqueous humor. It can be due to excessive aqueous humor production or its inadequate drainage through the trabecular meshwork. Certain medications, such as steroids, ocular trauma, other eye conditions including thin cornea, pseudoexfoliation syndrome, pigment dispersion syndrome, etc. can cause high eye pressure. Stress and hormonal dysfunction also are potential causes of ocular hypertension.

Complaints. Most people with ocular hypertension do not experience any symptoms at early stages.

Signs. IOP by Goldmann applanation tonometry above 21 mm Hg but below 30 mm Hg, the anterior chamber angle is opened, the optic nerve disc appears normal and no signs of glaucoma are found during perimetry.

Methods of Examination. Slit-lamp exam, gonioscopy, tonometry, pachymetry, ophthalmoscopy, funduscopy, perimetry, OCT, HRT.

Differential Diagnosis. Primary open-angle glaucoma, secondary open-angle glaucoma, chronic angle-closure glaucoma.

Treatment. The main goal of medical treatment for ocular hypertension is to reduce IOP (by at least 20 %) before it causes damage to the optic nerve. Medical treatment options include drugs that increase aqueous humor outflow (prostaglandin analogs, alpha-agonists and cholinomimetics) and those that decrease aqueous humor production (beta-blockers and carbonic anhydrase inhibitors). Combination products are also available.

Prognosis. Some patients in this group do not develop glaucomatous lesions in spite of continuation of elevated IOP but the others will develop primary open-angle glaucoma. The probability that a patient will develop glaucoma increases with higher IOP, age over 40, diabetes, high myopia and a family history of glaucoma.

Complications. Poor control of elevated IOP may lead to POAG development, which causes glaucomatous optic neuropathy resulting in progressive vision loss.

Prophylaxis. Regular eye examinations and IOP control are very important preventive measures to screen for any optic nerve damages caused by high pressure, so its progression to glaucoma can be prevented.

6.2. Angle-Closure Glaucoma

Angle-closure glaucoma (or **closed-angle glaucoma**) is a glaucoma associated with a mechanical narrowing or closing of the anterior chamber angle by the iris root (fig. 12.9). This condition may arise as a consequence of the anatomy of the eye: some people's angles are naturally very narrow, which makes the angle more vulnerable to blocking off; other susceptible patients include those with a thin iris, a thick lens and a shorter axial length of the eyeball. Due to blockage of the anterior chamber angle progressive trabecular dysfunction develops and junction of the iris and cornea at the periphery of the anterior chamber forms; that results in IOP increase, optic nerve damage, and loss of visual function. Angle-closure glaucoma may be acute, subacute, intermittent, or chronic.

Acute Angle-Closure Glaucoma

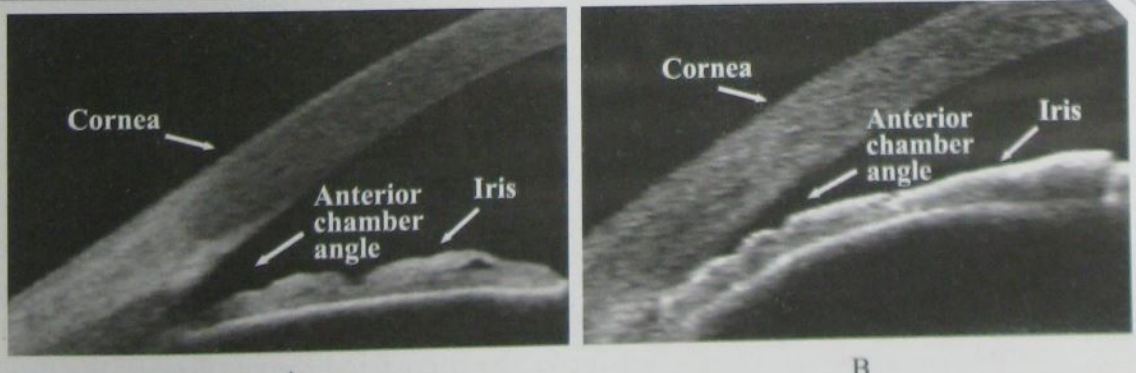
Definition. Acute angle-closure glaucoma (or acute glaucomatous attack) is a sudden and significant increase in IOP resulting from total closure of the anterior chamber angle that blocks aqueous humor outflow.

NOTE!

Acute angle-closure glaucoma is an emergency condition that requires immediate medical treatment to prevent permanent loss of vision.

Etiology. Anatomically predisposed eyes with shallow anterior chambers pose a relative obstruction to the flow of the aqueous humor through the pupil. The pupillary block increases the pressure in the posterior chamber. The pressure displaces the iris anteriorly toward the trabecular meshwork, suddenly blocking aqueous humor outflow (angle closure).

In addition to predisposing ocular anatomy, other factors that provoke sudden dilation of the pupil can contribute to acute glaucomatous attack. This is emotional



A B

Fig. 12.9. OCT image of the opened anterior chamber angle (A) and closed anterior chamber angle (B)

stress or excitement, long and hard work with a tilted head, staying or watching TV or working on a computer in a darkened room, physical exhaustion, hypothermia or overheating of the body. Pharmacologic mydriasis and systemic psychotropic drugs can also trigger a glaucoma attack.

Clinical Picture. Glaucoma attacks often occur at night or when the patient enters a darkened room. The low light conditions cause the pupil to dilate increasing the contact between the lens and the iris, narrowing the angle.

In addition to eye pain, blurry vision, headache, and associated nausea or vomiting, high intraocular pressure leads to corneal swelling (edema), which causes the patient to see haloes around lights.

The full clinical syndrome of acute glaucoma will not always be present. Patients' subjective perception of pain intensity can vary greatly. Some people have milder symptoms, sometimes with intermittent attacks of blurring and haloes without pain. The attack may end when they go into a brighter room or go to sleep. Both of these cause the pupil to constrict and pull the iris away from the drainage channels.

Complaints. Severe eye pain, eye redness, blurry vision, seeing multi-colored haloes around light sources, headache that may be referred to the temples, back of the head, and jaws (via the three branches of the trigeminal nerve), nausea, vomiting, and extreme weakness.

Signs. Vision reduced to counting fingers, the eye is red (conjunctival and ciliary injection), dilated pupil that does not react to light, edema of the cornea and the optic nerve disc (fig. 12.10). IOP is very high (up to 35—60 mm Hg); the eye becomes stone hard on palpation; the anterior chamber is shallow or completely collapsed.

Methods of Examination. Slit-lamp exam, gonioscopy, tonometry, ophthalmoscopy, ultrasound biomicroscopy, OCT.

Differential Diagnosis. Acute conjunctivitis, acute iridocyclitis, endophthalmitis (table 12.1).

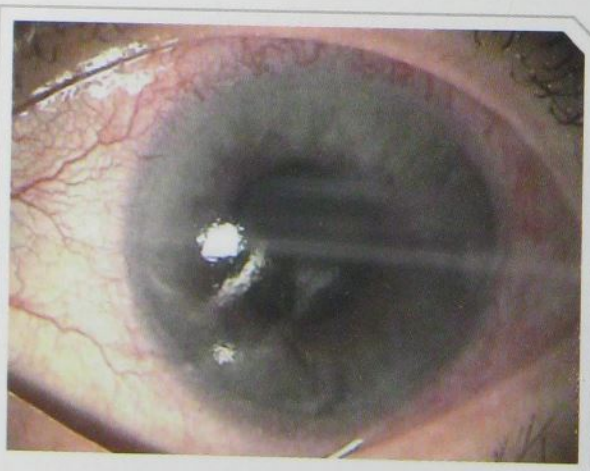


Fig. 12.10. Acute angle-closure glaucoma with presence of mixed injection, dilated fixed pupil, corneal edema

Treatment. Acute angle-closure glaucoma is an emergency, and a patient requires immediate treatment. The initial therapy is conservative; if it fails, surgical treatment is required.

In acute angle-closure glaucoma, several medications are used simultaneously to accelerate and maximize their pressure-lowering effects. The medications lower IOP by increasing the outflow of the fluid (aqueous humor) from the eye or by decreasing the production of fluid in the eye.

Table 12.1

Differential Diagnosis of Acute Conjunctivitis, Acute Iridocyclitis, and Acute Angle-Closure Glaucoma

Clinical Signs	Acute Conjunctivitis	Acute Iridocyclitis	Acute Angle-Closure Glaucoma
1	2	3	4
Visual acuity	Not changed	Reduced	Greatly reduced
Conjunctiva	Conjunctival injection	Ciliary injection, most marked at corneoscleral margin	Mixed (conjunctival and ciliary) injection
Cornea	Clear, sensitive	Clear, sensitive	Hazy, insensitive
Anterior chamber	Normal depth	Normal depth	Very shallow
Pupil size	Normal, round	Small, irregular	Dilated, oval
Pupil response to light	Active	Minimal	No reaction
IOP	Normal	Slightly raised	Extremely raised
Eye pain	Absent	Moderate	Severe
Photophobia	Slight	Significant	None
Multi-colored haloes around the source of light	Not seen	Not seen	Yes
Discharge	Sticky	Watery	None
General condition	Not changed	Not changed	Nausea, vomiting, and extreme weakness

Topical treatment:

— *Pilocarpine 1 %* eye drops every 15 min for an hour, followed by instillation every hour. Further instillation rate is reduced to 6 times a day. It is a *myotic drug* that causes the pupil to constrict (narrow) and help to move the iris away from the trabecular meshwork, which opens up the obstruction to the outflow of the aqueous humor.

— *Timolol 0.25—0.5 %* 2 times a day (a *beta-blocker* medication to reduce fluid production in the eye).

— *Dorzolamide 2 %* 3 times per day or *Brinzolamide 1 %* 2 times per day (*carbonic anhydrase inhibitors* to reduce aqueous humor production).

— *Dexamethasone* (a *topical steroid*) to reduce anterior chamber inflammation and the chance of both anterior and posterior synechiae formation.

Systemic treatment:

— *Acetazolamide 500 mg* intravenously; it may be given orally but the onset of action is not rapid, 2—3 times per day (*systemic carbonic anhydrase inhibitors* — inhibitors of aqueous humor production).

— *Mannitol 20 % 1—2 g/kg* intravenously, or *Glycerol 50 % 1—1.5 g/kg* orally (*hyperosmotic agents* for removing fluid from the eye).

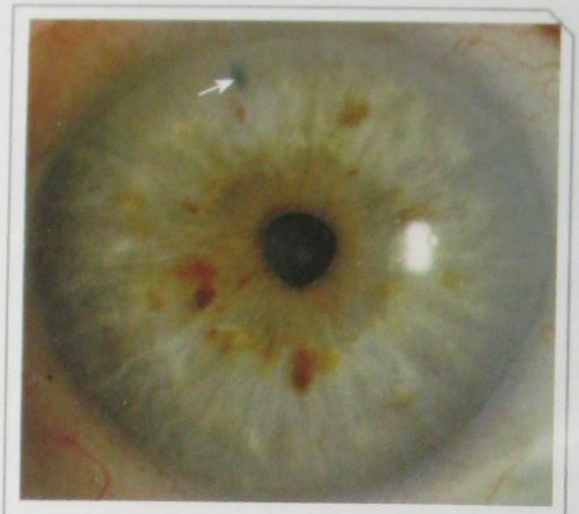


Fig. 12.11. The state after peripheral laser iridotomy with the presence of the hole in the iris (shown with an arrow)

— *Analgesics and sedatives.*

Along with drug therapy revulsive procedures are indicated: hot foot baths, mustard plasters on the gastrocnemius muscles, salt laxatives and hirudotherapy (leeches to the temple).

Laser treatment:

— *Peripheral laser iridotomy (LPI)* — a laser treatment to create communication between the posterior and anterior chambers by making an opening in the peripheral iris (fig. 12.11).

If the attack failed to stop within 12—24 hours, surgical treatment is indicted.

— *Peripheral iridectomy* — a surgical (or incisional) treatment with the help of which a small, triangular hole in the iris is created.

— *Phacoemulsification* — lens extraction. Thickening of the lens contributes to angle closure. A normal cataract is about 4.5 mm thick, and the lens implant is about 1 mm, so there is about 3.5 mm extra space for the aqueous humor to circulate.

Prognosis is good with early detection and treatment, vision in the affected eye may return to a level that is almost the same as what it was before the episode began.

Complications. Peripheral anterior synechiae, cataract, atrophy of the retina and optic nerve, absolute glaucoma, loss of vision are the most common complications.

Prophylaxis. Regular eye examinations by an ophthalmologist may identify people who are at risk of acute angle-closure glaucoma. In some people who are at high risk a laser iridotomy may be performed to prevent an attack of acute angle-closure glaucoma.

6.3. Congenital Glaucoma

Definition. Congenital glaucoma (childhood or pediatric glaucoma) is a type of glaucoma that is present at birth as a result of the drainage canals not forming properly. However, its manifestations may not be recognized until infancy or early childhood. It occurs once every 12,000—18,000 births and accounts for about 1 % of all glaucomas.

Etiology. Congenital glaucoma is a rare condition that may be inherited or caused by incorrect development of the eye's drainage system before birth. This leads to increased intraocular pressure, which in turn damages the optic nerve.

Clinical Picture. The eye of a young child enlarges in response to increased intraocular pressure because it is more pliable than the eye of an adult. Children with this disorder are irritable, poor eaters, and rub their eyes often. Physicians should be alert to parents who boast about their child's "big beautiful eyes" and should measure IOP. It is essential to diagnose the disorder as early in the child's life as possible to minimize the risk of loss of or irreversible damage to the child's vision.

Complaints. Symptoms of congenital glaucoma include enlarged eyes, clouding cornea, photophobia and tearing (fig. 12.12).

Signs. Corneal enlargement and clouding, conjunctival erythema, abnormally deep anterior chamber, myopia and/or astigmatism, elevated IOP and enlarged optic nerve cupping.

Normal intraocular pressure is lower in infants and young children than in adults. A newborn has an average intraocular pressure of 10—12 mm Hg, increasing to 14 mm Hg by 7 or 8 years of age. An asymmetric or elevated measurement in the presence of other clinical signs helps make the diagnosis of glaucoma.

NOTE!

A newborn's cornea is typically 9.5—10.0 mm in diameter and increases to 10.5—11.5 mm by age 1.

Any diameter in newborns above 10 mm, and if there is asymmetry between the two eyes, this is a sign of glaucoma!

Methods of Examination. Slit-lamp exam, corneal measurements, tonometry, gonioscopy, ophthalmoscopy.

Differential Diagnosis. Congenital clouding of the cornea, congenital anomalies of the nasolacrimal duct.

Treatment. The medical treatment of childhood glaucoma is not effective; it is treated surgically. Medical therapy is used only as a temporary measure prior to surgery and to maximize pressure control after surgery.

The aim of pediatric glaucoma surgery is to reduce IOP either by increasing aqueous humor outflow from the eye or decreasing humor production within the eye. One operation for pediatric glaucoma is goniotomy. Other surgical options are trabeculotomy, trabeculectomy, and glaucoma drainage tubes.

Prognosis is good in 80—90 % of the patients if treated early. Congenital glaucoma results in blindness in 2—15 % of childhood patients. When childhood glaucoma is not recognized and treated promptly, permanent visual loss ensues.

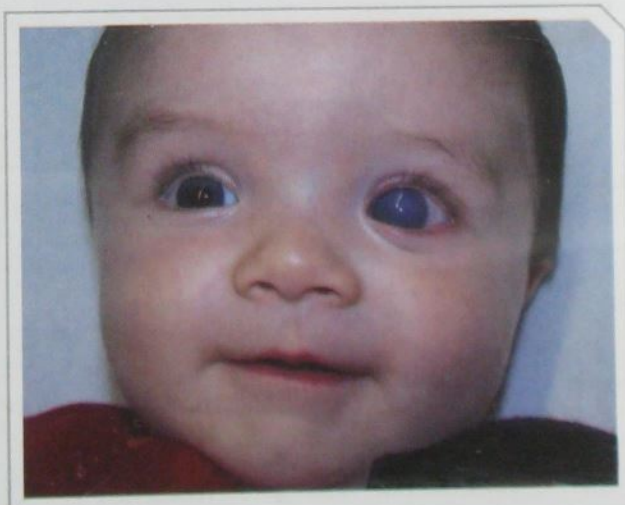


Fig. 12.12. Congenital glaucoma of the left eye (from http://ru-babyhealth.ru)

Complications. Some patients may have the following complications — amblyopia, myopia, astigmatism, retinal detachment, and lens dislocation.

Prophylaxis. There is no known way to prevent primary congenital glaucoma. Early detection and treatment are essential to maximize visual potential.

6.4. Secondary Glaucoma

Secondary glaucoma is a type of glaucoma that develops as a result of other eye conditions, which lead to increased IOP, glaucomatous changes, and vision loss. It can be caused by an eye trauma, inflammation, advanced cases of cataract, diabetes or vascular diseases and tumors. It may also arise in patients with prolonged use of corticosteroids, laser or conventional surgery, retinal detachment or retinal vein occlusion or blockage.

Treatment of secondary glaucoma is directed toward both the underlying problem as well as lowering pressure with standard glaucoma therapies. In early stage when no damage has been done to the optic nerve removing the cause of the disease leads to its cure, but some types of secondary glaucoma are difficult to treat.

According to the cause secondary glaucoma can be of different types with its peculiar clinical picture and symptoms.

Pigmentary glaucoma is a type of inherited open-angle glaucoma, which is caused by deposition of the iris pigment granules in the trabecular meshwork leading to progressive trabecular dysfunction and ocular hypertension with associated glaucomatous optic neuropathy.

Exfoliative glaucoma (pseudoexfoliation or **pseudoexfoliative glaucoma)** is a type of open-angle glaucoma caused by abnormal accumulation of protein in the drainage system and other structures of the eye that leads to blockage of aqueous humor outflow resulting in IOP increase (fig. 12.13). As a group, patients with exfoliative glaucoma show higher pressures and faster disease progression than patients with classic primary open-angle glaucoma. Exfoliative glaucoma arises secondary to pseudoexfoliation syndrome, a systemic disease. Exact etiology of this condition remains unknown.

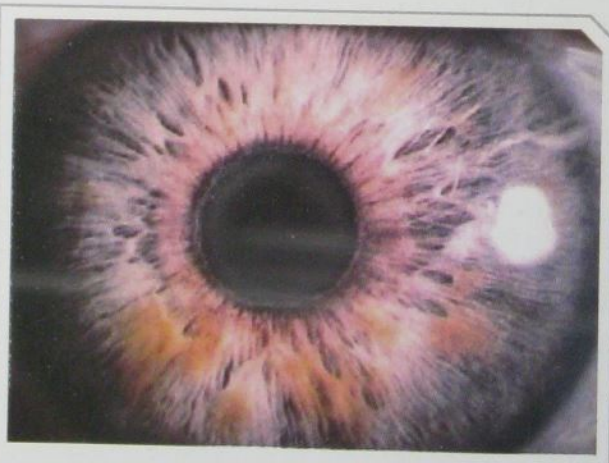


Fig. 12.13. Pseudoexfoliation glaucoma with deposition of exfoliative material on the pupillary margin and anterior lens capsule

Neovascular glaucoma is a form of open-angle glaucoma caused by

abnormal formation of new blood vessels on the iris and over the eye's drainage channels that block the outflow of the aqueous humor causing an increase in IOP. It is more often associated with advanced diabetes, central retinal artery occlusion, intraocular tumors, long-standing retinal detachment and chronic intraocular inflammation.

Uveitic Glaucoma (uveitis glaucoma) is a type of open-angle glaucoma caused by chronic inflammation of the uvea. Uveitis can cause increased IOP when inflammatory debris obstructs the trabecular meshwork resulting in decreased humor outflow from the eye. In the long-term, inflammation can also cause scar tissue that further obstructs fluid outflow. In addition, long-term corticosteroid treatment can also cause elevated IOP as a side effect.

Steroid-induced glaucoma is a type of open-angle glaucoma caused by prolonged use of corticosteroids in any form that results in IOP increase. Withdrawal of the medication usually eliminates these effects.

Phacolytic glaucoma is a type of acute open-angle glaucoma occurring secondary to hypermature cataract and occlusion of the trabecular drainage meshwork with the lens material.

Phacomorphic glaucoma is a type of angle-closure glaucoma caused by excessive enlargement of the lens and its opacification that leads to narrowing or even closure of the anterior chamber angle resulting in IOP increase. It can be successfully treated surgically but only at initial stages until the optic nerve is damaged.

Traumatic glaucoma (or **post-traumatic glaucoma)** is a type of glaucoma caused by an eye injury. It can develop as a result of either blockage of the anterior chamber angle with blood cells, adjacent anatomical structures, or by damage to the drainage meshwork that leads to intraocular pressure increase. This form of glaucoma can occur immediately after the injury or develop years later.

Review:

1. Key Points

Glaucoma is a progressive optic neuropathy leading to irreversible loss of visual functions and blindness. Vision lost due to glaucoma cannot be restored. It has been nicknamed "the silent sight thief" as it usually causes no symptoms early in its course. Glaucoma cannot be prevented; however, regular examinations by an ophthalmologist may help to identify the disease in early stages before irreversible damage occurs. There is no cure for glaucoma at present, but the course of the disease can be slowed or controlled.

Glaucoma can be classified as following: according to the presence or absence of causative factors — *primary* or *secondary*; according to the state of the anterior chamber angle — *open-angle, angle-closure* or *mixed*; according to the onset — *acute* or *chronic*; according to the age of onset — *congenital, infantile, juvenile* or *adult*; according to the IOP level — *normal tension glaucoma, high tension glaucoma, ocular hypertension*; according to the stage of the disease — *mild, moderate, advanced, far advanced, end stage*; according to the dynamics of vision — *stabilized* or *unstabilized.*

The anterior chamber of the eye is filled with the aqueous humor, which is produced constantly by the ciliary body and drains out of the eye through the trabecular or uveoscleral outflows. The relationship between aqueous humor production and its outflow determines IOP.

The *clinical triad* typical for different types of glaucoma is: permanent or periodical increase of IOP; specific defects of the visual field; pathologic excavation of the optic nerve head. The *examination* consists of visual acuity, slit-lamp examination, gonioscopy, ophthalmoscopy, tonometry, pachymetry, perimetry, ultrasound biomicroscopy, fundoscopy, OCT, HRT, GDx.

Open-angle glaucoma is a type of glaucoma, in which there is no mechanical narrowing or closing of the anterior chamber. *Angle-closure glaucoma* is a glaucoma associated with a mechanical narrowing or closing of the anterior chamber angle.

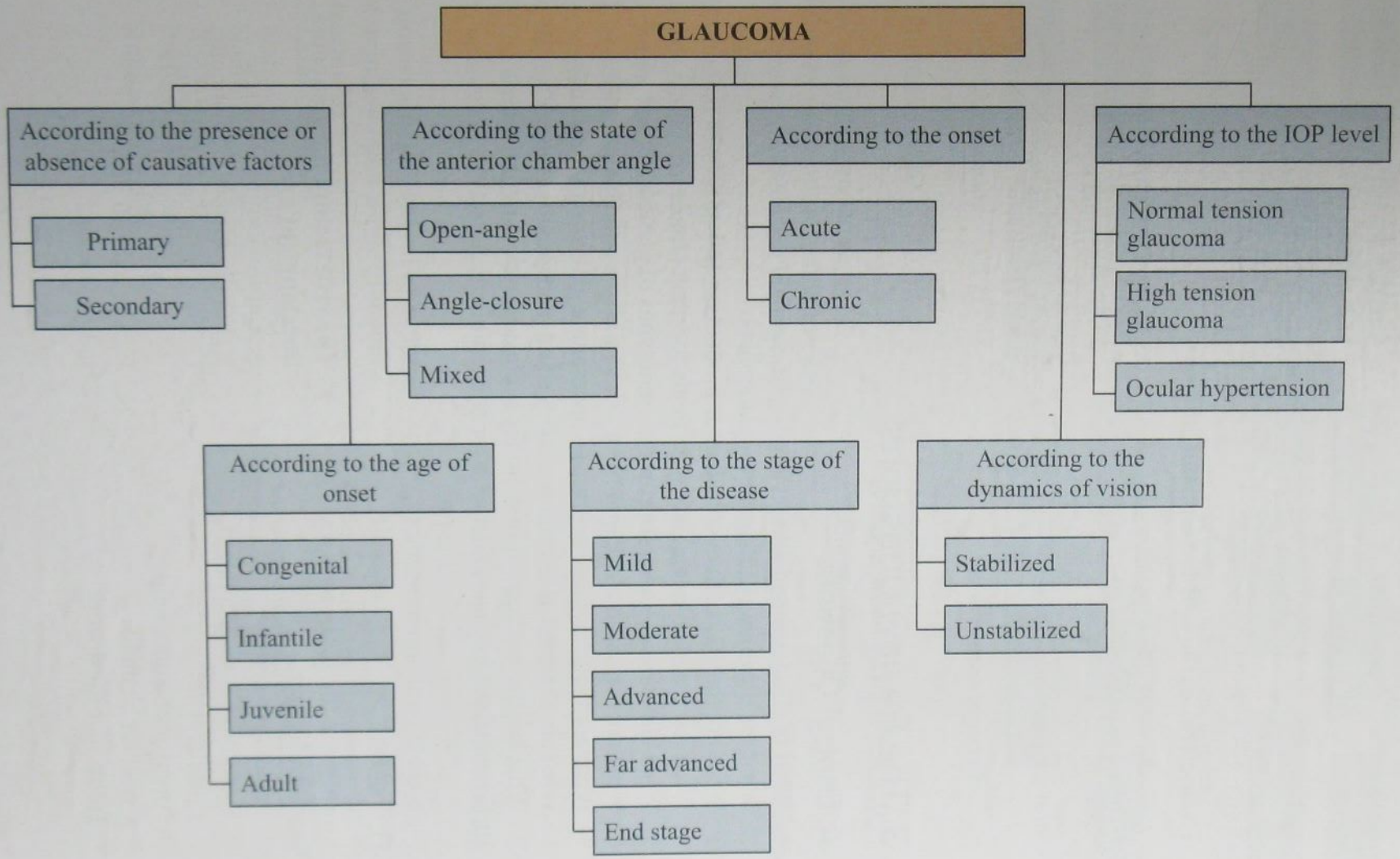
Primary open-angle glaucoma treatment consists of three sequential steps — medical, laser, surgical treatment. The aim of treatment is IOP lowering and neuroprotection. Medical treatment is a treatment of the first choice in POAG. Laser and surgical treatments are indicated only in cases of ineffective medical treatment, although they may be accompanied by simultaneous instillation of eye drops.

Acute angle-closure glaucoma is an emergency condition that requires immediate medical treatment to prevent permanent loss of vision. The initial therapy is conservative; if it fails, surgical treatment is required.

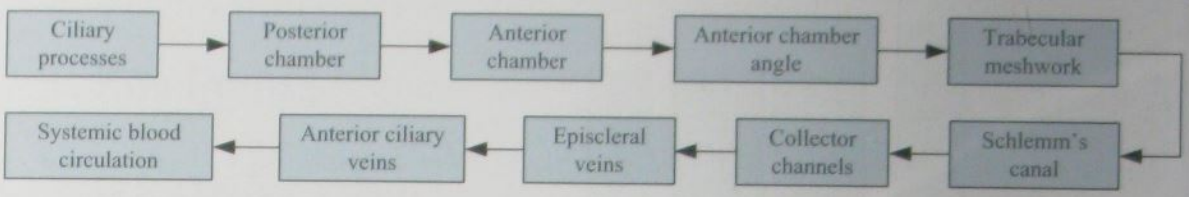
Congenital glaucoma is present at birth. It is treated surgically.

Secondary glaucoma develops as a result of other eye conditions. Treatment of secondary glaucoma is directed toward the underlying problem and standard glaucoma therapies.

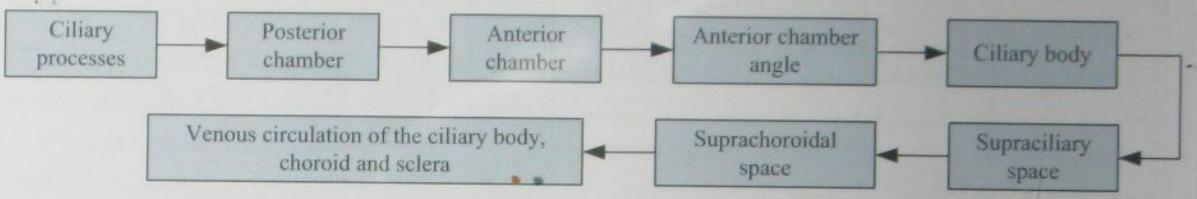
2. Diagrams


GLAUCOMA
According to the presence or absence of causative factors
Primary
Secondary
According to the state of the anterior chamber angle
Open-angle
Angle-closure
Mixed
According to the onset
Acute
Chronic
According to the IOP level
Normal tension glaucoma
High tension glaucoma
Ocular hypertension
According to the age of onset
Congenital
Infantile
Juvenile
Adult
According to the stage of the disease
Mild
Moderate
Advanced
Far advanced
End stage
According to the dynamics of vision
Stabilized
Unstabilized

Trabecular outflow of the aqueous humor



Uveoscleral outflow of the aqueous humor



3. The Review Questions

A. Control Questions

1. What is glaucoma and what types of glaucoma do you know?
2. What are the main risk factors for glaucoma?
3. What are the ways of aqueous humor production and outflow?
4. What are the main examination methods for glaucoma?
5. What clinical triad is typical for different types of glaucoma?
6. What are the cardinal symptoms of primary open-angle glaucoma?
7. What are the principles of primary open-angle glaucoma treatment?
8. What are the cardinal symptoms of an acute glaucomatous attack?
9. What are the methods of first aid for acute angle-closure glaucoma?
10. What are the treatment methods for congenital glaucoma?

B. Tests

1. **Enumerate the main pathways of aqueous humor outflow:**
   A. Schlemm's canal
   B. Systemic blood circulation
   C. Episcleral veins
   D. Anterior chamber angle
   E. Collector channels
   F. Trabecular meshwork

2. **The risk factors for glaucoma development are:**
   A. Cardiovascular diseases
   B. Family history of glaucoma
   C. Long term use of steroids
   D. Elevated IOP
   E. Age over 40 years
   F. Eye trauma

3. **The main clinical signs of glaucoma are:**
   A. Elevated IOP
   B. Watering eyes
   C. Excavation of the optic nerve head
   D. Worsening of dark adaptation
   E. Gradual loss of central vision
   F. Specific defects of the visual field

4. **The main diagnostic methods of glaucoma are:**
   A. Tonometry
   B. Gonioscopy
   C. Pachymetry
   D. Ophthalmoscopy
   E. Perimetry
   F. Biomicroscopy

5. **The main diagnostic signs of primary open-angle glaucoma are:**
   A. Excavation of the optic nerve head
   B. Gradual loss of central and peripheral vision
   C. Increased IOP
   D. Haloes around lights and blurry vision
   E. Severe eye pain and headache
   F. The anterior chamber angle is opened

6. **Enumerate the general principles of primary open-angle glaucoma treatment:**
   A. Laser treatment
   B. Nonselective beta-blockers
   C. Neuroprotection
   D. Medications of the second choice
   E. Prostaglandin analogs
   F. Surgical treatment

7. **The main symptoms of acute angle-closure glaucoma are:**
   A. Blurred vision
   B. Redness of the eye
   C. Severe pain in the eye
   D. Headache
   E. Extreme weakness and vomiting
   F. Haloes around light sources

8. **The general principles of acute angle-closure glaucoma medical treatment are:**
   A. Pilocarpine 1 % every 15 min during 1 hour
   B. Analgesics and sedatives
   C. Revulsive therapy
   D. Antibiotics
   E. Pilocarpine 1 % 3 times a day
   F. Osmotic therapy

9. **The main symptoms of congenital glaucoma are:**
   A. Increased IOP
   B. Buphthalmos
   C. Megalocornea
   D. Anisocoria
   E. Ptosis
   F. Corneal edema

10. **Indications for glaucoma surgery are:**
    A. Failure of maximal medical therapy
    B. Phacolitic glaucoma
    C. Phacomorphic glaucoma
    D. Congenital glaucoma
    E. Ocular hypertension
    F. Acute angle-closure glaucoma

C. Clinical Cases

Case 1

A 52-year-old female patient complains of mist before his right eye in the mornings that disappears without assistance. Periodically she has burning ache in the eye, headache on the right side. Objectively: VA — 1.0, the anterior camber is of normal width, the anterior chamber angle is opened, IOP — 29.0, the visual field is normal, distension of the physiological optic nerve head excavation. Make a diagnosis.

A. Primary open-angle glaucoma
B. Normal tension glaucoma
C. Ocular hypertension
D. Acute angle-closure glaucoma
E. Congenital glaucoma
F. Phacomorphic glaucoma

Case 2

What treatment should you prescribe as an initial treatment to the patient according to in Case 1?

A. Timolol 0.25—0.5 % 2 times a day
B. Analgesics
C. Corticosteroids
D. Prostaglandin analogs 1 time a day
E. Atropine 1 % 4 times a day
F. Pilocarpine 1 % 4 times a day

Case 3

An 81-year-old woman complains of severe eye pain, headache, blurred vision, rainbow-colored haloes around light sources, nausea and vomiting. Visual acuity in the affected eye is of hand motions; conjunctival and ciliary injection; the pupil is dilated and does not react to light; edema of the cornea. The anterior chamber is shallow, the anterior chamber angle is closed. IOP is 56 mm Hg. Make a diagnosis.

A. Primary open-angle glaucoma
B. Normal tension glaucoma
C. Ocular hypertension
D. Acute angle-closure glaucoma
E. Congenital glaucoma
F. Phacomorphic glaucoma

Case 4

What treatment should you prescribe to the patient from Case 3 in case of emergency?

A. Timolol 0.25—0.5 % 2 times a day
B. Analgesics

C. Corticosteroids
D. Osmotic therapy
E. Atropine 1 % eye drops 4 times a day
F. Pilocarpine 1 % eye drops every 15 min for an hour, then every hour

Case 5

A 67-year-old patient complains of sudden severe eye pain and ache around the eye, reduced vision with haloes seen around light sources, headache, extreme weakness. Under examination: vision acuity — counting fingers, red eye — the injection is mixed, the pupil is dilated and does not react to light, corneal edema, the anterior chamber is shallow, IOP 60 mm Hg. Maximal medical treatment for 12 hours could not stop the attack. Make a diagnosis and determine the methods of treatment.

A. Primary open-angle glaucoma, end stage — Timolol 0.25 0.5 % 2 times a day
B. Ocular hypertension — combined antiglaucoma preparations
C. Acute angle-closure glaucoma — instillation of Pilocarpine 1 % every hour
D. Acute angle-closure glaucoma — hyperosmotic agents intravenously and orally
E. Acute angle-closure glaucoma — peripheral laser iridotomy
F. Phacomorphic glaucoma — phacoemulsification with IOL implantation

Case 6

A 47-year-old woman complains of progressive loss of central and peripheral vision, particularly noticeable during migraine attacks. Objectively: VA with best correction 0.8—0.9, the anterior chamber and anterior chamber angle are normal, IOP OD — 18 mm Hg, OS — 16 mm Hg, glaucomatous disc changes, enlargement of the blind spot, and peripheral scotomas in the visual field. Make a diagnosis.

A. Primary open-angle glaucoma
B. Secondary open-angle glaucoma
C. Normal tension glaucoma
D. Ischemic optic neuropathy
E. Optic atrophy
F. Papilledema

Case 7

A mother referred to an ophthalmologist because she noticed watering eyes in her 6-month-old daughter: her eyes look cloudy and larger than normal. The girl rubs her eyes, tries to keep the eyes mostly closed during the day. Objectively: epiphora, conjunctival erythema, photophobia, blepharospasm, corneal edema, the cornea size is 12 mm, the anterior chamber is deep, IOP 16 mm Hg, enlarged optic nerve cupping. Make a diagnosis and the method of treatment.

A. Primary open-angle glaucoma — Timolol 0.25 % twice a day
B. Acute angle-closure glaucoma — laser iridotomy
C. Megalocornea — correction of refractive errors
D. Congenital glaucoma — medical treatment, Timolol 0.25 % twice a day

E. Congenital glaucoma — surgical treatment, goniotomy
F. Congenital cataract — cataract phacoemulsification with IOL implantation

Case 8

A 71-year-old man complains of gradual loss of vision, mild pain in the eye and floaters. Objectively: the anterior chamber is normal, the anterior chamber angle is moderately open OU, gray-white flakes on the pupillary borders and anterior lens capsule, increased trabecular meshwork pigmentation, poor reaction of the pupil to light, iridodenesis, phacodenesis, IOP of OD — 22 mm Hg, OS — 42 mm Hg, optic nerve cupping of both eyes. Make a diagnosis.

A. Primary open-angle glaucoma
B. Acute angle-closure glaucoma
C. Pseudoexfoliation glaucoma
D. Uveitic glaucoma
E. Neovascular glaucoma
F. Pigmentary glaucoma

Case 9

A 63-year-old man complains of acute pain, blurred vision, rainbow-colored halos around lights, nausea, and vomiting. Objectively: cornea edema, injection of the conjunctival and episcleral vessels, a shallow central anterior chamber, a mid-dilated, sluggish, irregular pupil, cataract, enlargement and forward displacement of the lens, the angle is closed, IOP — 35 mm Hg. Make a diagnosis.

A. Chronic angle-closure glaucoma
B. Acute angle-closure glaucoma
C. Phacolytic glaucoma
D. Phacomorphic glaucoma
E. Mature senile cataract
F. Hypermature senile cataract

Case 10

A 59-year-old woman complains of reduced vision, eye pain, photophobia, headache, nausea, and/or vomiting. The patient has advanced diabetes. Objectively: VA — counting fingers, conjunctival injection, corneal edema, a fixed, mid-dilated pupil, ectropion of the uvea, neovascularization of the iris, presence of neovascular capillaries at the pupillary margin, fibrovascular membranes at the trabecular meshwork, synechial angle closure, IOP — 50 mm Hg, optic nerve cupping. Make a diagnosis.

A. Chronic angle-closure glaucoma
B. Acute angle-closure glaucoma
C. Pseudoexfoliation glaucoma
D. Uveitic glaucoma
E. Neovascular glaucoma
F. Pigmentary glaucoma

CHAPTER 13

Diseases of the Retina

OBJECTIVES

Upon completion of the chapter the students should be able to:
- know the main pathologies of the retina and their classification;
- know the basic diagnostic methods of retinal examination;
- describe normal and abnormal fundus examination findings;
- evaluate and manage patients with retinal pathology;
- know the principles of retinal disease treatment.

Plan:

1. CLASSIFICATION OF RETINAL DISEASES
2. SYMPTOMS OF RETINAL DISEASES
3. EXAMINATION METHODS
4. RETINAL DISEASES
   - **4.1. Normal Ophthalmoscopic Findings**
   - **4.2. Degenerative Disorders**
     - Age-Related Macular Degeneration
     - Retinitis Pigmentosa
   - **4.3. Vascular Diseases**
     - Central Retinal Artery Occlusion
     - Central Retinal Vein Occlusion
   - **4.4. Retinal Detachment**
   - **4.5. Inflammatory Diseases**
     - Retinitis
   - **4.6. Tumors**
     - Retinoblastoma

1. Classification of Retinal Diseases

Retinal diseases:

— by the time of onset may be congenital (developmental or hereditary) or acquired;

— by clinical forms may be degenerative, vascular, inflammatory, retinal detachment, tumors.

1.1. Congenital Anomalies: coloboma, congenital anomalies of the retinal vessels, Coats' disease, Leber's congenital amaurosis, etc.

1.2. Degenerative Disorders: pigmentary retinal degeneration (retinitis pigmentosa), hereditary macular degeneration, age-related macular degeneration, macular edema, central serous retinopathy, peripheral retinal degeneration, retinoschisis.

1.3. Vascular Diseases: central retinal vein thrombosis, occlusion of the central retinal artery and its branches.

1.4. Retinal Detachment: rhegmatogenous, tractional, exudative; retinal breaks.

1.5. Inflammatory Diseases: retinitis, retinal vasculitis.

1.6. Tumors: retinoblastoma, astrocytoma, hemangioma.

2. Symptoms of Retinal Diseases

— Visual impairment or total loss of vision.
— Blurred central vision.
— Distorted vision (metamorphopsia — straight objects appear bent).
— A decrease (micropsia) or increase (macropsia) in the size of objects.
— Scotomas in or loss of central vision.
— Scotomas in the peripheral visual field.
— Appearance of spots or lines that appear to "float" in the vision and may look like insects.
— Flashes of light, which can appear as 'lightning streaks' to the side of vision.
— Reduction of night vision.

3. Examination Methods

— Direct ophthalmoscopy.
— Indirect ophthalmoscopy.
— Slit-lamp ophthalmoscopy.
— Ultrasonography.
— Ophthalmochromoscopy.
— Electroretinography.
— Funduscopy photography.
— Fluorescein angiography.
— Optical coherence tomography.
— Heidelberg retinal tomography.
— Perimeter test.

4. Retinal Diseases

The topicality of ophthalmic diseases is determined by the fact that a person gets 90 % of information about the world through the visual analyzer. Any pathological changes of the photoreceptor segment of the visual analyzer could lead to visual impairment or blindness.

The retina is the inner membrane of the eye, which is located between the choroid and the hyaloid membrane. It is the peripheral part of the visual analyzer. It converts light signals into electrical impulses that are transmitted to the brain, which finally interprets them as visual images.

4.1. Normal Ophthalmoscopic Findings

Ophthalmoscopy (may also be called funduscopy) is the method of interior eyeball examination that enables visualization of the condition of the retina, optic disc, and internal blood vessels.

Normal fundus (fig. 13.1):

— *The retina* is normally completely transparent without any intrinsic color. It receives its uniform bright red coloration from the vasculature of the choroid.

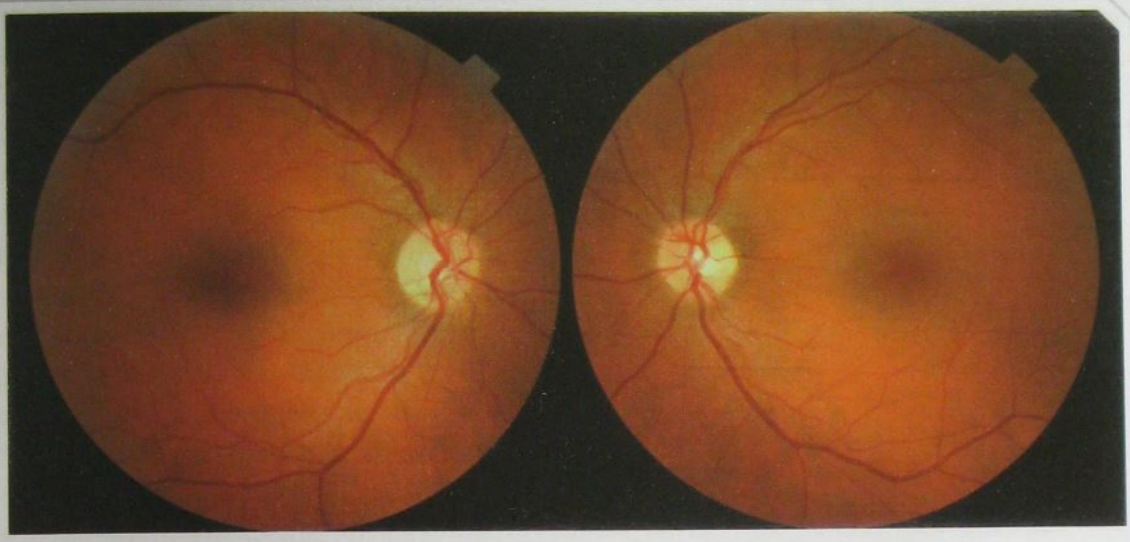


Fig. 13.1. Normal ophthalmoscopic picture of the fundus

The vessels of the choroid themselves are obscured by the retinal pigment epithelium. Loss of transparency of the retina is a sign of an abnormal process (for example, in retinal edemas the retina appears whitish yellow).

— *The optic disc* is located nasally. It measures about 1.5 mm in diameter and its usual form is round or vertically oval. It has pale red or yellowish color. The neuroretinal rim is pink; the edge of the optic disc may be slightly elevated. In the center of the optic disc there is a slight depression known as the optic or physiologic cup. The cup looks as a pale area located off-center in the disc.

— *The retinal arteries and veins* emerge from the nasal side of the optic disc. The vessels directed temporally have an arching course; those directed nasally have a radial course. The veins are relatively large and dark red, whereas the arteries are relatively thin and pale. Venous pulsation on the vessels on the disc may be seen (it is absent in approximately 20 % of normal individuals). The normal ratio of the thickness of the arteries and veins is 2:3.

— *The macula* is a 2.5 mm-diameter area that is located between the superior and inferior temporal blood vessel arcades. It looks slightly darker than the surrounding retina because of increased pigment. It is centered about 2.5 disc diameters temporally to the optic disc. The fovea lies in the center of the macula.

4.2. Degenerative Disorders

Age-Related Macular Degeneration

Definition. Age-related macular degeneration (AMD) is a progressive degeneration of the macula in elderly patients (fig. 13.2). It is a leading cause of vision loss

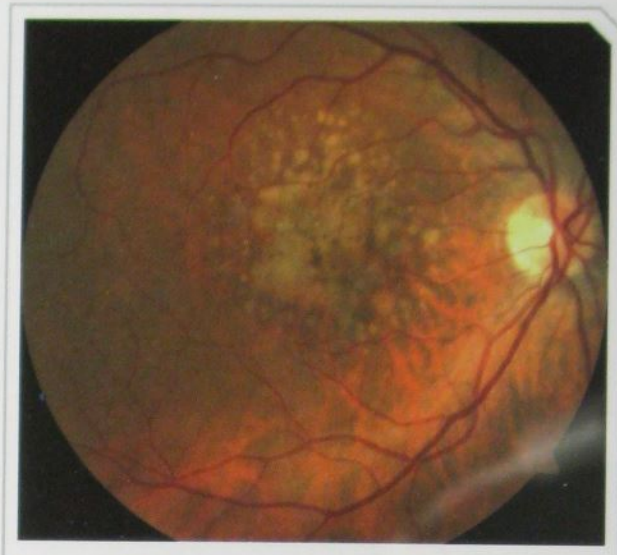


Fig. 13.2. Macular degeneration

among people aged 60 and older. Because people in this group are an increasingly larger percentage of the general population, vision loss from macular degeneration is a growing problem.

Etiology. The primary cause of AMD is unknown, but predisposing factors include older age, history of smoking, female gender, lighter skin pigmentation, high-fat diet, excessive exposure to sunlight, and genetic predisposition. Comorbidities of AMD are hypertension illness (AH), atherosclerosis, diabetes mellitus (DM), obesity.

AMD involves the choriocapillaris, Brooch's membrane and a retinal pigment epithelium failure.

Clinical Picture. There are two clinical types of AMD: dry (atrophic) and wet (neovascular or exudative).

Dry AMD is the most common type of macular degeneration, it affects 85—90 % of people with the condition. Dry AMD is characterized by atrophic changes in the macula and is bilateral. At early stages of the disease whitish or yellowish deposits, called drusen, with clear (heavy) or slightly effaced (soft) boundaries, that are located between the retinal pigment epithelium and Bruch's membrane, are detected in the macula area. As the disease develops, all of these drusen coalesce into larger conglomerates (drain drusen). The advanced stage is geographic atrophy — partial or complete depigmentation (atrophy) of the retinal pigment epithelium, with choroidal vessels that may become visible.

Loss of central vision usually takes a long time, maybe a couple of years to get to its final stage. At its worst, dry AMD causes a blank patch in the center of vision in both eyes, but it doesn't affect peripheral vision, so never leads to total blindness.

Wet AMD is less common, it affects about 10—15 % of people who have age-related macular degeneration but accounts for two-thirds of people who have significant visual loss. In the wet form of macular degeneration, after the stage of drusen formation newly created abnormal blood vessels grow through Bruch's membrane into the subpigment epithelial and subretinal spaces. These blood vessels are fragile, they leak, bleed, and can cause retinal swelling and hemorrhagic detachment of the retinal pigment epithelium. The end of this type of AMD is scar formation known as disciform macular degeneration.

Rapid vision loss, usually over days to weeks, is typical of this pathology. The first symptom is usually visual distortion, such as a central blind spot (scotoma) or curving of straight lines (metamorphopsia). Peripheral vision and color vision are generally

unaffected. Wet AMD usually affects one eye at a time; thus, symptoms of wet AMD are often unilateral.

Complaints. Patients complain of gradual decrease of central vision, reading trouble, especially in low-light conditions; sometimes patients' reports are focused on losing of individual letters, central scotoma. Also, patients may notice distortion of straight lines (metamorphopsia), changes in object sizes (macropsia or micropsia) and color perception.

Signs. In case of dry AMD — whitish or yellowish drusen in the macular region, changes in the retinal pigment epithelium, an area of chorioretinal atrophy. In case of wet AMD — localized retinal elevation, retinal edema, grey-green discoloration under the macula, exudates in or around the macula, detachment of the retinal pigment epithelium (visible as an area of retinal elevation), subretinal hemorrhage in or around the macula.

Methods of examination have to include visual acuity test, Amsler grid test, ophthalmoscopy, funduscopic examination, fluorescent angiography, electroretinography, OCT, etc.

Differential Diagnosis. Chronic recurrent central serous retinopathy, branch retinal vein occlusion, malignant melanoma.

Treatment. There is currently no cure for age-related macular degeneration, but treatment may prevent severe vision loss or slow the progression of the disease considerably.

Treatment options for *dry AMD* have to be complex, including:

— normalization of the general state of health (blood lipids, blood pressure, sugar level, etc.);

— the use of certain specific antioxidants, vitamins and minerals (vitamin C and E, beta carotene, lutein, zinc and copper) could possible prevent or delay the progression of the disease: Visiox Lutein; Lutein Complex; Ocuvite Lutein; Ocuvite Complete; Optix Forte — 1 capsule 1—2 times per day with a little sip of water. One course of treatment should be applied for 12 weeks at least. It's highly recommended to carry out 2—3 courses per year;

— the use of specific dietary supplements such as carotenoids and foodstuffs rich in omega-3 fatty acids (spinach, tomatoes, red and yellow sweet peppers, corn, carrot, salted fish: mackerel, herring, salmon, tuna, halibut, and cod);

— drug therapy that is focused on the preservation of visual analyzer function: microcirculatory vasodilator drugs (Xantinol Nicotinate 0.15; Cinnarizine; Drotaverine (No-Spa) 0.04; Pentoxifylline 0.2; Vinpocetin 0.005); nootropic and neuroprotective drugs (Aminobutyric Acid 0.025; Piracetam 0.4 or 0.8; Nootropil, Comb Drug). One course of treatment should be carried out for 1 month 2—3 times per year.

Treatment methods for wet AMD are directed at blocking the development of new blood vessels and leakage from them:

— laser photocoagulation — application of hot laser to seal and destroy abnormal blood vessels under the macula;

— photodynamic therapy — the use of non-thermal laser together with the intravenous drug Verteporfin (a photosensitive drug) to seal and stop or slow the progression of the condition;
— intravitreal injection of anti-angiogenic drugs: Bevacizumab; Ranibizumab; Pegaptanib; Aflibercept;
— submacular surgery to remove the abnormal blood vessels or blood;
— macular translocation surgery that involves detachment of the retina from its base, its slight rotation and replacement in a different position, so that the macula sits on a new, healthy base.

Prognosis. The course of the disorder is chronic and leads to a progressive loss of visual acuity. Treatment can only slow the progression of visual loss.

Complications. At any time, dry macular degeneration can progress to wet macular degeneration, which causes rapid vision loss and leads to retina perforation.

Prophylaxis. Healthy lifestyle that includes regular exercises, maintaining healthy weight, proper diet, no smoking, and management of other diseases (cardiovascular, hypertension, etc.).

Retinitis Pigmentosa

Definition. Retinitis pigmentosa (RP) is a group of hereditary progressive degenerative diseases of the retina that involve atrophy of rod cells followed by abnormalities in the adjacent retinal pigment epithelium and deterioration of cone photoreceptor cells. It is characterized by night blindness and progressive peripheral vision loss, eventually resulting in tunnel vision or total blindness.

Etiology. Retinitis pigmentosa is an inherited condition that has many different modes of inheritance. It is known to be caused by more than 100 different genetic mutations. RP involves both eyes. If it starts in one eye, the other eye usually develops the same condition in a couple of years. Most cases are familial (multiple family members affected or only one affected person), inherited in a variety of ways including dominant, recessive, sex-linked recessive or autosomal.

Clinical Picture. Retinitis pigmentosa causes slow loss of vision. It begins with decreased night vision and later progresses to loss of peripheral (side) vision creating a "tunnel vision" effect. Some people may also have difficulty identifying colors. The rate of vision change varies in different people depending on the genetic makeup of their disorder.

Complaints. Patients complain of difficulties with performing tasks at night or under low illumination, difficulties in driving in low light, at dusk or in foggy conditions. They also notice slowly progressive constriction of the visual field. Many patients with RP report seeing twinkling lights or small flashes of light (photopsia).

Signs. Loss of visual acuity, myopia, peripheral ring scotoma, nyctalopia, color vision defects. Fundus examination shows hyperpigmentation in a bone-spicule configuration in the midperipheral retina, narrowing of the retinal arterioles, a waxy yellow appearance of the optic disc, posterior subcapsular cataracts, cells in the vitreous.

Methods of Examination. Diagnosis should be based on the fundus picture, electroretinography, adaptometry, color testing and perimetry.

Differential Diagnosis. Central serous chorioretinopathy, juvenile retinoschisis, Leber congenital amaurosis.

Treatment. There is no way to reverse damage caused by retinitis pigmentosa, but treatment may help to slow disease progression:

— the use of certain antioxidants, vitamins and minerals: vitamin A (15,000 IU/day) and E (400 IU/day), lutein (Visiox Lutein; Lutein Complex; Ocuvite Lutein; Ocuvite Complete; Optix Forte — 1 capsule 1—2 times per day with a little sip of water. One course of treatment should be applied for 12 weeks at least. It's highly recommended to carry out 2—3 courses per year);
— dietary supplements of the omega-3 fatty acid DHA that include salmon, tuna, mackerel, sardines, and herring;
— metabolism-oriented drugs use: aloe 1.0 ml, 2.0 ml; Actovegin 2 ml, Solcoseryl 2.0 ml;
— complex of nucleotides — Encadum intramuscularly;
— vasodilators externally or intramuscularly: Nicotinic acid 1 ml 1 %, Pentoxifylline 2 % 5 ml or 0.2, Xantinol nicotinate 15 % 2 ml or 0.15, Vinpocetine 0.005, Cavinton 0.01;
— vitamins B_1, B_2, B_6, B_{12} externally or intramuscularly: Taufon 4 % intradermally or parabulbarly;
— physiotherapy: endonasal electrophoresis should apply Drotaverine, Papaverine;
— if cataract occurs, cataract phacoemulsification with IOL implantation;
— in case of cystoid macular edema orally or topically Acetazolamide, Dorzolamide.

There are numerous experimental trials underway ranging from electrical stimulation therapy, stem cell transplantation, subretinal microphotodiodes (prosthesis) implantation, gene therapy.

Prognosis. Retinitis pigmentosa is chronically progressive. The clinical course depends on the specific form of the disorder; severe forms lead to blindness.

Complications. Peripheral and central loss of vision will eventually occur.

Prophylaxis. Genetic counselling and testing may determine the risk of this disease occurring in a person's offspring.

4.3. Vascular Diseases

Central Retinal Artery Occlusion

Definition. Central retinal artery occlusion (CRAO) occurs when the central retinal artery is blocked with an embolus.

Etiology. CRAO is considered a stroke of the eye. The main causes of CRAO are the following: embolism (sudden obstruction of a blood vessel by a blood clot); ath-

NOTE!

Central retinal artery occlusion is an emergency condition that requires immediate treatment as it causes sudden vision loss, which can lead to irreversible blindness.

erosclerotic disease that results in progressive narrowing of arteries over time; endarteritis (chronic inflammation of the inner layer of arteries); angiospasm (a spasmodic contraction of a blood vessel with an increase in blood pressure).

CRA may become blocked by a blood clot or fat deposits that get stuck in the arteries. These blockages may occur due to hardening of the arteries in the eye. Also clots may travel from other parts of the body and block the central artery in the retina. The most common sources of clots are the carotid artery in the neck or the heart lining.

People affected by CRAO typically have high blood pressure, heart disease, or diabetes as an underlying condition. Other conditions that may increase the risk of CRAO include high cholesterol and glaucoma. The incidence is slightly higher in men and in people at the age of 60 or older.

Clinical Picture. CRAO causes sudden, painless, severe vision loss or visual field defect, usually unilaterally.

Complaints. Patients complain of sudden painless loss of vision or 'fallout' in the visual field in one eye.

Signs. Relative afferent pupillary defect, a pale retina with vessel attenuation, the arteries may even appear bloodless, milky-white ischemic edema of the posterior pole, an opaque fundus with a red fovea (a cherry-red spot) due to the vascular choroid shining through the thin macula, the disc is pale with blurred outlines (fig. 13.3).

Methods of Examination. Pupillary reflex, ophthalmoscopy, funduscopic examination, perimetry, fluorescein angiography.

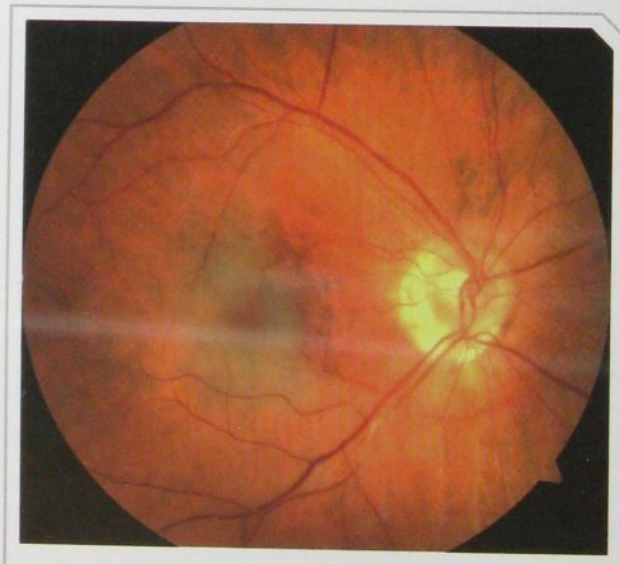


Fig. 13.3. Acute central retinal artery occlusion

Differential Diagnosis. Ocular ischemic syndrome, diabetic retinopathy, papilledema, retinal detachment, retinal vein occlusion.

Treatment. CRAO is an emergency situation that requires urgent hospitalization in an ophthalmological department. It represents an ischemic infarct of the retina. The retinal survival time is about 100 minutes: if treatment begins within this period, the patient has the highest possibility of regaining vision in the affected eye. After that time the ischemic retina will not recover, restoration of vision is unlikely.

Medical therapy is directed toward vasospasm removal, retinal vessel dilation and moving the occlusion from the central retinal artery to a branch artery, increasing retinal perfusion and oxygen delivery to hypoxic tissues, IOP lowering, and embolus dissolving:

— Nitroglycerin sublingual pill;
— inhalation of Carbon dioxide (5 % carbon dioxide and 95 % oxygen) 10 min every 2 hours per 2 days or breathing the air with a higher concentration of CO_2 (in a paper bag);
— immediate IOP lowering: instillation of β-blockers (Timolol 0.5 % 2 times per day);
— in case of hypertension it is highly recommended to decrease blood pressure:
— in case of a cholesterol embolus during the first three days eyeball massage over the closed lid (circular pressing movements) for ten seconds with five-second pauses every 10—15 min 5—6 times per day may also be performed in order to move the embolus into the distal vascular channel occlusion;
— intravenously Aminophylline 2 % 10 ml with Glucose solution 20 % 20 ml;
— intramuscularly Papaverine 2 % 2 ml + Dibasol 1 % 1 ml + Nicotinic acid 1 % 1 ml;
— Acetazolamide 0.25 g orally.

Hospital treatment and outpatient observation:

— parabulbar (p/b) injections of Atropine 0.1 % 0.5 ml; Dexamethasone 2 mg 10 days in combination with Heparin 500 p/b + Emoxipin 1 % 0.5 ml; hereafter corticosteroids: Triamcinolone 20 mg p/b once a week or Betamethasone 0.5 ml p/b every 7—10 days;
— parabulbar injections of Papaverine 2 % 0.2 ml; Novocaine 2 % 0.2 ml once a day for 10 days (injected in 40 min after Dexamethasone intake);
— intravenously Pentoxifylline 5 ml combined with sodium chloride 0.9 % 200 ml once a day for 4—7 days; afterwards — vasodilators ext. Nicotinic acid 0.02—0.05 g 2—3 times per day for 1 month; Pentoxifylline 0.1 g 3 times per day for 1—2 months;
— in case of normal blood viscosity intravenously Acetazolamide 0.25 g once per 2 days for 2 weeks (with potassium drugs);
— if hematocrit level corresponds to 30—35 % — intravenously Rheopolyglucin 200—400 ml or Neohaemodes 200 ml once per 2 days up to 3—5 infusions;
— drugs that restore the integrity and function of the vascular endothelium: intravenously Sulodexide 2 times per day for 30 days; Trimetazidine 20 mg 3 times per day for 2 months;
— antioxidants; Emoxipin 1 % p/b once a day for 10—15 days; Cytoflavin; Mexidol intravenously 200—300 mg 2—4 days, then 100 mg 3 times per day for 10—14 days; Vitamin E (Tocopherol) intravenously 200 mg once or twice during 1—1.5 months;
— B-multivitamin group intravenously or 1 pill 3 times per day for 1 month.

Prognosis. The prognosis for central retinal visual acuity is poor with only about one-third of patients recovering useful vision. The longest delay in getting treatment that has been associated with significant visual recovery was approximately 72 hours.

The presence of a retinal embolus is associated with a 56 % mortality rate over 9 years compared to 27 % in patients without arterial emboli. Life expectancy of patients with CRAO is 5.5 years compared to 15.4 years for an age-matched population without CRAO.

Complications: retina degeneration, optic nerve atrophy, further emboli to the brain in a cerebrovascular accident or to the same or contralateral eye, resulting in visual loss.

Prophylaxis. Individuals affected by underlying conditions such as high blood pressure, heart disease, diabetes, glaucoma, and elevated cholesterol should treat their conditions appropriately to minimize the possibility of retinal artery occlusion.

Central Retinal Vein Occlusion

Definition. Central retinal vein occlusion (CRVO) is a blockage of the central retinal vein by a blood clot or a thrombus. It is a common cause of vision loss in older individuals, and the second most common retinal vascular disease after diabetic retinopathy.

Etiology. CRVO development could be caused primarily by hypertensive diseases (in the majority of cases), atherosclerosis, diabetes, systemic vasculitis, any disease accompanied by increased blood viscosity (macroglobulinemia, multiple myeloma, polycythemia), different forms of thrombophilia. Development of this condition should also be preceded by local factors: optic disc swelling or drusen, high IOP; generally orbit blood vessel compression takes place (orbital tumors, thyroid ophthalmopathy).

Clinical Picture. Central retinal vein occlusion is divided into two types: non-ischemic or ischemic, each having its clinical presentation, prognosis, complications and management options. An intermediate form also exists, but 80 percent of these intermediate cases progress to the ischemic variety over time.

Non-ischemic CRVO is the most common type, accounting for about 75 %. Presentation is with sudden, unilateral blurred vision. Funduscopy shows tortuosity and dilatation of all branches of the central retinal vein, dot/blot and flame-shaped hemorrhages, throughout all four quadrants and most numerous in the periphery, and optic disc and macular edema. Some cotton-wool patches, particularly in hypertensive patients, may be present.

Ischemic CRVO is the severe form of the disease that is characterized by rapid onset of venous obstruction resulting in decreased retinal perfusion, capillary closure and retinal hypoxia. Patients with severe central retinal vein occlusion typically have sudden unilateral painless loss of vision. It presents with a marked afferent pupillary defect, severe tortuosity, and engorgement of all branches of the central retinal vein,

extensive deep blot and flame-shaped hemorrhages involving the peripheral retina and posterior pole, severe disc edema and hyperemia. This may lead to profound vascular leakage and intraocular pressure increase.

Complaints. Patients complain of sudden painless decrease or loss of vision in one eye, which develops in a few minutes or hours, veil in front of the eyes. The disease should be preceded by periodic blurring of vision. Patients also may notice distorted objects, floaters and dark spots in front of the eyes.

Signs. In the fundus, the optic disc borders are effaced or undetectable, the veins are dilated, curved. In severe cases, retinal edema may occur in the optic disc and on the major vessels, which extends to the macular area. Soft exudates should resemble clumps of wool. Numerous shaped hemorrhages usually occur (a 'squashed tomato' clinical picture) (fig. 13.4, 13.5). In the early stage of thrombosis or pre-thrombosis hemorrhages could be small and bar-shaped. In a case of the non-ischemic type of thrombosis, hemorrhages are mainly located on the periphery, in severe cases — at the posterior pole of the eye. 'Soft exudates' seldom appear. While thrombosis progresses similarly to the ischemic type, hemorrhages are localized basically at the posterior pole, having been an edema of retina shift. This type of thrombosis is characterized by numerous 'soft exudates'.

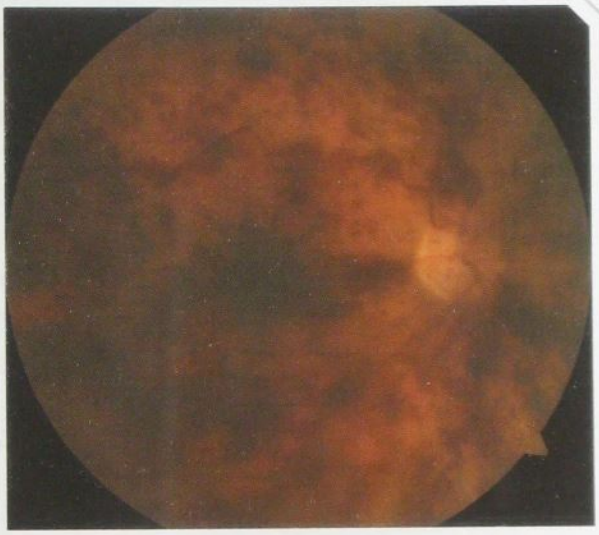


Fig. 13.4. Central retinal vein thrombosis

Methods of Examination. Funduscopy, color fundus photography, fluorescein angiography, optical coherence tomography.

Differential Diagnosis. Branch retinal vein occlusion, ocular ischemic syndrome.

Treatment. The earlier CRVO is diagnosed and treated, the lower the risk of permanent structural damage to the eye. It may require immediate treatment that includes thrombus dissolving, IOP lowering, treatment

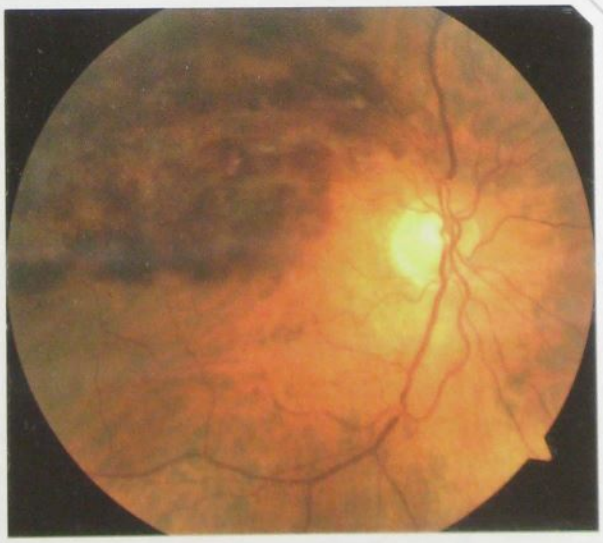


Fig. 13.5. Thrombosis of the central retinal vein branch

of associated systemic diseases and complications of CRVO that may include macular edema and neovascularization:

- intravenously 10 ml of 2 % Aminophylline solution diluted in 20 ml of 20 % glucose solution;
- intramuscularly 10 ml of 25 % Magnesium sulfate solution;
- leeches on the temple area, hot foot bath;
- decreasing blood pressure;
- parabulbar — Dexamethasone 2 mg with Heparin 750 U and Rheopolyglucin 30,000—40,000 0.2 ml once a day for 10—12 days; or Dexamethasone 2 mg with 0.5 ml 1 % Emoxipin for 10—15 days; afterwards corticosteroids — parabulbar Kenalog once a day every 7—14 days;
- at the hematocrit level of 30—35 % — Rheopolyglucin intravenously 200—400 ml or Neohaemodes 200 ml every 2 days, Furosemide 1—2 ml and Dexamethasone 4—8 mg, totally 3—5 infusions;
- intravenously Pentoxifylline 5 ml should be dissolved in 200 ml of 0.9 % Sodium chloride solution — 10 infusions; afterwards Pentoxifylline internally 100 mg 3 times per day for 1—2 months;
- reduction of IOP: Timolol 0.5 % 2 times per day, Brinzolamide 2—3 times per day; and diuretics: ext. Acetazolamide 1 tablet (0.25 g) once a day in the morning before breakfast for 3 days. When a patient should have a 3-days' break off drugs, the given therapy is repeated in 3 days. The doctor could abide different ways of treatment regime: 1 tablet during any other day. Such treatment should be combined with potassium drugs;
- hemostatic drugs (2 ml of intravenous Etamsylate 12.5 % solution) during the first 3—5 days, and after 5—7 days of the disease thrombolytic drugs (enzymes) could be administered in the form of intravitreal injection. Later, the patient should be transferred to enzyme treatment: Wobenzym 8—10 pills 3 times per day for 2 weeks, thereafter 3 pills for 6—8 weeks; Phlogenzym 2 pills 3 times per day 30—60 min before meals; drinking enough water for 2—3 months;
- endothelium protectors: Sulodexide for 30 days, Troxevasin 1 capsule 2 times per day for 1—2 months;
- antiplatelet drugs for the period of 3—6 months: Acetylsalicylic acid ext. 50—125 mg 1 pill per day nocte; thrombotic ACC 50 mg 2 times per day or 100 mg once a day;
- antioxidants: Emoxipin 1 % p/b 0.5 ml once a day for 10—15 days, Cytoflavin intravenously 1 pill once a day No. 5; Mildronate p/b 0.5 ml once a day, Mexidol orally 200—300 mg 2—4 days — the first course, the next course: 100 mg 3 times per day for 10—14 days; Vitamin E (tocopherol) 200 mg 1—2 times during 1—1.5 months; Trimetazidine orally 20 mg 3 times per day for 2 months.

For macular edema secondary to CRVO intravitreal injections of anti-vascular endothelial growth factor (anti-VEGF) drugs — Ranibizumab, Bevacizumab, Afliber-

cept that blocks the growth of new blood vessels and intravitreal injections of triamcinolone are used. This treatment is still being studied.

In cases of neovascular complications of CRVO panretinal photocoagulation is used to block the growth of new vessels that can cause glaucoma.

In case of vitreous hemorrhages vitrectomy is conducted to remove the posterior hyaloid.

Prognosis. In case of non-ischemic CRVO the prognosis is reasonably good with return of vision to normal or near normal in about 50 % cases. The prognosis of ischemic CRVO is extremely poor due to macular ischemia and a high risk of neovascular glaucoma or retinal neovascularization. About 10 % of patients can develop CRVO or other type of vein occlusions within either the same eye or the contralateral eye within two years.

Complications. Potential complications include macular edema, secondary neovascular glaucoma due to retinal neovascularization, vitreous hemorrhage, and optic nerve atrophy.

Prophylaxis. Central retinal vein occlusion is a sign of a general blood vessel (vascular) disease. Measures used to control associated systemic diseases (hypertension, diabetes, thrombophilia, etc.) may reduce the risk of retinal vein occlusion.

4.4. Retinal Detachment

Definition. Retinal detachment is separation of the inner neurosensory retina from the underlying retinal pigment epithelium.

NOTE!

Retinal detachment is a critical emergency. Since the retina cannot work properly under this condition, a patient can lose vision if the detached retina is not repaired promptly.

Etiology. Retinal detachment is caused by separation of the rods and cones layer from the retinal pigment epithelium due to accumulation of fluid between them. As a result, there develop nourishment disorders in the outer retina layers leading to fast loss of vision. The possibility of retinal detachment is usually caused by its morphological structure, as well as degeneration of the retina structure, traction influence of the vitreous, injuries and numerous ophthalmic diseases such as retinitis, uveitis, choroidal tumors, cysticercoids, diabetic retinopathy and kidney diseases, retinopathy of prematurity, vascular lesions, etc.

Clinical Picture. Retinal detachment occurs by three basic mechanisms and thus is classified into the following three main types:

Rhegmatogenous retinal detachment occurs due to a tear or break in the retina that allows fluid to get under the retina and separate it from the retinal pigment epithelium (RPE), the pigmented cell layer that nourishes the retina. This type of retinal detachment is the most common.

The tractional type occurs when scar tissue on the retina's surface contracts and causes the retina to pull away from the back of the eye. This is a less common type of detachment that typically affects people with diabetes.

The exudative type of detachment is caused by retinal diseases, including inflammatory disorders and injury to the eye. In this type, fluid leaks into the area underneath the retina, but there are no tears or breaks in the retina.

The classical clinical presentation of impending retinal detachment is sensations of flashing lights accompanied by black floating dots and streaks followed by appearance of a blur in the field of vision encroaching from the periphery, described as a "shadow" or "curtain". When the detachment crosses the macular region, field loss progresses to sudden loss of central visual acuity. The time interval is usually a few days or weeks but it may be longer when detachment is below.

The flashing lights are due to traction of the vitreous on the peripheral retina because the retina has no pain fibres and responds to abnormal stimulation by giving a sensation of light. The black spots are due to hemorrhage into the vitreous from the retinal tear. In case of a small retinal hole or holes the onset may be insidious with gradual loss of peripheral visual field; a very gradual onset of lower retinal detachments may even be discovered accidentally when unsuspected.

Complaints. Patients with retinal detachment complain of sudden loss of visual activity; appearance of many floaters (small bits of debris in the field of vision that look like spots, hairs or strings and are floating before the eye); sudden flashes of light (photopsia) in the affected eye; appearance of a "veil" moving across the vision field; bending objects (metamorphopsia); loss of vision in the peripheral visual field occurs on the side opposite to detachment location.

NOTE!

The detached area of the retina is in the opposite direction to the field loss, e.g. loss of the lower field of vision indicates upper detachment.

Signs. The loss of the red reflex. The detached area of the retina appears greyer than the surrounding fundus and bulged forward as a bubble, the vessels on its surface are darker and more tortuous (fig. 13.6). The detached retina often mobile.

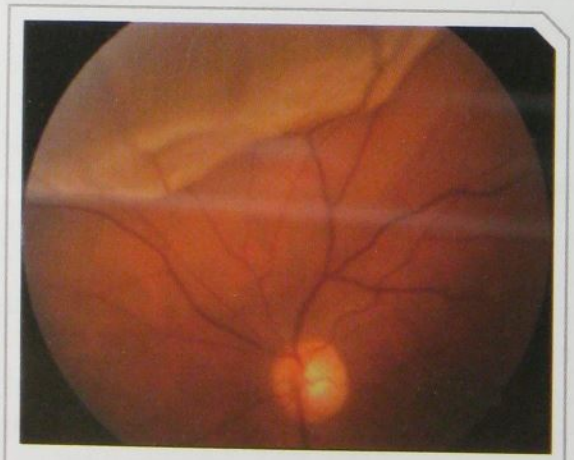


Fig. 13.6. Retinal detachment

The retinal tear appears as a red area where the choroid contrasts sharply with the greyish detached retina.

Methods of Examination. Ophthalmoscopy, perimetry, biomicroscopy, ultrasonography, electrophysiological methods of examination, OCT.

Differential Diagnosis. Retinoschisis, choroidal mass, optic neuritis, age-related macular degeneration.

Treatment. Retinal detachment requires care right away. Without treatment, vision loss can progress from minor to severe or even to blindness within a few hours or days. Surgery is the only treatment of retinal detachment. The goals of surgery are to reattach the retina and to prevent or reverse vision loss. It is usually successful and, in many cases, restores good vision.

For treatment purposes the following methods can be applied:

1. In case of a retinal hole or tear *laser photocoagulation* or *cryopexy* (intense cold) is used to seal the area around the hole and reattach the retina.

2. *Scleral buckling* involves the use of fine bands of silicone rubber or sponge that are permanently stitched onto the sclera in the area where the retina has detached. The bands act like a buckle and press the sclera towards the middle of the eye, so the torn retina can lie against the wall of the eye. Scleral buckling is combined with retinopexy, typically cryotherapy. This is the oldest method of repair that still has excellent results.

3. *Vitrectomy* is the most commonly performed operation. It works by removing the vitreous from the inside of the eye and replacing it with either a gas or silicone bubble. This holds the retina in its position from the inside.

4. *Pneumatic retinopexy* involves intravitreal injection of a bubble of gas near the area of retinal detachment to press the retina back into place. Laser or freezing treatment is often used to create scar tissue that keeps the retina in the correct place. The bubble is slowly absorbed into the eye over the following weeks.

Prognosis. As the retina is a neuro-sensitive tissue, visual prognosis can be difficult to predict. It depends on the severity of the condition and how quickly a patient received medical care. Generally, retinal detachment without macular involvement tends to have a better final visual prognosis. In some cases, more than one treatment is necessary. Some blurring of vision may remain in people who have detachments that involve the macula (central vision). This is why treatment is an emergency if the macula is still attached.

Complications. Loss of vision to hand motion or light perception is a frequent complication of retinal detachments that involve the macula.

Prophylaxis. In general, there is no way to prevent retinal detachment. However, there are steps to avoid retinal detachment that results from an injury by wearing protective eyewear when playing sports or using tools. In case of diabetes — blood sugar control. Early diagnostics of vitreous detachment and retinal tears, which can be treated with laser or cryotherapy, reduces the risk of retinal detachment.

4.5. Inflammatory Diseases

Retinitis

Definition. Retinitis is an inflammation of the retina that can be caused by toxic-allergic processes, immune aggression or infection spread into the retina through the central retinal artery and its branches.

Etiology. Infectious diseases (tuberculosis, syphilis, toxoplasmosis, viral (cytomegalovirus, herpes zoster) and purulent infection), infectious and allergic conditions (rheumatism and other types of collagen diseases), allergic processes, intoxication, irradiation, tropical infections spread in countries with hot climate. We distinguish primary infectious retinitis, radiant energy retinitis, and secondary retinitis that develops in case of uveal tract inflammation.

Clinical Picture. It may be present as pure retinitis (only retinal inflammation), chorioretinitis (inflammation of the retina and choroid), neuroretinitis (inflammation of the optic disc and surrounding retina), or retinal vasculitis (inflammation of the retinal vessels).

Complaints. Patients often complain of photopsia and metamorphopsia. Generally, you should pay attention to cases of micro- and macropsia. Lesions of the peripheral retina are accompanied by concentric or sector-oriented changes near the boundaries of the field of view. Violation of color perception is typical for central localization of the process.

Signs. In retinitis, the fundus is of white, yellow or grey color. Focal opacities located along the nerve fibres in the macular area form a shape of a star. Sometimes small foci coalesce into a large area of turbidity. In the majority of cases, numerous retinal hemorrhages of various shapes and sizes can appear. The retinal vessels have a tendency to change their form, as well as to narrow or expand; occasionally there are aneurysms. The vitreous body could be clouded. On the fundus wide areas of necrosis, scarring, clusters of pigment and atrophy can be seen (fig. 13.7).

Methods of Examination. Diagnosis of retinitis should be carried out by ophthalmoscopy, fluorescent angiography, and perimetry.

Treatment. Treatment of retinal inflammatory diseases may vary depending on both its severity and underlying causes. First of all, treatment must be etiologically focused.

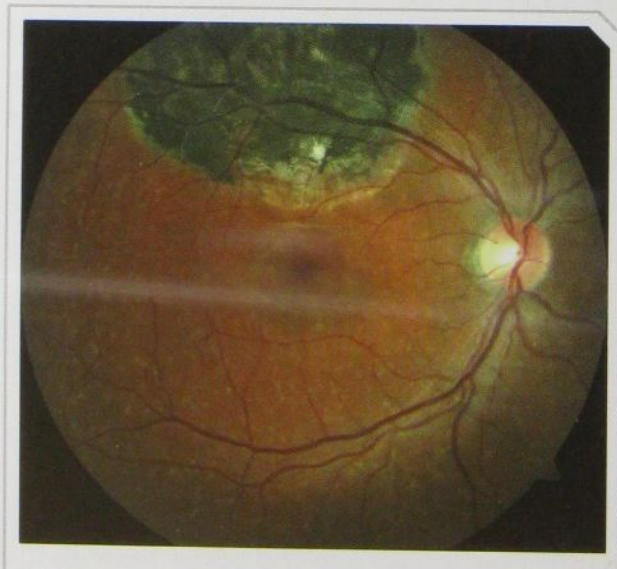


Fig. 13.7. Retinitis

Anti-inflammatory and immunosuppressive therapy: Fluoroquinolones, Penicillin, corticosteroids, salicylates, Indomethacin, Ibuprofen, etc. Retrobulbar administration of 0.4 % 0.5 ml Dexamethasone solution (10 injections per one course) should be provided in the first place. Electrophoresis with emulsion of 0.1 % hydrocortisone.

Orally drugs are administered for vascular wall protection: Dicynone, Rutin, Ascorutine, Ascorbic acid.

Prognosis. Untreated retinitis will progress to blindness due to retinal necrosis, optic nerve in-

volvement, or retinal detachment. Macular lesion leads to visual acuity decrease and central scotoma appearance; on the other hand, visual acuity of peripheral localization is slightly reduced.

Complications. As the process spreads into the choroids, retinochoroiditis could occur; sometimes optic nerve swelling and neuritis, secondary foci of retinal degeneration are formed.

Prophylaxis: diagnosis and primary treatment of infectious and viral diseases, allergic conditions, prevention of exposure to toxic substances, radiant energy, etc.

4.6. Tumors

Retinoblastoma

Definition. Retinoblastoma is the most common malignant ocular tumor in childhood that originates from immature retinal cells. The onset generally occurs between the third month of pregnancy and 4 years of age. It is the most common malignant ocular tumor in children occurring in approximately one in 20,000 births. In 30 % cases, it is bilateral.

Etiology. Retinoblastoma is caused by the so-called retinoblastoma gene, which is a mutation in the long arm of chromosome 13. It may be sporadic or hereditary. A somatic mutation is detected in about 90 % of patients. In the other patients (10 %), it is inherited as an autosomal-dominant trait. The sporadic form of retinoblastoma can be caused by de novo gene RBI mutations located in the retina cells. Chromosomal analysis of mutation of this gene can find the hereditary form of the disease.

Clinical Picture. Retinoblastoma develops in any part of the retina. At the beginning of the disease, the tumor appears as a violation of the fundus reflex. Medical examination shows a greyish, muddy center with indistinct contours. Further clinical picture depends on the growth of retinal blastoma — of endophytic, exophytic or mixed nature. Fast-growing endophytic retinoblastoma occurs in the inner retinal layers, being characterized by vitreous growth. Exophytic tumor develops in the outer retinal layers and distributes under the retina causing its detachment. The mixed nature of retinoblastoma is characterized by symptoms of both endophytic and exophytic tumor growth.

Retinoblastoma has several stages according to its extension. They are:

— intraocular: the tumor is inside the eyeball;
— extraocular: it is extended beyond the eye, usually along the optic nerve; it may be confined to the tissues around the eye, or it may spread to other parts of the body;
— trilateral: in patients with bilateral retinoblastoma a related tumor develops in the pineal gland at the base of the brain;
— recurrent: the cancer has come back or progressed after it has been treated; it may recur in the eye or elsewhere in the body.

Complaints. Over 80 % of retinoblastomas are first detected by a family member or a friend (compared to less than 20 % detected by a doctor) as a white pupillary reflex identified on photos, misaligned eyes. A child doesn't fix gaze on objects and doesn't control eye movements. Sometimes the iris color changes.

Signs. There are several signs which can indicate retinoblastoma but the most typical are:

— leukocoria (also known as cat's eye reflex) — white pupillary reflex when light is shone into the eyes;
— strabismus — when one eye does not seem to be looking at the intended target, either crossed inwards or deviated outwards;
— iris heterochromia;
— nystagmus.

Retinoblastoma can cause secondary changes in the eye, including glaucoma, retinal detachment, buphthalmos, hyphema, hemophthalmus, and inflammation secondary to tumor necrosis.

Methods of Examination. Ophthalmoscopy, ultrasonography, fluorescein angiography, photographic imaging, optical coherence tomography, computed tomography, and magnetic resonance imaging. Genetic testing.

Differential Diagnosis. Cataract, primary strabismus, retinopathy of prematurity, retinal detachment, Coats' disease.

Treatment is usually individualized patient-specific and is based on the extent of the disease within the eye, whether it is in one or both eyes, and whether it has spread beyond the eye. Treatment options consider both cure of the cancer and preservation of sight or the affected eye, and include the following:

— treatment of a small tumor that is completely inside the eye involves laser treatment (photocoagulation), cryotherapy or thermotherapy;
— larger tumors are treated with one or a combination of internal or external radiation therapy methods delivered by plaques of radioactive ruthenium or iodine (brachytherapy), chemotherapy or targeted chemotherapy in order to prevent tumor metastasis;
— if the tumor is large, it requires immediate enucleation of the eye.

Prognosis. Early diagnosis and treatment can lead to cure in over 90 % cases. There is a 6 % risk of a second malignancy. Untreated, the tumors invade adjacent structures and then metastasize causing death within two years.

Complications. The tumor can spread to the brain, spinal cord, bones or lymph nodes. Patients who have been treated with radiation are at higher risk of secondary tumors.

Prophylaxis. Family/genetic counselling and screening for prevention. Following the diagnosis, the fellow eye should be examined with the pupil dilated every 3 months for 5 years. After that, follow-up examinations can be performed at longer intervals.

Review:

1. Key points

Diseases of the retina by origin may be congenital (developmental or hereditary) or acquired, and by clinical forms — degenerative, vascular, inflammatory, retinal detachment, and tumors.

The main symptoms of retinal diseases are visual impairment or total loss of vision, metamorphopsia, macropsia or micropsia, scotomas in the central or peripheral vision fields, spots or lines that "float" in the vision field and may look like insects, flashes of light reminding "lightning streaks". The examination consists of ophthalmoscopy, ophthalmochromoscopy, ultrasonography, electroretinography, fluorescein angiography, OCT, HRT, perimetry.

Degenerative disorders are age-related macular degeneration that can be of dry or wet form, retinitis pigmentosa. Signs of dry AMD: whitish or yellowish drusen in the macular region; wet AMD: retinal edema, discoloration under the macula, exudates in or around the macula, detachment of the retinal pigment epithelium. Typical signs of retinitis pigmentosa: retinal hyperpigmentation, narrowing of the retinal arterioles, a waxy yellow appearance of the optic disc. Decrease of vision and visual disorders are gradual. There is no cure for retinal degenerative diseases, but treatment may prevent severe vision loss and help to slow disease progression.

Vascular diseases are central retinal artery or central retinal vein occlusion that are emergency situations as untreated they can cause irreversible blindness or retinal impairment. Patients complain of sudden painless loss of vision or "fallout" in the visual field in one eye. An obvious fundoscopic sign of CRAO is a cherry-red-spot in the macula region, of CRVO — soft exudates and a "squashed tomato" clinical picture on the fundus. Medical therapy is directed toward vasospasm removal, retinal vessel dilation, and embolus dissolving.

Retinal detachment is a critical emergency that can lead to sudden loss of vision. The classical clinical presentation of impending retinal detachment is sensations of flashing lights accompanied by appearance of floaters and photopsia. Retinal detachment requires immediate treatment. For treatment purposes the following methods can be applied: laser photocoagulation or cryopexy, scleral buckling, vitrectomy, and retinal retinopexy.

Inflammatory diseases are retinitis that can be caused by toxic-allergic processes, immune aggression, and a couple of other infections. The fundus in case of retinitis is of white, yellow or grey color. Treatment of retinal inflammatory diseases varies depending on both its severity and underlying causes.

Tumors: retinoblastoma is a malignant ocular tumor in childhood that originates from immature retinal cells. The most typical signs are leukocoria (white pupillary reflex) and strabismus. Early diagnosis and treatment can lead to cure in over 90 % cases. Left untreated — lethal. Treatment depends on the size and extension of the tumor: photocoagulation, cryotherapy, thermotherapy, internal or external radiation therapy, chemotherapy, enucleation of the eye.

2. Diagrams

DISEASES of the RETINA
Congenital
Coloboma
Leber's congenital amaurosis
Coats' disease
Congenital anomalies of the retinal vessels
Degenerative
Age-related macular degeneration
Retinitis pigmentosa (pigmentary degeneration
Central serous retinopathy
Peripheral retinal degeneration
Vascular
Central retinal artery occlusion
Branch retinal artery occlusion
Central retinal vein occlusion
Branch retinal vein occlusion
Inflammatory
Retinitis
Retinal vasculitis
Detachment
Rhegmatogenous
Tractional
Exudative
Tumors
Retinoblastoma

3. Review Questions

A. Control Questions

1. What are the methods of retina investigation?
2. What are the clinical signs of central retinal artery occlusion?
3. First aid in case of central retinal artery occlusion.
4. What are the clinical signs of central retinal vein occlusion?
5. Which somatic diseases can cause central retinal vein occlusion?
6. Specify treatment of retinal detachment.
7. Which forms of AMD can you classify?
8. Which forms of dystrophic retinal diseases do you know?
9. What are the main signs of retinoblastoma?
10. What methods of retinoblastoma treatment do you know?

B. Tests

1. Methods of retinal disease diagnostics are:
A. Ophthalmoscopy
B. Ophthalmochromoscopy
C. Perimetry
D. Optic coherence tomography
E. All the answers are correct

2. Signs of central retinal artery occlusion:
A. Visual acuity is abruptly reduced
B. The optic disc is pale, its borders are effaced
C. The arteries are sharply narrowed
D. 'Cherry spot' symptom
E. All the answers are correct

3. Typical features of central retinal vein occlusion:
A. Visual acuity is reduced
B. Presence of optic disc edema, hemorrhage
C. The veins are abruptly expanded and convoluted
D. Prominent swelling of the retina, multiple hemorrhages
E. All the answers are correct

4. Which disorders can cause sudden blindness?
A. Cataract
B. Retinal detachment
C. Thrombosis of the central retinal vein
D. Central retinal artery occlusion
E. All the answers are correct

5. Symptoms of pigment retinitis:
A. A decrease in visual acuity
B. Concentric narrowing of the visual field
C. The optic disc is yellowish-pale
D. The arteries and veins are narrowed
E. All the answers are correct

6. The main symptoms of age-related macular degeneration are:
A. A decrease in central vision
B. Metamorphopsia
C. Macro- or micropsia
D. Pathology of color perception
E. All the answers are correct

7. **Retinal detachment should be differentiated from:**
   A. Age-related macular degeneration
   B. Retinoschisis
   C. Retinoblastoma
   D. Choroidal mass
   E. All the answers are correct

8. **Types of retinal detachment are:**
   A. Tractional
   B. Inflammatory
   C. Exudative
   D. Rhegmatogenous
   E. All the answers are correct

9. **Etiology of retinitis:**
   A. Infectious diseases
   B. Collagenoses
   C. Intoxication
   D. Allergic processes
   E. All the answers are correct

10. **In a case of retinoblastoma, life prognosis depends on:**
    A. Tumor localization
    B. Tumor size
    C. Growth rate
    D. Metastases activity
    E. All the answers are correct

C. Clinical Cases

Case 1

A patient with rheumatic heart disease suddenly went blind in his right eye. Visual acuity is 0 (zero). There are no signs of inflammation. The optical media are transparent. Fundus examination showed that the optic disc is pale, swollen, in the macula region — milky white swelling with a red spot in the center. The retinal arteries are narrowed abruptly. What is the diagnosis?

A. Cataract
B. Retinal detachment
C. Occlusion of the central retinal vein
D. Central retinal artery occlusion
E. All the answers are correct

Case 2

What drugs should you administer to the patient in case of emergency care as in the clinical case 1?

A. Antispasmodics
B. Antibiotics
C. Corticosteroids
D. Diuretics
E. Laser treatment

Case 3

The patient complains of severe sudden loss of vision. The patient had hypertension. Visual acuity was 0.05, cannot not be corrected. There are no signs of inflamma-

tion. The optical media are transparent. Fundus examination: the optic disc is hyperemic, edematous, its boundaries are effaced. The retinal vein is dilated, tortuous, the artery is narrowed. In the fundus centre, there are multiple hemorrhages in the form of flames. What is the disease?

A. CRA occlusion
B. CRV thrombosis
C. Ischemic neuropathy
D. Hypertensive retinopathy

Case 4

Enumerate possible complications in the clinical case 3:

A. Retinal degeneration
B. Secondary glaucoma
C. Retinal embolism
D. Optic nerve atrophy

Case 5

A patient with a high myopia after considerable physical exertion complains of a sudden decrease in vision in the left eye preceded by the appearance of flashing lights before the eye. Medical examination showed absence of ocular inflammation. The cornea and lens are transparent. On the fundus: loss of the red reflex, the retina appears grey and bulged forward as a bubble. In the back side of the grey bubble there is a red spot of a horseshoe shape. What is the disease?

A. Intraocular tumor
B. Retinal detachment
C. Hemophthalmus
D. CRA occlusion

Case 6

A 14-year-old patient complains of poor eyesight under low illumination, which has developed recently. Since his childhood, visual acuity of both eyes has been 1.0. The anterior part of the eye hasn't changed. The optical media are transparent. In both eyes the optic discs are pale yellow. The retinal artery and veins are partially narrowed. There are black foci of degeneration on the fundus periphery. The field of vision in both eyes is concentrically narrowed. Adaptometry has shown a decrease in dark adaptation. Specify the diagnosis and treatment options.

A. Retinitis pigmentosa
B. Age-related macular degeneration
C. Acute retinitis
D. All the answers are correct

Case 7

A patient, 75 years old, complains of gradual acute decrease in visual acuity in both eyes. Visual acuity of both eyes is 0.7, which can't be corrected. In the macular region — numerous yellow-grey foci. What diagnosis should be supposed?

A. Acute retinitis
B. dry form of AMD
C. Detachment of the retina
D. Central retinal vein occlusion

Case 8

A patient, 56 years old, complains of decreasing visual acuity after hypertonic crisis. Visual acuity was 0.3, which can't be corrected. Eye fundus status: optic disc borders are undetectable, veins are dilated, curveted, hemorrhages of different form and size, a retinal edema has been found. What diagnosis should be supposed?

A. Retinal embolism
B. Hypertensive retinopathy
C. Central retinal vein outflow occlusion due to thrombosis
D. All the answers are correct

Case 9

Eye fundus examination with a dilated pupil, focused on the periphery and on the surface, showed a large white focus with boundary pigmentation. What diagnosis should be supposed?

A. Acute retinitis
B. Retinal degeneration
C. Chronic inflammatory focus
D. All the answers are correct

Case 10

A patient complains of central scotoma, decreasing visual activity, photopsia, metamorphopsia. Find out the localization of the pathological process of the retina.

A. Fovea
B. Peripheral retina
C. Optic nerve disc
D. All the answers are correct

CHAPTER 14

Diseases of the Optic Nerve

OBJECTIVES

To know:

— classification of optic nerve diseases;
— symptoms and signs of optic nerve disorders;
— diagnostic techniques for optic nerve pathology;
— pathogenesis and causes of optic nerve diseases;
— treatment options for optic nerve diseases.

Plan:

1. Classification of Optic Nerve Diseases

— According to origin — congenital (hereditary or developmental) and acquired.
— According to the character of the pathological process — acute and chronic.
— According to the dynamics of the pathological process — fixed and progressive.
— According to the pathogenesis — inflammatory, degenerative, compressive, vascular, nutritional and toxic, tumors, traumas (direct or indirect).

1.1. Congenital anomalies — optic atrophy, Leber's hereditary optic neuropathy, optic nerve hypoplasia, optic disc coloboma, myelinated nerve fibres, morning glory syndrome, optic disc drusen, pseudopapillitis.
1.2. Inflammatory diseases — optic neuritis: papillitis, neuroretinitis, retrobulbar neuritis, chiasmal optic neuritis
1.3. Degenerative — optic atrophy.
1.4. Compressive optic neuropathies — papilledema.
1.5. Vascular disorders — ischemic optic neuropathy (anterior and posterior; arteritic and non-arteritic).
1.6. Nutritional and toxic optic neuropathies — tobacco-alcohol amblyopia, methyl alcohol amblyopia, vitamin deficiency, etc.
1.7. Tumors — glioma, meningioma.
1.8. Trauma — direct or indirect.

2. Symptoms of Optic Nerve Diseases

— Mild to severe central vision loss that can be acute and gradual.
— Blurred vision.
— Peripheral vision scotomas.
— Reduced color vision.
— The presence of a relative afferent papillary defect.
— Changes in the optic nerve disc.

3. Diagnostic Techniques

- Visual acuity.
- Ophthalmoscopy.
- Pupillary light reflex.
- Color vision testing.
- Optical coherence tomography.
- Heidelberg retinal tomography.
- Visual field/perimetry.
- Fluorescein angiography.
- Visual evoked potential.
- Color Doppler flow imaging.
- CT and MRI scans.

4. Optic Nerve Diseases

4.1. Normal Ophthalmoscopic Picture of the Optic Nerve Head

The optic nerve head (or optic disk, or optic papilla) is the location where ganglion cell axons exit the eye to form the optic nerve. There are no light sensitive rods or cones to respond to a light stimulus at this point thus it is also known as the blind spot.

Examination of the optic disc can show the health of the optic nerve. In particular, it evaluates the color, sharpness of the edges, and the cupping size (as a ratio of the cup to disc size).

The optic nerve head is usually round or slightly oval in shape. Its size is 1.5 to 1.7 mm and it is larger in the vertical direction. It is orange-pink to light red in color.

The borders of the optic disc should be clear, distinct and well defined, particularly temporally.

The optic nerve head has a central white depression called the physiological cup. The tissue between the cup and the disc margin is called the neural rim or neuroretinal rim. In normal individuals, the rim has a relatively uniform width and a color that ranges

from orange to pink. The normal cup to disc ratio (the diameter of the cup divided by the diameter of the whole nerve head or disc) is about 1/3 or 0.3.

The central retinal artery and central retinal vein enter the eye within the optic disc slightly nasally. Retinal arteries are viewed as lighter, thinner blood vessels whereas the retinal veins appear thicker in caliber and are darker in color. Their lumens show a relationship of 2:3 (fig. 14.1). In 70 % of normal eyes vein pulsation is seen, arterial pulsation is pathological.

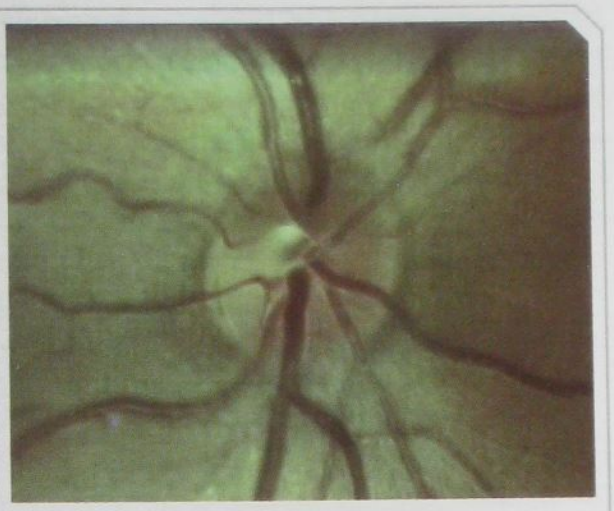


Fig. 14.1. Normal optic nerve head

4.2. Inflammatory Diseases

Optic Neuritis

Optic neuritis is an inflammation of the optic nerve causing sudden visual loss in part of the visual field. Inflammation of the optic nerve may be caused by a wide variety of diseases. They include demyelinating diseases (loss of the optic nerve's protective myelin sheath); viral infections, including infections that have spread from a nearby structure; current infections or inflammation located elsewhere in the body; decreased blood supply to the nerve; tumors or cancers; nutritional and metabolic disorders; and poisoning or exposure to toxic chemicals. Direct injury to the optic nerve can also cause neuritis.

Optic neuritis affects individuals between 20 and 40 years of age. More than 75 % of those affected are women, and 85 % are Caucasian.

The condition is unilateral rather than bilateral. It may affect some part of the nerve and disc within the eyeball (papillitis), the optic disc and surrounding temporal retina (neuroretinitis) or a portion of the optic nerve behind the eyeball (retrobulbar optic neuritis).

Papillitis

Definition. Papillitis is an acute inflammation of the optic nerve head (papilla or optic disc) associated with rapid loss of vision.

Etiology. There are many possible causes of papillitis. These include diseases that result in demyelinating diseases such as multiple sclerosis (MS) and encephalomyeli-

tis; viral or bacterial infections such as poliomyelitis, measles, pneumonia, or meningitis; nutritional or metabolic disorders such as diabetes, pernicious anemia, and hyperthyroidism; secondary complications of other diseases; reactions to toxic substances such as methanol, quinine, salicylates, and arsenic; and trauma.

In patients over 60 years of age, a common cause of papillitis is temporal arteritis (giant cell arteritis). In such cases, papillitis can spread to the other eye resulting in bilateral blindness.

Clinical Picture. An acute loss of vision that is usually unilateral. There is sudden onset and rapid progress, complete blindness sets in rapidly in untreated cases.

Complaints. Rapid vision loss within hours or days, eye movement pain.

Signs. Reduced visual acuity (VA), afferent pupillary defect, impaired color vision, central or paracentral scotomas, swollen optic disc.

Methods of Examination. VA test, testing of color vision, swinging flash light test, perimetry, ophthalmoscopy, MRI.

Differential Diagnosis. Papilledema, pseudopapilledema.

Treatment is directed toward the underlying cause of papillitis that is why it is important to have a full medical exam when vision loss and pain do occur.

There are two stages of treatment — immediate (before the etiology is clarified) and etiological. It is primarily treated with corticosteroid drugs. If MRI of the brain shows lesions of multiple sclerosis, the patient should receive immediate intravenous methylprednisolone for 3 days followed by oral prednisolone.

Other treatment is symptomatic and supportive. It includes antibiotic therapy and by indications — antiviral drugs. Combined therapy is focused on the following processes: to normalize the blood flow in small vessels — anticoagulants; to accelerate the process of metabolism and to restore damaged nerve tissues — biogenic stimulators, immunostimulatory agents, enzymes, osmotherapy; in order to improve visual functions vitamins B_1, B_6, B_{12}, C are prescribed.

Prognosis depends on the underlying disorder. Patient's eyesight often returns to normal without treatment in about two to three weeks. People who have reoccurring attacks of papillitis often suffer permanent vision loss.

Complications. Optic nerve atrophy with partial or total vision loss.

Prophylaxis is timely and adequate treatment of diseases, which can cause the development of optic neuritis, regular examinations by an ophthalmologist, healthy life style, regular checkups by specialists.

Retrobulbar Neuritis

Definition. Retrobulbar neuritis is an inflammation of that portion of the optic nerve that lies behind the eyeball.

Etiology. Many cases of this disease are caused by multiple sclerosis while others may be due to viral or infectious disorders such as meningitis, syphilis or allergic reactions. In most cases there may be no apparent cause.

Clinical Picture. This disease usually affects one eye and is characterized by pain associated with movement of the eye, headache and a sudden and progressive loss of vision that can result in complete blindness. Symptoms usually worsen for two weeks and then stabilize.

Complaints. An acute loss of vision that may progress over a few days and then slowly improve, pain on eye movement, tenderness of the eyeball to touch or pressure, neuralgia and headache, change in color perception.

Signs. Loss of visual acuity, relative afferent pupillary defect, central scotoma. In the early stages the optic disk is normal. Only in a month or later the optic nerve head may become pale.

NOTE!

In retrobulbar neuritis, the patient sees nothing due to central scotoma; the ophthalmologist sees nothing as the fundus appears normal.

Methods of Examination. VA test, color vision evaluation, swinging flash light test, perimetry, ophthalmoscopy, MRI.

Differential Diagnosis. Indirect optic neuropathy, posterior ischemic optic neuropathy.

Treatment is directed towards finding out and treating the underlying cause. The treatment must be carried out on the same principles as the treatment of patients with papillitis.

There are two stages of treatment — immediate (before the etiology is clarified) and etiological. It is primarily treated with corticosteroid drugs. If MRI of the brain shows lesions of multiple sclerosis, the patient should receive immediate intravenous methylprednisolone for 3 days followed by oral prednisolone.

Other treatment is symptomatic and supportive. It includes antibiotic therapy and by indications — antiviral drugs. Combined therapy is focused on the following processes: to normalize the blood flow in small vessels — anticoagulants; to accelerate the process of metabolism and to restore damaged nerve tissues — biogenic stimulators, immunostimulatory agents, enzymes, osmotherapy; in order to improve visual functions vitamins B_1, B_6, B_{12}, C are prescribed.

NOTE!

Don't use oral prednisolone alone as a primary treatment because of increased risk of reoccurrence.

Prognosis depends on the cause. Cases in which there is no obvious cause or in which the cause is multiple sclerosis often improve after two weeks, but the vision may never completely return to normal. People who have reoccurring cases of retrobulbar neuritis often suffer from permanent vision loss.

Complications. Optic nerve atrophy with partial or total vision loss.

Prophylaxis is timely and adequate treatment of diseases, which can cause the development of optic neuritis, regular examinations by an ophthalmologist, healthy life style, regular checkups by specialists.

4.3. Degenerative Diseases

Optic Nerve Atrophy

Definition. Optic nerve atrophy (optic atrophy) is damage to the optic nerve resulting in its degeneration or destruction. Optic atrophy is not a disease, but rather a sign of a potentially more serious condition that can cause decreased vision, including blindness.

Etiology. There are many possible causes of optic nerve atrophy (ONA). These causes may include glaucoma, vascular disorders, inflammation, degenerative and compressive disorders, toxins or nutritional deficiencies, syphilis, trauma and systemic disorders. This condition can also be caused by diseases of the brain and central nervous system, such as: brain tumor, multiple sclerosis, stroke. It may be inherited or acquired. Sometimes optic atrophy occurs without a known or proven cause.

Clinical Picture. Optic nerve atrophy is classified according to ophthalmoscopic appearance as primary and secondary. According to the direction of optic nerve degeneration it can be ascending or descending. Etiological classification of ONA is based on the etiology of the pathologic process — vascular, inflammatory, compressive, glaucomatous, traumatic, toxic, congenital or hereditary, etc.

Primary ONA is most commonly caused by MS, compression conditions from intracranial tumors, vascular and inflammatory disorders, exogenous toxins. This type reflects a chronic process and is not preceded by edema of the optic disc. It is characterized by pale or white disc with sharply delineated margins.

The most often cases of secondary ONA are edema or inflammation of the nerve head. A characteristic feature of secondary optic atrophy is a white optic disc with blurred indistinct margins.

Ascending (consecutive) optic atrophy — the lesion is located anterior to the lamina cribrosa in the ocular portion of the optic nerve or retina.

Descending optic atrophy — the lesion is located posterior to the lamina cribrosa in retrobulbar or cranial location.

Atrophy can be total or partial; depending on its course it can be stable or progressive. Total stable optic atrophy is uncurable; partial can be treated especially if visual functions are dynamic.

Complaints. Reduced VA (visual acuity), blurred vision, decrease in sharpness and clarity of vision, colors seem faded, difficulties with peripheral vision.

Signs. The optic disc is pale — to help elicit this sign the comparison with the fellow eye may help. There is usually a reduction of the small blood vessels crossing its surface and, in the case of secondary atrophy, the disc margin may be poorly delineated (this is due to gliosis rather than edema) (fig. 14.2). Where the atrophy is glaucomatous in origin, disc cupping will also be present. Pupillary reactions are absent.

Methods of Examination. VA, pupillary light reflex, color vision testing, perimetry, tonometry, ophthalmoscopy, electroretinography (ERG), visual evoked potential (VEP), OCT and HRT, MRI test when MS or brain tumor are suspected.

Differential Diagnosis. Axial myopia, optic nerve pit, optic disc drusen, optic nerve hypoplasia, myelinated nerve fibres.

Treatment. Unfortunately, there is no effective treatment for optic atrophy. Once the nerve fibres of the optic nerve are lost they never heal or restore. However, early diagnosis and treatment of the underlying causes of optic atrophy can help prevent further damage from the disease.

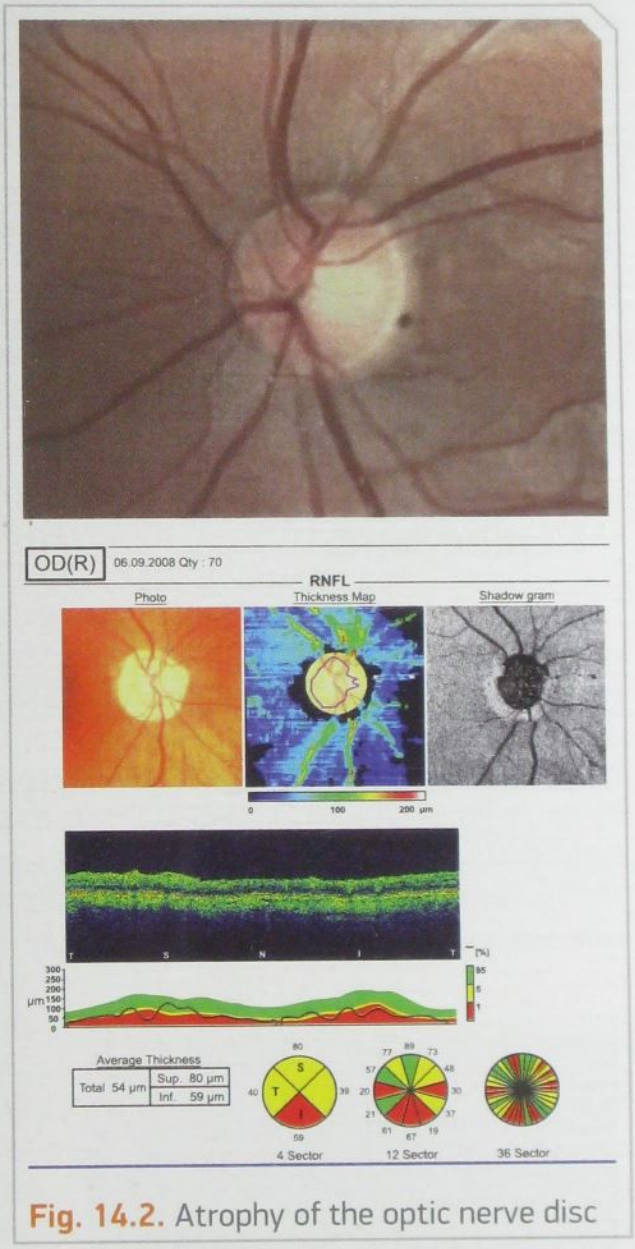


Fig. 14.2. Atrophy of the optic nerve disc

Treatment of optic nerve atrophy is directed at blood flow increase and improvement of optic nerve trophism, stimulation of the vital function of preserved nerve fibres and those that are in the stage of parabiosis. It includes stem cell treatment, B vitamins, nicotinic acid and other vasodilatators, oxygen theraphy, extract of ginseng, eleutherococcus. Recently such therapies as acupuncture, the use of magnetic fields, ultrasound, physiotherapy and electrical stimulation got spread. Promising in the treatment of partial optic atrophy is the use of endovascular surgery, which is based on redistribution of the blood supply of the brain, can improve blood supply of the optic nerve at atrophy due to ischemia.

Prognosis depends on the cause of the disorder. Vision lost due to optic nerve atrophy cannot be recovered. If the cause can be found and controlled, further vision loss and blindness may be prevented. In some cases, ONA can lead to total blindness.

Complications are related to the disease that causes the atrophy. In some cases, a complete and irreversible loss of vision can occur.

Prophylaxis. Many cases of optic nerve atrophy can not be prevented, since it is caused by a variety of diseases or conditions. Nevertheless, timely and optimal treat-

ment of the underlying problem, routine eye exams can help with an early diagnose of any damage to the optic nerve and slow vision loss.

4.4. Compressive Optic Neuropathy

Compressive optic neuropathy is the condition caused by a compression of the optic nerve. The main causes of this type of neuropathy are infection, swelling, trauma, aneurysms, orbital and intracranial diseases, cerebral tumors or abscess that result in disc swelling and vision impairment.

Papilledema

Definition. Papilledema (edema of the optic nerve disc) is a non-inflammatory swelling of the optic nerve head secondary to increased intracranial pressure (fig. 14.3).

Etiology. Causes of papilledema include all factors that may increase the intracranial pressure such as lesions and tumors in the central nervous system (CNS), malignant hypertension, intracranial hemorrhages, hydrocephalus, cerebral abscess, intracranial infection such as meningitis and encephalitis, head injury. The use of some medications, for example, tetracycline and corticosteroids may cause the pathology.

Clinical Picture. As a rule, papilledema is bilateral and not associated with significant loss of central vision or visual field unless it continues untreated for a long period. In the early stages vision remains normal but the patient notices temporary blurring of vision (lasting seconds). Another most common early visual finding is enlargement of the blind spot. Color vision and pupillary exam are often normal. If the disease progresses, vision loss is gradual and painless.

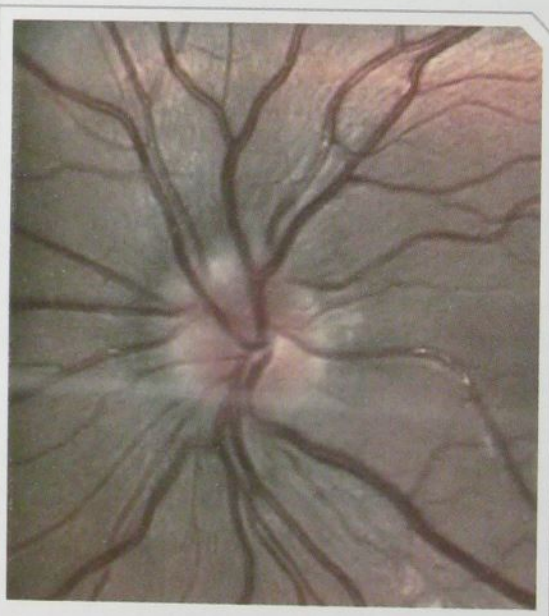


Fig. 14.3. Edema of the optic nerve disc

Complaints. Short-term blurring or loss of vision, headache, nausea, vomiting, constricted visual field, double vision.

Signs. On the basis of fundus appearance papilledema may present in the following four phases.

Early phase. Hyperemia of the optic disc, blurring of all borders, swelling of the nasal and then superior and inferior margins ("C-shape"). The temporal mar-

gin is normal. The optic cup is preserved. Retinal venous pulsations are not present. Concentric peripapillary retinal folds known as Paton's folds.

Developed (acute) phase. Blurring and swelling of all borders, increased diameter of the optic nerve head, obscuration of vessels at the disc margin. The color of the optic disc is grayish red.

Chronic phase. Significant swelling of the entire nerve head, blurring of all borders. The disc appears pale, the cup may be obscured. The veins are dilated and tortuous.

Atrophic phase. A gray disc that loses its central cup. The vessels largely but not completely obscured on the disc surface and lost at the margins. With time, the disc may develop small glistering crystalline deposits.

Methods of Examination. VA, pupillary light reflex, color vision testing, perimetry, ophthalmoscopy, HRT, CT scans, MRI.

Differential Diagnosis. Papillitis, pseudopapilledema, ischemic optic neuropathy, glaucoma (table 14.1).

Table 14.1

Differential Diagnosis of Papillitis and Papilledema

Diagnostic Findings	Papillitis	Papilledema
Visual Acuity	Reduced	Usually normal
Pupil	Poor response to direct light	Normal
Visual Field	Central scotoma	Normal (increased blind spot)
Color Vision	Defective	Normal
Site	Usually unilateral	Usually bilateral

Treatment. The treatment of papilledema is primarily focused on the treatment of the underlying cause of increased intracranial pressure. If a mass is present, primary therapy should be directed towards that. If medications (tetracyclines, vitamin A analogues, etc.) are felt to be causative, they should be discontinued.

Hydration and osmotherapy combined with a weight reduction program may be useful in cases of papilledema that is caused by an abnormally high production of cerebrospinal fluid. Corticosteroids have been shown to be effective in relieving the symptoms in some patients with papilledema caused by inflammatory disorders.

Alternative treatments for conditions that cause the occurrence of papilledema include acupuncture, aromatherapy, hydrotherapy, massage, and herbal remedies.

Once intracranial pressure has been normalized, papilledema will resolve within few weeks or months.

Prognosis. With prompt medical care of the underlying cause of papilledema, a person will not have permanent damage to vision. However, prolonged papilledema can result in damage to the optic nerve, which could lead to blindness.

Complications. Left untreated papilledema may eventually lead to permanent blindness.

Prophylaxis. Preventing papilledema is only possible if the underlying condition causing papilledema can be found. Treatment of this underlying condition may prevent recurrences of papilledema.

4.5. Vascular Disorders

Ischemic Optic Neuropathy

Definition. Ischemic optic neuropathy is a damage to the optic nerve resulting from insufficient blood supply, sometimes called stroke or infarct of the optic nerve.

Etiology. Ischemic optic neuropathy occurs most commonly in people over age 55. Most ischemic optic neuropathy is unilateral. Bilateral, sequential cases occur in about 20 %, but bilateral simultaneous involvement is uncommon.

This pathology can be caused by an inflammatory disorder of the arteries that typically affects the temporal arteries (giant-cell arteritis) — arteritic ischemic optic neuropathy, or by cardiovascular diseases such as hypertension, atherosclerosis, hypotension, diabetes, rheumatoid artritis — non-arteritic ischemic optic neuropathy.

Clinical Picture. Ischemic optic neuropathy may be of two types depending on the part of the optic nerve involved:

Anterior ischemic optic neuropathy is caused by acute ischemia of the anterior part of the optic nerve (optic nerve head), which is mainly supplied by the posterior ciliary arteries. This results in a severe loss of vision and inferior visual field defects, a pale, swollen optic disc with peripapillary hemorrhages.

Posterior ischemic optic neuropathy is less common and occurs due to acute ischemia of the posterior part of the optic nerve, located some distance behind the eyeball. Impairment of visual acuity in ischemic optic neuropathy may vary from slight — with a corresponding decrease in color vision — to no light perception. The optic disc in this case is not swollen.

Complaints. Rapid (over minutes, hours or days) and painless loss of vision usually in one eye.

Signs. In case of anterior ischemic optic neuropathy an afferent pupillary defect is present, the optic disc is pale and swollen, the disc margins are blurred, multiple hemorrhages (fig. 14.4).

In case of posterior ischemic optic neuropathy the ophthalmoscopic picture may be normal and reveal only arteriosclerotic changes of the retinal vessels.

Methods of Examination. Ophthalmoscopy, perimetry, Doppler flow imaging, CT, MRI.

Differential Diagnosis. Central retinal vein occlusion, papilledema.

Treatment of ischemic optic neuropathy must be complex, taking into account the overall vascular pathology, coagulation status, and lipid metabolism. With simultaneous intensive treatment of the underlying diseases, ischemic optic neuropathy is treat-

ed with vasodilatators, thrombolytics, corticosteroids, neurotrophic drugs, vitamins, various types of fibre optic nerve stimulation.

In recent years, the so-called targeted therapy of ischemic damage to the optic nerve is developing, which involves the use of special vascular catheters for endovascular drug administration directly to the lesion.

Prognosis. There is no effective treatment for the arteritic variety, and most lost vision is not recovered; however, in nonarteritic cases, up to 40 % of patients spontaneously recover some useful vision.

Complications. Irreversible loss of vision.

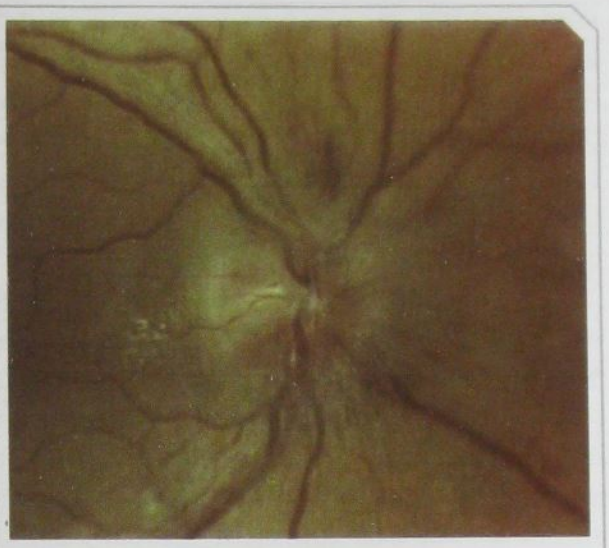


Fig. 14.4. Edema of the optic nerve disc with hemorrhages

Prophylaxis. Ischemic optic neuropathy can be prevented by optimal management of the underlying problem.

4.6. Nutritional and Toxic Optic Neuropathies

Definition. Nutritional and toxic optic neuropathy is a damage to the optic nerve that results from nutritional deficiencies, some medications and toxin or poison substances and cause gradual or sudden vision loss.

Etiology. The most common optic neuropathy from poisons and toxins is called tobacco-alcohol amblyopia, thought to be caused by cyanide from tobacco smoking. Other toxins are methanol (wood or methyl alcohol), ethylene glycol (antifreeze), cyanide, lead, etc. Nutritional optic neuropathy (sometimes called nutritional amblyopia) typically involves deficiencies in vitamin B_{12} (cobalamin), vitamin B_1 (thiamine), vitamin B_2 (riboflavin), and folic acid. Nutritional optic neuropathy is clearly more common among tobacco and alcohol abusers.

Clinical Picture. Visual loss is painless, bilateral, and simultaneous though the involvement can be asymmetric in the acute phase. There is usually no relative afferent pupillary defect since the disease is almost always bilateral and symmetric. Most patients will present with normal-appearing optic discs in the early stages. Optic disc edema may be seen in some toxic optic neuropathies. Papillomacular bundle loss and optic atrophy (especially temporal atrophy) finally develop in the chronic stages. Bi-

lateral central or cecocentral scotomas with preservation of the peripheral visual field are characteristic of these optic neuropathies.

Complaints. Patients complain of painless, progressive, bilateral, symmetrical visual loss, changing in color perception or seeing faded colors. In case of methanol ingestion patients may have no perception of light.

NOTE!

Ethylene glycol and particulary methanol poisoning can cause sudden and complete loss of vision. Both substances can cause other serious symptoms such as abdominal pain, vomiting, difficulty breathing, coma and even death.

Signs. The optic disc may appear normal, swollen, or hyperemic in the early stages. Temporal pallor of the optic disc is seen in later stages. The pupils usually demonstrate a normal response to light. In those who are practically blind, the pupils will be dilated with a weak or absent response to light. Dyschromatopsia. Visual field shows central or cecocentral scotoma.

Methods of Examination. Ophthalmoscopy, perimetry, MRI.

Differential Diagnosis. Optic neuritis, compressive optic neuropathy.

Treatment. The first step in managing toxic optic neuropathy is to remove the offending agent. This may cause some reversal of the process, especially if removed early in the course of optic nerve dysfunction. For nutritional optic neuropathies, improved nutrition is essential.

Treatment in the early stages includes total detoxication, decongestants, anti-inflammatory therapy, use of vitamins, especially C, thiamine, folic acid, and vitamin B_{12} injections monthly for several months. For visual functions recovery neurotrophic, retina-protective drugs, various types of stimulation of the retina and the optic nerve are used.

If ethylene glycol or methanol poisoning is the cause, rapid treatment with hemodialysis and an alternative alcohol taking may help.

Prognosis. With prompt treatment, most people recover some of the lost acute and color vision.

Complications may include severe bilateral visual loss and various non-ophthalmological symptoms and signs from underlying causes, such as tobacco use and poor nutrition.

Prophylaxis. Avoiding alcohol and other chemicals or drugs that may be toxic. Well-balanced diet and vitamin supplements.

Review:

1. Key points

Diseases of the optic nerve by origin may be *congenital* (hereditary or developmental) *or acquired*; by the character of the pathological process — *acute and chronic*; by the dynamics of the pathological process — *fixed and progressive*; by pathogenesis — *inflammatory, degenerative, compressive, vascular, nutritional and toxic, tumors, traumas* (direct or indirect). The main sympptoms of optic nerve diseases are blurred vision, central vision loss that can be gradual or sudden, changes in color perception, peripheral vision scotomas. The examination consists of visual acuity, ophthalmoscopy, color vision testing, pupillary light reflex, perimetry, visual evoked potential, CT and MRI scans. Examination of the optic nerve disc should consider the following aspects — size and shape, margins, color, physiological excavation and cupping size.

Congenital anomalies are optic atrophy, Leber's hereditary optic neuropathy, optic nerve hypoplasia, optic disc coloboma, myelinated nerve fibres, morning glory syndrome, optic disc drusen, pseudopapillitis.

Inflammatory diseases are optic neuritis that may affect the optic disc — papillitis, or the optic nerve behind the eyeball — retrobulbar optic neuritis. They are characterized by rapid loss of vision and pain on eye movement. In case of optic neuritis the optic disc is swollen, in retobulbar neuritis — normal at early stages and becomes pale temporally at later stages.

Optic atrophy may be ascending or descending and shows a pale optic nerve head, the disc margins may be blurred or sharply outlined depending on the etiology of the atrophy, disc cupping will also be present.

Papilledema is characterized by swelling of the optic nerve head. Patients notice short-term blurring or loss of vision.

Ischemic optic neuropathy according to the etiology may be arteritic or non-arteritic, depending on the part of the optic nerve involved — anterior or posterior. In case of anterior ischemic optic neuropathy the optic disc is pale and swollen, the disc margins are blurred, multiple hemorrhages. In case of posterior ischemic optic neuropathy the ophthalmoscopic picture may be normal.

Nutritional and toxic optic neuropathy may result in painless, progressive, bilateral, symmetrical visual loss, changes in color perception. In case of methanol ingestion patients may have no perception of light. The optic disc may appear normal, swollen, or hyperemic in the early stages. Temporal pallor of the optic disc is seen in later stages.

The prognosis of optic nerve diseases is usually unfavourable and depends on timely treatment. As prevetion methods treatment of underlying causes and maintaining a healthy lifestyle are important.

2. Diagrams

DISEASES of the OPTIC NERVE
Congenital (Hereditary Developmental)
Optic Disc Coloboma
Leber's Hereditary Optic Neuropathy
Optic Nerve Hypoplasia
Morning Glory Syndrome
Myelinated Nerve Fibers
Optic Disc Drusen
Pseudopapillitis
Inflammatory
Optic Neuritis
Papillitis
Retrobulbar Neuritis
Degenerative
Optic Atrophy
Primary or Secondary
Ascending or Descending
Total or Partial
Stable or Progressive
Compressive
Papilledema
Vascular
Ischemic Optic Neuropathy
Anterior
Arteritic
Non-Arteritic
Posterior
Nutritional and Toxic
Tobacco-Alcohol Amblyopia
Methyl Alcohol Amblyopia
Vitamin Deficiency
Tumors
Traumas

3. The Review Questions

A. Control Questions

1. What are the main groups of optic nerve diseases?
2. What main examination methods of optic nerve diseases do you know?
3. What are the main symptoms of optic nerve diseases?
4. What are the factors affecting the state of the optic nerve?
5. What are the inflammatory diseases of the optic nerve and their clinical picture?
6. What are the vascular disorders of the optic nerve and their clinical picture?
7. What is optic atrophy and what are the causes of degenerative diseases of the optic nerve?
8. What is papilledema, its etiology and clinical picture?
9. What are nutritional and toxic neuropathies and their causes?
10. What are the main principles of treatment and preventive methods of optic nerve diseases?

B. Tests

1. **Symptoms of optic nerve diseases are:**
   A. Short-term blurring of vision
   B. Exophthalmos
   C. Painless loss of vision
   D. Dyschromatopsia
   E. Double vision
   F. Photophobia

2. **The main diagnostic methods of optic nerve diseases are:**
   A. Pupillary light reaction test
   B. Perimetry
   C. Visual evoked response
   D. MRI
   E. Color vision test
   F. Ophthalmoscopy

3. **Examination of the optic nerve disc should consider the following aspects:**
   A. Size and shape
   B. Blood vessels caliber
   C. Color
   D. Macula color
   E. Margins
   F. Cupping size

4. **What are the main causes of optic neuritis?**
   A. Bacterial infections
   B. Allergic reactions
   C. Multiple sclerosis
   D. Sinusitis
   E. Diabetes
   F. Gastrointestinal disease

5. **The symptoms of papilledema are:**
   A. Sudden loss of vision
   B. Short-term blurring or loss of vision
   C. Color vision pathology
   D. Double vision
   E. Enlargement of the blind spot
   F. Poor response to direct light

6. **The main causes of optic atrophy are:**
   A. Vascular disorders
   B. Glaucoma
   C. Multiple sclerosis
   D. Brain tumor
   E. Toxins or nutritional deficiencies
   F. Compressive disorders

7. **What are the symptoms of ischemic optic neuropathies?**
   A. Enlargement of the blind spot
   B. Headache
   C. Rapid loss of vision
   D. Color vision decrease
   E. Bilateral loss of vision
   F. Cecocentral scotoma

8. **What are the symptoms of nutritional and toxic optic neuropathy?**
   A. Unilateral loss of vision
   B. Bilateral loss of vision
   C. Pain on eye movement
   D. Dyschromatopsia
   E. Cecocentral scotoma
   F. Normal response to light

9. **What are the main ophthalmoscopic signs of optic atrophy?**
   A. Crystalline deposits
   B. Swelling of the optic disc
   C. Disc cupping
   D. Multiple hemorrhages
   E. Pale optic disc
   F. Sharply delineated margins

10. **The treatment methods of optic nerve diseases are:**
    A. Neurotrophic drugs
    B. B vitamins
    C. Vasodilatators
    D. Corticosteroids
    E. Treatment of underlying causes
    F. Endovascular surgery

C. Clinical Cases

Case 1

A 40-year-old female complains of cloudy vision of the right eye. Two weeks prior to her clinic visit she noticed visual loss in her right eye. It was accompanied by pain on eye movements and a dull retro-orbital ache. She also noted decreased perception of color and contrast. Ophthalmoscopy of the right and left eyes shows no optic disc edema, light temporal pallor of the optic disc in the right eye. What is the diagnosis?

A. Papillitis
B. Retrobulbar neuritis
C. Papilledema
D. Ischemic optic neuropathy
E. Optic atrophy
F. Nutritional and toxic optic neuropathy

Case 2

A 60-year-old male patient complains of rapid and painless loss of vision in the left eye which he noticed upon awakening. Vision on the left eye is obscured by a dark shadow. VA — light perception, color vision is lost. Perimetry reveals an inferior altitudinal defect. Ophthalmoscopy — the optic disc is pale and swollen, the disc margins are blurred, multiple hemorrhages. What is the diagnosis?

A. Papillitis
B. Retrobulbar neuritis
C. Papilledema
D. Ischemic optic neuropathy
E. Optic atrophy
F. Nutritional and toxic optic neuropathy

Case 3

A 37-year-old patient complains of short-term blurring of vision on both eyes, headache, nausea, vomiting especially when rising from a lying or sitting position. He also notices constriction of the visual field, decreased color perception and diplopia. In anamnesis — head trauma. Ophthalmoscopic examination reveals a bilateral hyperemic and swollen optic disk, blurred margins, concentric peripapillary retinal folds, absent venous pulsation. Visual acuity 0.7. Pupillary response to light is normal. Visual field testing detects an enlarged blind spot. What is the diagnosis?

A. Papillitis
B. Retrobulbar neuritis
C. Papilledema
D. Ischemic optic neuropathy
E. Optic atrophy
F. Nutritional and toxic optic neuropathy

Case 4

A 54-year-old patient complains of progressive bilateral loss of vision after drinking unknown alcohol. He had abdominal pain, nausea, vomiting and unconsciousness. VA — no perception of light. The pupils are dilated and do not respond to light. Ophthalmoscopy reveals bilateral temporal optic atrophy. What is the diagnosis?

A. Papillitis
B. Retrobulbar neuritis
C. Papilledema
D. Ischemic optic neuropathy
E. Optic atrophy
F. Toxic optic neuropathy

Case 5

A 62-year-old patient complains of reduced and blurred vision, decrease in sharpness and clarity of vision, dimness of colors, difficulties with peripheral vision. VA — 0.4. Pupillary reactions are absent. Color vision testing reveals dyschromatopsia. Ophthalmoscopic picture — the optic discs are grayish, pallor temporally, the margins are sharply delineated, disc cupping, reduction of the small blood vessels crossing the margins. What is the diagnosis?

A. Papillitis
B. Retrobulbar neuritis
C. Papilledema
D. Ischemic optic neuropathy
E. Optic atrophy
F. Toxic optic neuropathy

CHAPTER 15

Diseases of the Vitreous Body

OBJECTIVES

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

— know the main pathologies of the vitreous body and their classification;
— know the basic diagnostic methods of vitreous body examination;
— know the signs and symptoms of vitreous body pathology;
— evaluate and manage the patients with vitreous body diseases;
— know the principles of vitreous body disease treatment.

Plan:

1. **CLASSIFICATION OF VITREOUS BODY DISEASES**
2. **SYMPTOMS OF VITREOUS BODY DISEASES**
3. **EXAMINATION METHODS**
4. **DISEASES OF THE VITREOUS BODY**
   - **4.1. Developmental Vitreous Body Abnormalities**
     - Persistent Hyperplastic Primary Vitreous
     - Persistent Hyaloid Artery
   - **4.2. Age-Related Vitreous Body Changes**
     - Vitreous Liquefaction
     - Posterior Vitreous Detachment
   - **4.3. Vitreous Opacities**
   - **4.4. Vitreous Hemorrhage**
   - **4.5. Vitreous Inflammation**

1. Classification of Vitreous Body Diseases

Diseases of the vitreous body can be classified:
— according to the origin — developmental or acquired;
— according to the pathogenesis — age-related, degenerative, dystrophies, inflammatory, traumatic;
— according to the type of opacity — muscae volitantes, asteroid hyalosis, synchysis scintillans, amyloidosis, red cell opacities, inflammatory cells opacities, tumor cells opacities, vitreous hemorrhage.

1.1. Developmental Vitreous Body Abnormalities — persistent hyperplastic primary vitreous, persistent hyaloid artery.
1.2. Age-Related Vitreous Body Changes — vitreous liquefaction, posterior vitreous detachment.
1.3. Vitreous Opacities — muscae volitantes, asteroid hyalosis, synchysis scintillans, amyloidosis, red cell opacities, inflammatory cell opacities, tumor cell opacities, vitreous hemorrhage.

2. Symptoms of Vitreous Body Diseases

— Floaters
— Flashes
— Cloudy vision
— Visual loss

3. Examination Methods

— Slit-lamp exam
— Ophthalmoscopy
— B-scan ultrasound
— CT scan
— MRI

4. Diseases of the Vitreous Body

4.1. Developmental Vitreous Body Abnormalities

Persistent Hyperplastic Primary Vitreous

Definition. Persistent hyperplastic primary vitreous (PHPV) is a developmental anomaly, due to persistence of embryonic remnants of the primary vitreous and part of the adjacent vascular structures.

Etiology. The cause of PHPV is unknown. Normally the embryonic vascular system in the vitreous body and lens disappears completely by the time of birth, leaving only the *hyaloid canal*. PHPV, also known as persistent fetal vasculature (PFV), results from incomplete regression of the primary embryonic vitreous and hyaloid vasculature in the anterior and/or posterior chambers. Most examples of PHPV are unilateral (in 90 % cases) and non-hereditary.

Clinical Picture. PHPV is characterized as a condition with leukocoria (white pupil) due to the fibrous vascularized membrane behind the lens, often associated with congenital cataract, glaucoma, microphakia, microphthalmia, and often retinal detachment, which lead to poor visual prognosis. It can be present in three forms: anterior, posterior, and a combination of both. There are usually no associated systemic findings.

Complaints. White eye reflex, small and lazy eye.

Signs. Leukocoria, microphthalmia, cataract, microphakia, strabismus, poor vision.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, B-scan ultrasonography, CT scan, MRI.

Differential Diagnoses. Congenital cataract, retinopathy of prematurity, retinoblastoma.

NOTE!

Leukocoria should be regarded as a retinoblastoma until proven otherwise.

Treatment should be started as early in life as possible to avoid the risk of damage to the eyeball and amblyopia. Surgical management of PHPV depends on its type — most often lensectomy, capsulorhexis of the posterior capsule, insertion of an intraocular lens in the posterior chamber, glaucoma management, posterior vitrectomy and intensive amblyopia therapy.

Prognosis depends primarily on the severity of the disorder. However, adequate surgical intervention can often save the eye and stabilize visual acuity even if at a very low level.

Complications. Secondary glaucoma, tractional retinal detachment, persistent intraocular hemorrhages, loss of vision of the affected eye.

Prophylaxis. There are no preventive methods of the condition.

Persistent Hyaloid Artery

Definition. Persistent hyaloid artery (PHA), or tunica vasculosa lentis, is a unilateral congenital anomaly of the eye that is associated with incomplete regression of the hyaloid artery.

Etiology. The hyaloid artery is a part of the embryonic vascular supply of the lens. It extends from the optic disc to the lens via the vitreous body. Regression of the hyaloid artery leaves a clear central zone in the vitreous humor called the hyaloid or Cloquet's canal. Normally the hyaloid artery regresses by the time of birth. PHA is usually unilateral and occurs in 95 % of premature infants.

Clinical Picture. PHA may be partial or complete. Partial remnants of the anterior portion of the artery appears as a tiny thread-like strand attached to the posterior capsule of the lens with its loose end floating in the vitreous. It can be seen as a small pinpoint-like scar on the posterior surface of the lens (Mittendorf's dot). If a partial posterior remnant persists, it appears as a grey cord attached to the optic disc with its free end moving in the vitreous with every eye movement. The optic disc may have a conical shape — Bergmeister's papilla. Rarely, the entire hyaloid artery (patent or occluded) may be present as a whitish cord extending from the optic disc to the posterior lens capsule.

Complaints. Often asymptomatic, dark shadows in the visual field.

Signs. Mittendorf's dot located at the posterior lens capsule or as Bergmeister's papilla located at the optic disc, a single sinuous vessel extending from the posterior lens capsule or the optic disc that moves at eyeball movements.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, B-scan ultrasound.

Differential Diagnoses. Posterior capsular or subcapsular cataract, optic disc coloboma, optic nerve hypoplasia.

Treatment. PHA alone rarely requires treatment as it usually doesn't influence visual functions. If the associated cataract formation leads to visual impairment, however, surgery may be indicated.

Prognosis is good.

Complications. Visual impairment, amblyopia.

Prophylaxis. There are no preventive methods of the condition.

4.2. Age-Related Vitreous Body Changes

Ageing is accompanied by significant physical, structural and biochemical changes in the vitreous. The most important changes are vitreous liquefaction and posterior vitreous detachment.

Vitreous Liquefaction

Definition. Vitreous liquefaction is a normal age-related degeneration of the vitreous as a result of which it loses its gelatinous consistency becoming liquid.

Etiology. In the infant the vitreous is a homogeneous gel-like body. With aging, the gel volume decreases and the liquid volume increases. By age of 40 years the vitreous is 80 % gel and 20 % liquid, and by 70 or 80 years it is 50 % liquid, with most of the liquefaction occurring in the central vitreous.

In addition to the aging vitreous, liquefaction can be caused by high myopia, uveitis, blunt or penetrating trauma, thermal and radiation burns.

Clinical Picture. The vitreous gel naturally undergoes liquefaction, resulting in small pockets of more liquid vitreous lying within the firmer gel. This process typically begins in front of the macula or in the central vitreous cavity. The boundary between each liquid pocket and the gel may be noticeable to a patient as eye floaters.

As the result of the vitreous gel volume reduction the concentration of the collagen fibrils within it increases. As a part of this process, fine collagen fibrils clump into bundles that obstruct light as it passes through the eye causing shadows on the retina, which appears as floaters.

Complaints. Patients notice black floaters in front of the eye, usually seen on bright background.

Signs. Visible pockets of liquefaction in the vitreous gel and shadows on the retina are observed at biomicroscopy.

Methods of Examination. Slit-lamp exam, ophthalmoscopy.

Differential Diagnoses. Posterior vitreous detachment, vitreous hemorrhage, any other eye diseases that can cause vitreous opacification.

Treatment. There is no medical treatment for vitreous liquefaction. If floaters are too excessive and disturb vision, vitrectomy may be necessary.

Prognosis. Decrease of vision.

Complications. Posterior vitreous detachment, macular hole.

Prophylaxis. There are no preventative measures to the development of vitreous liquefaction as it is a part of the natural ageing process.

Posterior Vitreous Detachment

Definition. Posterior vitreous detachment (PVD) is the separation of the vitreous body from the retina.

Etiology. PVD occurs in the majority of population over 70 years as a result of aging vitreous liquefaction and shrinkage. It can be caused by trauma, some degenerative or inflammatory conditions, high myopia, cataract surgery.

Clinical Picture. Many people are not aware that they have developed PVD but some notice symptoms such as floaters or flashing lights. Floaters can take many forms from little dots, circles, lines, to clouds or cobwebs. Sometimes people experience one large floater, which can be distracting and make difficult to read. With time, floaters will usually become less noticeable or disappear entirely on their own.

Complaints. Mobile dense shadows that obscure vision, flashes of light or sparkles at the side of vision, darkness of the peripheral visual field, decreased vision.

Signs. Biomicroscopic examination reveals a ring-like opacity (Weiss ring or Fuchs ring) anteriorly to the optic disc — the thickened posterior surface of the vitreous that has now become visible as it has pulled away from the optic disc. It looks like a thin irregular ring of translucent material floating in the vitreous.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, B-scan ultrasound, OCT.

Differential Diagnoses. Vitreous opacities, vitreous degenerations, vitreous hemorrhage, retinal detachment.

Treatment. There is no treatment that will put the vitreous back in position. If PVD has occurred without associated retinal tears, therapy is not required or indicated. The vitreous will continue to age and liquefy and floaters will usually become less noticeable. If a retinal tear has occurred, laser treatment or cryopexy is usually recommended. Surgery will be indicated if the tear has progressed to a retinal detachment or vitreous hemorrhage.

Prognosis. The majority of patients recover fully from their symptoms. But if PVD is followed by retinal tears or retinal detachment, it can result in blindness.

Complications. Retinal tear, retinal detachment, vitreous hemorrhage, retinal hemorrhages, cystoid maculopathy, vision loss.

Prophylaxis. There are no prevention methods of PVD caused by natural aging processes.

4.3. Vitreous Opacities

EYE FACTS

The word vitreous is derived from the Latin word vitrum, which means glass.

The vitreous is an optically transparent gel-like structure that transmits light rays to the retina. Any structural and biochemical changes or inclusions result in vitreous opacification that blocks light from reaching the retina causing fluttering shadows in the visual field, mild to severe vision loss.

Etiology. Commonly, mild opacification occurs due to age-related vitreous degeneration. However, a number of conditions can also lead to vitreous clouding. They are ocular and systemic inflammatory and idiopathic diseases, diabetes, trauma, laser treatment and surgery, intraocular tumors and vitreous parasitic diseases. Thickened collagen fibrils, red and white blood cells, inflammatory or tumor cells, deposits, calcium material, intraocular foreign bodies obstruct light and cast shadows on the retina leading to formation of floaters.

EYE FACTS

The majority of people (7 out of 10) may experience at some period of their lives. They are a normal sign of the aging process and do not require treatment as usually don't interfere with visual functions.

Floaters are shadows or spots in the field of vision that move or "float" when you look around. They can appear as different shapes, such as little dots, circles, lines, clouds or cobwebs. They can be transparent or dark or even black in color depending on the consistency. They may be especially noticeable when you look at something bright, such as white paper or the blue sky.

Clinical Picture. Common conditions associated with vitreous opacities are:

Muscae volitantes (or ***flying flies***) are physiological vitreous opacities that represent the residues of primitive hyaloid vasculature. Patients perceive them as fine spots or filaments seen floating before the eyes against a bright background (e. g., clear blue sky). They are not visible objectively with an ophthalmoscope.

NOTE!

Conditions requiring urgent ophthalmic examination are:

- acute onset of floaters;
- recent change of floater pattern;
- floaters associated with flashing;
- visual loss.

Asteroid hyalosis is a common degenerative process in which very small, white, rounded deposits of calcium containing lipids are suspended in the vitreous. It is an asymptomatic unilateral condition that is associated with ageing, diabetes, hypertension and hypercholesterolemia. At ophthalmoscopy these deposits are quite refractive giving the appearance of stars (or asteroids) shining in the night sky; except that ocular asteroids are mobile.

Synchysis scintillans (or ***cholesterolosis bulbi***) is a degenerative condition resulting in a liquefied vitreous body and accumulation of cholesterol crystals within it.

This condition develops after ocular trauma, inflammation or other ocular diseases and may occur at any age. This phenomenon appears as a beautiful "shower of golden rain" on ophthalmoscopic examination (fig. 15.1).

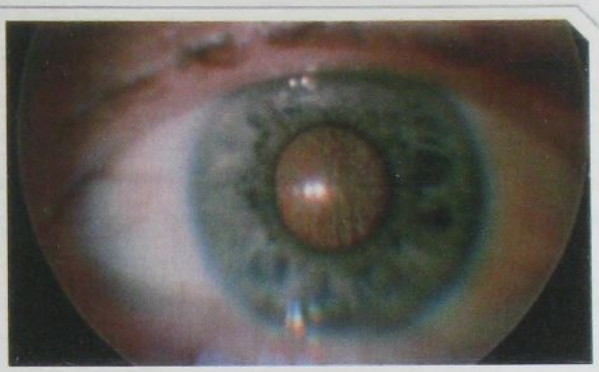


Fig. 15.1. Vitreous destruction of the "golden rain" type

Complaints. Floaters of different size, color and degree of transparency, haziness or decrease of vision.

Signs. Shadows on the retina, deposits of different form and color.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, B-scan ultrasonography.

Differential Diagnoses. Vitreous detachment, vitreous hemorrhages, macular hole, retinal detachment.

Treatment. In most cases vitreous opacification doesn't require treatment unless it causes vision impairment. It has been shown that our brain eventually learns to ignore the presence of floaters and a person does not notice them by that time. In severe cases vitrectomy may be performed. A novelty laser surgery called vitreolysis may be performed to remove deposits.

Prognosis. For patients who develop vitreous opacification from ageing the prognosis is quite good because with time they learn to ignore it. In patients, who have developed vitreous opacification from other causes, the outcome depends on its management and resolution may be quite limited.

Complications. None or vision impairment or loss depending on the underlying cause.

Prophylaxis. Most vitreous opacities occur as part of the natural aging process so they can't be prevented. If other conditions are related to vitreous opacification, their management may help.

4.4. Vitreous Hemorrhage

Definition. Vitreous hemorrhage is the leakage of blood into the vitreous or into the space created by vitreous detachment.

Etiology. Different causes such as vascular and metabolic disorders (diabetic and hypertensive retinopathy, anemia, leukemia, retinal vein occlusion), inflammatory diseases (chorioretinitis, retinal periphlebitis), blunt or penetrating eye trauma, posterior vitreous detachment, intraocular tumor may lead to either pre-retinal or vitreous hemorrhage. It is a relatively common cause of severe vision loss, having an annual incidence of approximately seven cases per 100,000 in the population.

The mechanisms of vitreous hemorrhage fall into three main categories: *abnormal vessels* (result of neovascularization) that are prone to bleeding, *normal vessels* that are damaged, or *blood from an adjacent source*.

Clinical Picture. Depending on the location of the hemorrhage and its volume, clinical picture can range from mild peripheral floaters to profound vision loss. A small hemorrhage at the periphery can be asymptomatic, in the macula region it can cause sudden painless loss of vision. For example, approximately 10 µl of blood is sufficient to reduce visual acuity to perception of hand movements in front of the eye.

Complaints. Appearance of small dots, fine particles of hair, shadows, cobwebs or red hue in the field of vision, cloudy or hazy vision, vision loss.

Signs. Black shadows against the red fundus reflex or absence of the red fundus reflex, no view to the fundus, presence of blood in the vitreous.

Methods of Examination. Slit-lamp exam, ophthalmoscopy, B-scan ultrasonography (fig. 15.2).

Differential Diagnoses. Vitreous detachment, retinal detachment, central retinal vein or artery occlusion, diabetic retinopathy.

Treatment consists of the following steps:

— *Bed rest with the head elevated by 30—45°* and with occasional bilateral patching to allow the blood to settle down, allowing a view of the superior peripheral fundus.

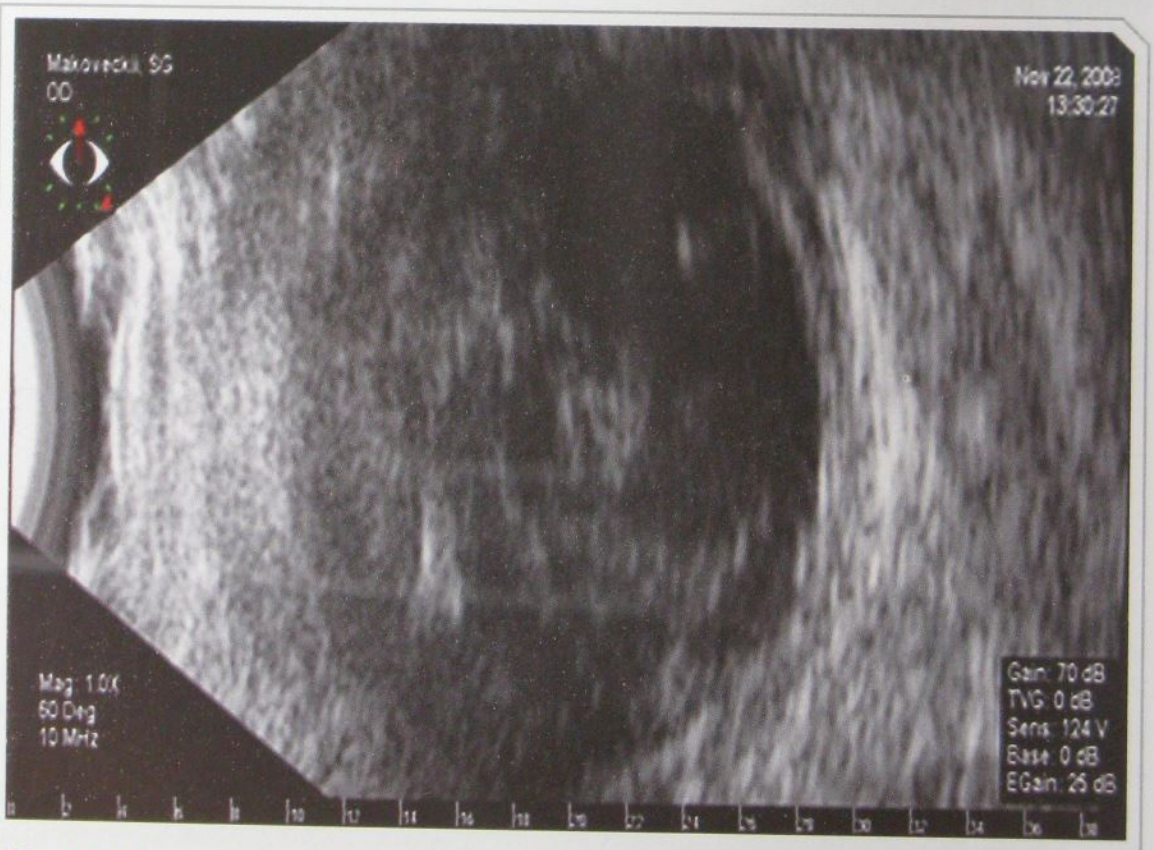


Fig. 15.2. Vitreous hemorrhage seen on ultrasound

— *Find the source of bleeding and stop it.* Once the source of the bleeding has been identified, treatment will depend on the cause. For example,
  - laser photocoagulation is used for treatment of fragile abnormal vessels, retinal tears and detachments;
  - intravitreal injections of anti-VEGF agents such as Bevacizumab, Ranibizumab, and Aflibercept may be used for regression of the abnormal new vessels, which have formed in the eye;
  - sometimes cryotherapy is used as a treatment for retinal tears and retinal detachments.

— *Restore the media transparency.*
  - Systemic and topical use of hemostatic and vasoprotective drugs to prevent new cases of bleeding for 2—3 days:
    - instillation of Calcium chloride 3 %, solution of Glucose with Ascorbic acid, Pilocarpine 1 % twice daily;
    - parabulbar injections of Etamsylate 12.5 % and Dexamethasone;
    - systemic use of vitamins C and K, Etamsylate in tablets and intramuscular injections, Calcium gluconate 10 % intramuscular injections.
  - Absorptive therapy after 2—3 days:
    - instillation of Potassium iodide 2 %, Lidasa;
    - intravenous or subconjunctival injections of Fibrinolysin or Heparin;
    - physiotherapy is indicated: electrophoresis with lidasa and aloe, phonophoresis.
  - Vitrectomy will be required if hemorrhage is not absorbed after 3 months.

Vitrectomy is a microsurgical procedure that involves removal of the vitreous body and its replacement with Ringer's solution, gas, or silicone oil.

There are two types of vitrectomy — anterior vitrectomy and pars plana vitrectomy. These names are based on the region of the eye, where the procedure is performed.

Anterior vitrectomy refers to the procedure where only small portions of the anterior vitreous are removed. This procedure is performed in case of a traumatic cataract removal and secondary intraocular lens placement.

Pars plana vitrectomy (PPV) refers to a group of procedures that involve the deeper part of the eye. During this procedure, almost all of the vitreous fluid is removed. It is carried out in cases of unabsorbed vitreous hemorrhage; tractional retinal detachment; removal of intravitreal displaced lenses or foreign bodies; severe postoperative or post-traumatic inflammatory vitreous changes.

The procedure is done in an operating room, usually under local anesthesia with sedation or occasionally under general anesthesia. The surgeon uses an operating microscope and contact lenses placed on the corneal surface, which allows a clear view of the vitreous cavity and retina at various magnifications.

Three incisions are made through the pars plana located just behind the iris but in front of the retina, where cannulas are placed, through which instruments can be inserted into the vitreous cavity. They are:

— *a vitrector*, or cutting device, that cuts the vitreous fluid into tiny bits and aspirates it;

— *an infusion line* that supplies a fluid replacing the vitreous being removed, in order to maintain constant eye pressure;
— *an endo-illuminator*, a light source, which lights up the interior of the eye in order to give a clear view as the surgery is being performed.

After removing the vitreous opacities, the retina can be treated intraoperatively if there are any problems with it such as detachment, tears or holes. At the end of the surgery, silicone oil or a gas bubble is injected into the eye to press slightly the retina against the wall of the eye. Oil cannot be absorbed by the body, so, if a silicone oil is used, it must be removed during the second operation after the retinal detachment has healed.

After the procedure is finished, the cannulas are removed and the tiny openings in the sclera are closed with self-absorbing stitches.

Prognosis depends both on the underlying cause and extent of hemorrhage. For example, patients with vitreous hemorrhage resulting from posterior vitreous detachment will have better prognosis compared to those with vitreous hemorrhage secondary to proliferative diabetic retinopathy or age-related macular degeneration. The prognosis in penetrating eye injury is often poor.

Complications. Complicated cataract, retinal atrophy, retinal detachment, vision loss.

Prophylaxis. Primary prevention should be directed at controlling risk factors for systemic vascular diseases such as diabetes, hypertension, and smoking. Proper eye protection should be provided during activities likely to cause an eye trauma.

4.5. Vitreous Inflammation

Since the vitreous has no blood vessels or nerves, there are no primary inflammatory processes in it. *Vitritis* is a medical term used to denote infiltration of inflammatory cells or exudates into the vitreous body. Thus vitritis is actually a symptom of an inflammatory disease occurring in some adjacent structures of the eye, for example, iridocyclitis, choroiditis, panuveitis and endophthalmitis. In some cases, vitritis can develop secondary to systemic inflammatory conditions such as sarcoidosis, Wegener's granulomatosis, systemic lupus erythematosus, toxoplasmosis, and syphilis.

Patients complain of floaters, blurred vision, decrease of visual acuity. Examination will reveal infiltration of the vitreous body by inflammatory cells (often mobile), obscure fundus.

Vitritis can be sight-threating, so it requires intensive topical and systemic treatment. The outcome of treatment depends on the underlying cause.

Review:

1. Key Points

Diseases of the vitreous according to the origin may be *developmental* or *acquired;* according to the pathogenesis — *age-related, degenerative, dystrophic, inflammatory, traumatic.*

The main symptoms of vitreous diseases are floaters, flashes, cloudy vision, visual loss. The main examination methods are slit-lamp exam, direct and indirect ophthalmoscopy, ultrasonography.

Developmental vitreous body abnormalities include persistent hyperplastic primary vitreous, persistent hyaloid artery. *Age-related vitreous body changes* — vitreous liquefaction, posterior vitreous detachment. *Vitreous opacities* — muscae volitantes, asteroid hyalosis, synchysis scintillans, red cell opacities, inflammatory cell opacities, tumor cell opacities, vitreous hemorrhage.

Age-related vitreous disorders and most cases of opacities do not require treatment unless they cause vision impairment. Vitreous hemorrhages require immediate treatment for vision loss prevention. Vitreolysis may be performed to remove large deposits. In severe cases vitrectomy may be performed.

2. Diagrams

DISEASES OF THE VITREOUS
Developmental Abnormalities
Persistent Hyperplastic Primary Vitreous
Persistent Hyaloid Artery
Acquired Disorders
Age-Related Vitreous Body Changes
Vitreous Liquefaction
Posterior Vitreous Detachment
Vitreous Opacities
MuscaeVolitantes
Asteroid Hyalosis
Synchysis Scintillans
Vitreous Hemorrhage
Vitritis

3. The Review Questions

A. Control Questions

1. What is the classification of vitreous body diseases?
2. What are the main symptoms of vitreous disorders?
3. What are the age-related changes of the vitreous body and their symptoms?
4. What developmental abnormalities of the vitreous do you know?
5. What types of vitreous opacities and their main symptoms do you know?
6. What diseases may cause vitreous hemorrhage and are the main treatment methods in this case?

B. Tests

1. The main symptoms typical for vitreous diseases are:
A. Ocular pain
B. Floaters
C. Flashes
D. Photophobia
E. Cloudy vision
F. Decrease of vision

2. The main diagnostic methods for vitreous disorders are:
A. VA
B. Slit-lamp exam
C. Ophthalmoscopy
D. CT scan
E. Tonometry
F. B-scan ultrasonography

3. What are the complications of developmental vitreous body disorders?
A. Posterior vitreous detachment
B. Glaucoma
C. Retinal detachment
D. Vitreous hemorrhages
E. Amblyopia
F. All the answers are correct

4. Conditions that may cause vitreous liquefaction are:
A. High myopia
B. Cataract
C. Blunt or penetrating trauma
D. Radiation burns
E. Acute conjunctivitis
F. Uveitis

5. Floaters can be seen in all of the following cases except:
A. Vitreous hemorrhage
B. Posterior vitreous detachment
C. Vitreous deposits
D. Vitreous degeneration
E. Vitreous liquefaction
F. Acute glaucoma

6. What are the frequent causes of vitreous hemorrhage?
A. Diabetic retinopathy
B. Retinal vein occlusion
C. Blunt or penetrating eye trauma
D. Posterior vitreous detachment with or without retinal tears
E. Hypertensive retinopathy
F. Age-related macular degeneration

7. **Treatment methods for vitreous hemorrhages are:**
   A. Bed rest with the head elevated by 30—45°
   B. Laser photocoagulation of retinal disorders
   C. Cryotherapy
   D. Hemostatic and vasoprotective therapy
   E. Absorptive therapy
   F. Vitrectomy

C. Clinical Cases

Case 1

A mother referred to an ophthalmologist because she noticed white eye reflex in the left eye of her 2-year-old daughter, the girl's left eye is smaller compared to the right one, and poor vision. Objectively: leukocoria, microphthalmia, cataract, microphakia, strabismus, poor vision. What is the diagnosis?

Case 2

A 61-year-old woman with a history of ocular trauma complains of floaters, haziness, and decrease of vision in the right eye. Ophthalmoscopic examination reveals that the vitreous is filled with golden reflective crystals. What is the diagnosis?

Case 3

A 25-year-old male with diabetes complains of sudden painless loss of vision in one eye for the past 2 weeks. There is no history of trauma. On examination the anterior segment is normal, but there is no fundal glow. What is the most likely cause of this condition?

CHAPTER 16

Ocular Trauma

OBJECTIVES

Upon completion of the chapter the students should:

— know the classification of ocular trauma;
— know the basic methods of examination in ocular trauma;
— know the main clinical symptoms of traumatic damage to the ocular adnexa and the globe;
— describe the clinical picture of non-penetrating, penetrating, and perforating globe injury, blunt eye trauma, orbital fracture, trauma to the eyelids, chemical, thermal, and radiation burns;
— evaluate and manage patients with a superficial corneal or conjunctival foreign body, corneal and scleral penetrating injury, laceration of the eyelids, blunt orbital trauma, chemical and thermal burns;
— be able to render first aid to patients with non-penetrating, penetrating, blunt trauma of the eyeglobe; know the principles of suturing of eyelid laceration;
— know the algorithm of urgent aid in thermal and chemical burns of the eye.

Plan:

1. CLASSIFICATION OF OCULAR TRAUMA
2. SYMPTOMS OF OCULAR TRAUMA
3. EXAMINATION METHODS
4. OCULAR TRAUMA
   - **4.1. Mechanical Trauma to the Eyeglobe**
     - Contusion
     - Non-Penetrating Eyeball Injuries
       - Superficial Foreign Bodies
     - Penetrating Eyeball Injuries
     - Perforating Eyeball Injuries
     - Intraocular Foreign Bodies
   - **4.2. Injuries of the Ocular Adnexa**
     - Eyelid Hematoma
     - Eyelid Laceration
     - Orbital Blow-out Fracture
   - **4.3. Burns**
     - Chemical Burns
     - Thermal Burns
     - Radiation Burns
       - Ultraviolet Burns
       - Ionizing Radiation Burns
       - Laser Radiation Injuries
       - Infrared Burns

1. Classification of Ocular Trauma

Ocular injuries can be classified:
— according to the causative conditions — industrial, car accident, agricultural, domestic, children's, sports-related, military;
— according to the location — injuries to the eyeglobe or ocular adnexa;
— according to the mechanism of injury — mechanical (contact) or burns;
— according to the extent of the mechanical trauma — contusion (blunt trauma), non-penetrating, penetrating, or perforating injuries;
— according to the cause of burn — chemical, thermal, radiation;
— according to the grade of damage or visual outcome — mild, moderate, severe, very severe.

Mild grade — the trauma doesn't cause any risk of visual impairment.
Moderate grade — the trauma causes the risk of visual impairment.
Severe grade — the trauma causes the risk of total visual loss.
Very severe grade — the trauma causes the risk of eyeball loss.

The grading of trauma severity is quite conditional, because it is difficult to predict the course of the traumatic process and all possible post-traumatic complications.

In clinical practice, the ocular trauma classification system (OTCS), based on the Birmingham Eye Trauma Terminology System (BETTS), is used for mechanical trauma for the purpose of treatment measure differentiation. According to this classification mechanical ocular traumas are subdivided into closed and open globe injuries, which are characterized by four parameters — type, grade, presence or absence of relative afferent pupillary defect (RAPD), and extend of the injury (table 16.1).

With the use of this classification system ophthalmologists can collect important data at the time of initial examination without the use of specialized equipment or testing. These data help to determine appropriate clinical examination methods and management and can be prognostic of final visual outcome.

This classification system also standardizes and provides unambiguous definition of terms and has become an international language of ocular trauma terminology (table 16.2).

Table 16.1

Classification of Mechanical Injuries of the Eye According to the Ocular Trauma Classification System (OTCS)

Parameter	Closed Globe Injury	Open Globe Injury
Type (mechanism of injury)	— contusion — lamellar laceration — superficial foreign body — mixed	— laceration • penetrating injury • perforating injury • intraocular foreign body (IOFB) — rupture — mixed
Grade (visual acuity at initial examination)	≥ 0.5 0.4—0.2 0.19—0.025 0.02 — to light perception No light perception	≥ 0.5 0.4—0.2 0.19—0.025 0.02 — to light perception No light perception
Pupil (presence or absence of RAPD)	RAPD (+) RAPD (–)	RAPD (+) RAPD (–)
Zone (wound location)	The external segment (the bulbar conjunctiva, cornea, sclera). The anterior segment (injury to the structures in the anterior chamber including the lens and zonules). The posterior segment (the vitreous, retina, optic nerve, choroid, and ciliary body)	Limited to the cornea, corneo-scleral (within 5 mm of the limbus), posterior scleral (posterior to 5 mm from the limbus)

Table 16.2

Definition of Terms According to the Birmingham Eye Trauma Terminology System (BETTS)

Terms	Definitions
Eye wall	The outer layer of the globe — the cornea and sclera
Closed globe injury	No full-thickness wound of the eye wall
Contusion	Blunt force injury with no wound of the eye wall
Lamellar laceration	Partial-thickness wound of the eye wall caused by a sharp object
Superficial foreign body	One or more foreign objects lodged in the conjunctiva and/or eye wall
Open globe injury	Full-thickness wound of the eye wall
Laceration	Full-thickness wound of the eye wall caused by a sharp object
Penetrating injury	An entrance wound is present
Perforating injury	Both an entrance and an exit wound, caused by the same agent, are present
Intraocular foreign body	One or more foreign objects retained inside the eye
Rupture	Full-thickness wound of the eye wall caused by a blunt object

2. Symptoms of Ocular Trauma

Symptoms of ocular trauma may differ a lot depending on the affected eye structure and severity of the injury. Sometimes patients will complain only of the appearance and do not notice any changes in their visual functions (e.g. black eye). In cases of severe ocular injury such as severe contusion, globe rupture, perforating injury with an intraocular foreign body or severe burn the symptoms are intense pain, loss of vision, and loss of the eye itself.

From the standpoint of diagnosis it is very important to differentiate penetration injuries from other types of mechanical ocular trauma, so their signs are divided into absolute and relative:

Absolute, or reliable signs:

— presence of a corneal or scleral wound;
— a hole in the iris;
— prolapse of the intraocular contents (the iris, ciliary body, choroid, vitreous);
— leakage of the intraocular humor — positive Seidel test;
— intraocular foreign body;
— air bleb in the vitreous.

Relative, or suggestive signs:

— the anterior chamber is shallow, or absent, or too deep (shallow in corneal lacerations, deep — in scleral);
— change of the pupil shape (drop-shaped and displaced toward the penetration wound);
— hypotony of the eye;
— punctate lens capsule defect or opacification;
— focal iris-corneal adhesion;
— bleeding in the anterior chamber (hyphema) and vitreous body (hemophthalmos);
— marked conjunctival edema (chemosis) or subconjunctival hemorrhage.

3. Examination Methods

Methods of examination differ according to the type of the injury, its location, character, and severity and may include:

Ophthalmological methods:

- History of the trauma.
- External inspection.
- Eye movements.
- Pupil reaction to light.
- Slit-lamp exam.
- Eyelid eversion.
- Ophthalmoscopy.
- Perimetry.

Investigative methods:

- B-scan ultrasonography.
- CT scan.
- X-ray, plain and with a prosthesis-localizer.
- X-ray by Fogt.

4. Ocular Trauma

Ocular trauma is a leading cause of preventable monocular blindness worldwide and is a serious public health concern in developed and developing countries. According to the data of the World Health Organization (WHO) approximately 55 million eye injuries occur yearly, of which 750 000 people require hospitalization, 1.6 million people are blind from eye injuries, 2.3 million are bilaterally visually impaired, and 19 million have unilateral visual loss.

The age distribution for the occurrence of serious ocular trauma is bimodal, with the

EYE FACTS

Eye injuries contribute 20 % of all eye pathology.

Eye injuries in > 75 % cases are a cause of monocular blindness.

Eye injuries in 70 % cases affect population of < 30 years of age, 80 % males.

Eye injuries are the most common cause of enucleation.

Eye injuries in 90 % cases are preventable.

maximum incidence in young adults and a second peak in the elderly. So, in addition to the impact on affected individuals there are profound social implications regarding the productivity lost by young men and the requirement for caring facilities and rehabilitation for the elderly.

The spectrum of injuries ranges from very mild, non-sight-threatening to extremely serious with potentially blinding consequences. Ocular injuries may also be associated with other injuries including facial fractures, in this case a severe visual impairment may occur.

Objective examination of patients with eye injury, proper diagnostics, prompt and appropriate management may prevent serious vision impairment or loss of the eye. Following this, the actuality of the subject for the doctors of different specialization is beyond any doubt. It is important that every general practitioner and health care staff member is able to recognize an ocular injury and provide initial treatment.

4.1. Mechanical Trauma to the Eyeglobe

Contusion

Definition. Contusion is a blunt force injury of the globe without the damage of the eye wall, instead affecting the inner structures of the eye.

Etiology. Contusion can be caused by impact of an object with a low velocity and a relatively big area (a fist, a ball, a stone, a champagne cork, etc.) to the eye and surrounding tissues. Contusion can be direct — when an object hits the eye directly; indirect — due to a blow to the surrounding structures of the face or shaking of the body and facial skeleton (may be due to explosions); or mixed, which occurs in case of combined effects of these traumatic factors.

Clinical Picture. When the eye is struck, it is compressed antero-posteriorly and correspondingly stretched in the equatorial plane, which causes a combination of contusional and tearing damage. The character and severity of contusion depends on the intensity and direction of the impact, individual features of the eye structures, patient's age, previous ocular history, etc. This may result in a spectrum of injuries ranging from a simple "black eye" to severe intra-ocular disruption, including orbital fracture and rupture of the globe (fig. 16.1).

The most common finding of mild contusion is ***subconjunctival hemorrhage*** (bleeding from the conjunctival vessels between the bulbar conjunctiva and the sclera) (fig. 16.2). It can be caused by severe cough, vomiting, lifting of heavy objects, blood hypertension or may accompany a blunt trauma to the eyelids, orbital soft tissues or nasal sinuses.

Blunt trauma to the globe may cause ***corneal damage***, such as corneal abrasion, corneal edema, corneal endothelial breaks. Blood staining of the cornea (***hematocornea***) may arise from endothelial breaks in the presence of hyphema in the anterior

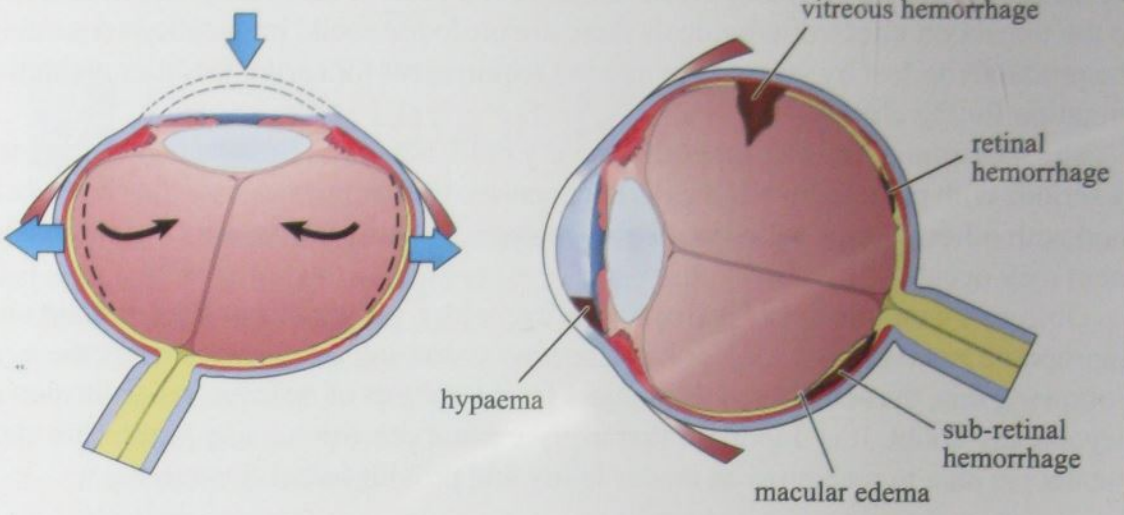


Fig. 16.1. Possible clinical presentations of blunt eye trauma

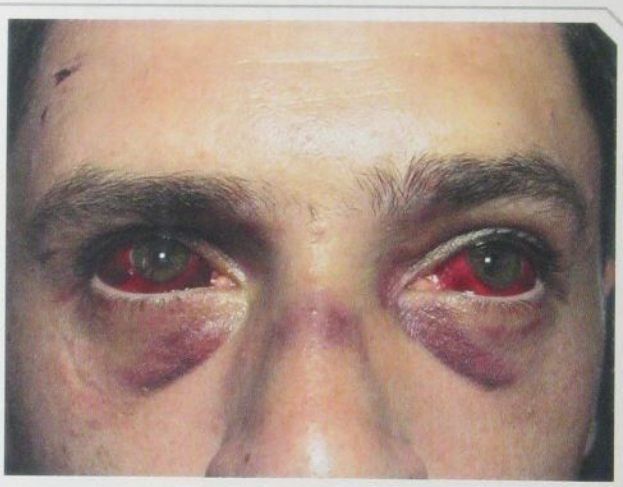


Fig. 16.2. Subconjunctival hemorrhages of both eyes (spectacle hematoma)

chamber and elevation of intraocular pressure. In this condition the cornea becomes reddish-brown, then greenish-yellow, then white-grey. Very rarely a blunt trauma may lead to corneal rupture.

Damage of the sclera in blunt trauma may range from mild mixed injection (hyperemia due to dilation of the episcleral and conjunctival vessels) to dramatic ruptures of the sclera that typically occur in the weakest parts — 3—4 mm parallel to limbus in the superior-medial and superior-lateral quadrants. Severe swelling of the conjunctiva (red chemosis) may point at subconjunctival scleral rupture (fig. 16.3).

In blunt trauma, the cornea is flattened, the anterior chamber is compressed and this causes the aqueous humor to force peripherally, leading to ***damage to the drainage angle***. This results in a ***rapid rise of intraocular pressure***, which, if extensive enough, predisposes to glaucoma later.

Compression of the anterior chamber forces the pupil to dilate rapidly, which may break fibres of the sphincter pupillae muscle, causing ***traumatic mydriasis***. Irritation of the ciliary nerves leads to ***traumatic miosis*** often associated with ***spasm of accommodation***. The tension on the peripheral iris may tear it from its root partially (***iridodialysis***) (fig. 16.4) or completely (***aniridia***).

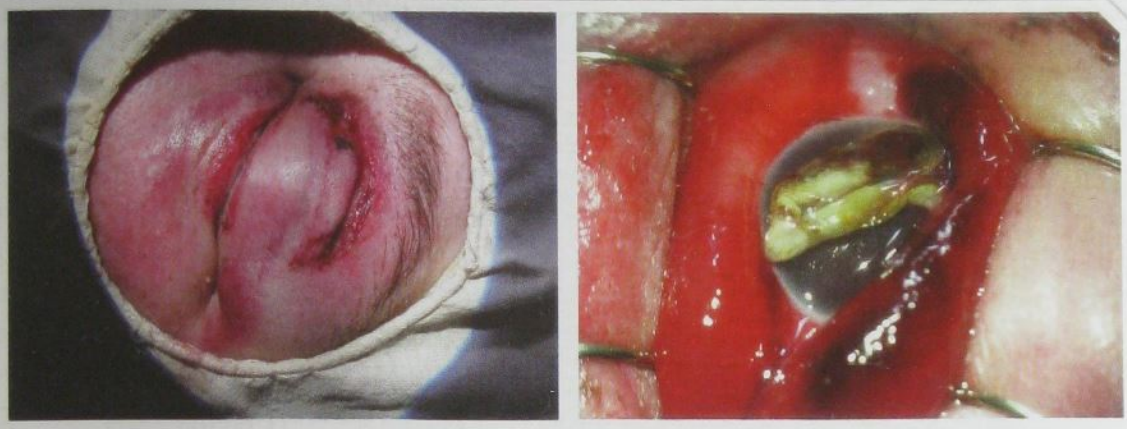


Fig. 16.3. Severe contusion with eyelid hematoma and laceration, rupture of the eyeball and prolapse of the internal structures

Bleeding from damaged vessels of the iris and ciliary body causes a ***hyphema*** (accumulation of blood in the anterior chamber forming a horizontal level that moves according to the patient's head position) (fig. 16.5). It can be partial or total. Damage to the iris and ciliary body occurs in 60 % cases of blunt trauma.

NOTE

Hyphema may be the hallmark of severe damage of the intraocular structures in blunt trauma.

The supporting lens zonules may rupture partially or completely causing ***lens subluxation*** or ***complete dislocation***, respectively. The lens usually dislocates posteriorly into the vitreous cavity, but may be found in the anterior chamber (fig. 16.6). Blunt trauma can also cause changes in the lens structure resulting in ***concussion cataract*** or ***Vossius ring*** (a 2—3 mm pigmented ring like an iris imprint on the anterior lens capsule).

Stretching of the choroid causes a ***choroidal rupture***, which appears as ***subretinal hemorrhage***. In the posterior segment, the vitreous is firmly attached in the re-

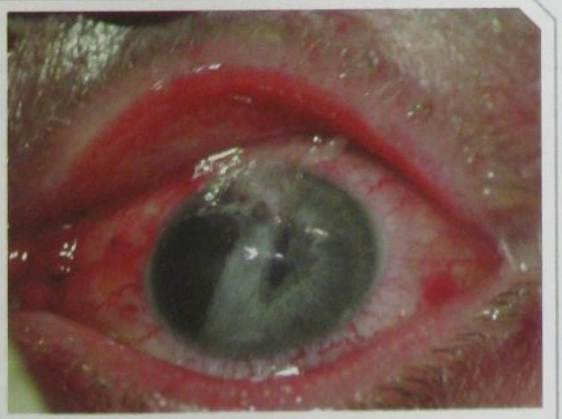


Fig. 16.4. Iridodialysis

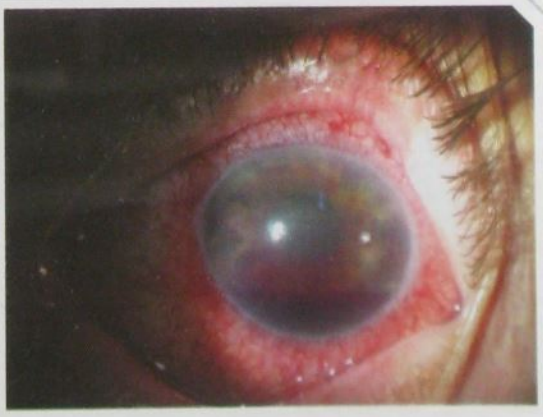


Fig. 16.5. Hyphema

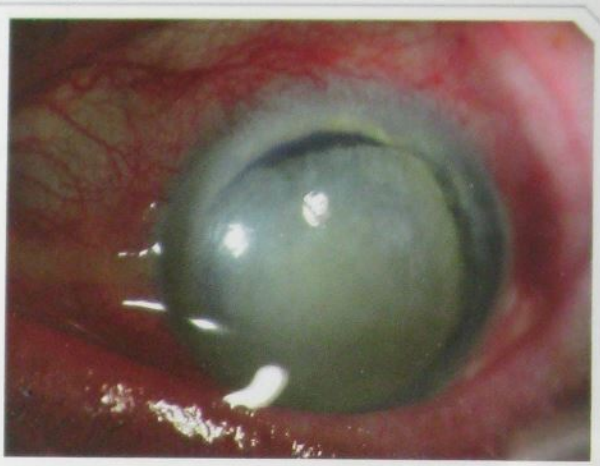


Fig. 16.6. Traumatic cataract and lens dislocation into the anterior chamber

gion of the ora serrata, and stress on this area causes disinsertion of the peripheral retina (***retinal dialysis***), which will result in ***retinal detachment*** (fig. 16.7). Other types of retinal damages may include ***macular edema*** (***commotio retinae*** or ***Berlin's edema***), ***retinal tears***, ***retinal hemorrhages***. Any damage in the posterior segment may result in ***vitreous hemorrhage***, usually due to a uveal tract rupture, retinal blood vessels injury, retinal tears.

Contusion may lead to ***fracture of the floor or medial wall of the orbit*** due to a sudden increase in intraorbital pressure. Orbital fracture can result in prolapse of the orbital fat and oculomotor muscles in the fracture defect. The eye may recede into the orbit (***enophthalmos***) and the palpebral fissure may narrow. The displacement of the orbital bones and retrobulbar bleeding cause the globe to protrude (***exophthalmos*** or ***proptosis***).

Orbital fracture may be accompanied by ***optic nerve trauma***: incarceration between bone pieces, rupture at different levels or even separation of the optic nerve from the globe.

Complaints. Contusions may present with a wide range of symptoms, from minimal discomfort to severe pain and loss of vision depending on the type and severity of the condition.

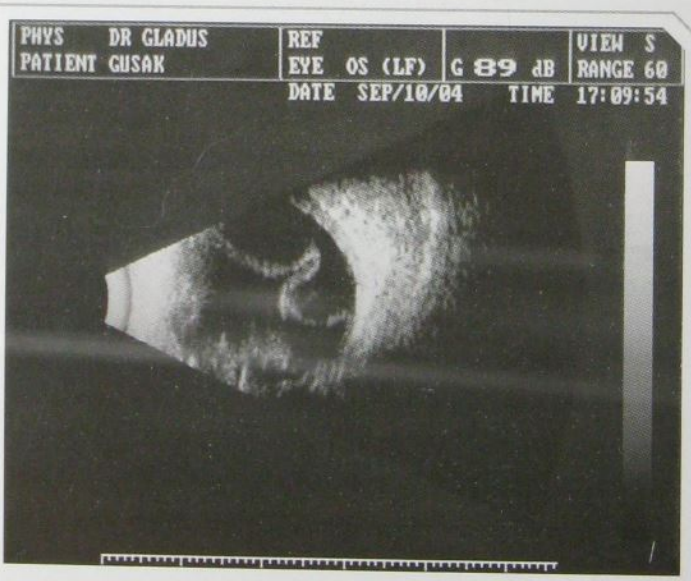


Fig. 16.7. Retinal detachment detected at B-scan examination

In subconjunctival hemorrhages the condition is usually painless and vision is not affected. The only complaint is a red and sore eye.

In corneal damage patients complain of acute ocular pain, foreign body sensation, photophobia, tearing, blepharospasm, and decreased vision.

Other complaints as a result of intraocular damage may include blurred vision, ocular pain ranging from mild discomfort to severe deep dull pain, a mild to mod-

erate decrease of visual acuity, restriction and pain on eye movements, diplopia, halos around lights, flashes, floaters, defects of visual fields and even no light perception.

Signs. Ophthalmologic findings depend on the ocular structures damaged in case of blunt trauma and may be as following:

— eyelids — swelling and hematoma, laceration of the eyelids;
— conjunctiva — subconjunctival hemorrhage, usually flat, that may enlarge within a few days, chemosis;
— cornea — corneal abrasions, folds in Descemet's membrane, deep stromal edema, lamellar corneal laceration, blood staining of the cornea, corneal rupture;
— sclera — partial or full rupture that results in hypotony, hemophthalmos, prolapse of the inner structures (lens, iris, choroid);
— anterior chamber — exudates, hyphema, recession of the angle, elevated IOP, irregularity in the anterior chamber depth due to cornea and/or sclera rupture;
— iris and ciliary body — traumatic miosis, traumatic midriasis, irregularity of the pupillary shape, radiating lacerations, iridodialysis, aniridia, cyclodialysis;
— lens — Vossius ring, concussion cataract, subluxation or dislocation of the lens;
— choroid — choroidal rupture, subretinal hemorrhage;
— retina — Berlin's edema (commotio retinae), retinal hemorrhages, retinal tears, retinal detachment;
— vitreous — liquefaction, vitreous opacities, vitreous detachment, hemophthalmos, vitreous loss in cases of globe rupture;
— optic nerve — optic disc edema, optic disc pallor, atrophy of the optic nerve, avulsion.

Methods of Examination. Thorough trauma history, external inspection, VA test, pupils shape and pupillary responses, extraocular movements, slit-lamp exam, corneal staining, gonioscopy, ophthalmoscopy, perimetry, ultrasonography, CT scan, X-ray.

Differential Diagnosis. Other cases of acute red eye, corneal abrasion, lamellar laceration, penetrating injury, perforating injury, intraocular foreign body.

Treatment options depend on the type and severity of the damage and eye structure affected:

— subconjunctival hemorrhage is typically a self-limiting condition that requires no treatment in the absence of infection or associated significant trauma. Severe swelling of the conjunctiva (red chemosis) may point at subconjunctival scleral rupture, in this case conjunctival incision, hemorrhage drainage with a revision of the sclera may be needed;

NOTE

Detailed trauma history is very important, and as much information about the details of the injury should be obtained as possible. The history related to the injury may provide clues to the nature of the ocular injury, assist in appropriate diagnosing and help to avoid missing sight-threatening conditions.

The clinical examination should be as complete as possible, but any further injury to the globe should be avoided.

— corneal abrasion is treated with administration of topical antiseptics or antibiotics (Ofloxacin, Ciprofloxacin, Norfloxacin) and epithelizing agents (Solcoseryl, Dexpanthenol) 4—6 times daily. To decrease pain a bandage contact lens and oral pain medicine may be prescribed;
— hyphema — a small hyphema usually resolves within a few days with no treatment. A large or total hyphema requires strict bed rest with the head raised at 30—45 degrees, hemostatic therapy (Fibrinolysin, Gemaza, Dicynone), and even a binocular eye bandage initially to prevent the recurrent bleeding. With no effect after 4—5 days anterior chamber paracentesis and blood draining are indicated. The use of corticosteroids and cycloplegics is controversial;
— subluxation and luxation of the lens is treated surgically by intracapsular lens extraction;
— traumatic miosis usually resolves within few days without specific treatment;
— traumatic midriasis can be treated with myotics (Pilocarpine 1 %) for 2 weeks, a soft cosmetic contact lens, or if resistant — plastic iris surgery to form a "normal" pupil;
— iridodialysis is treated with surgical closure of the defect not earlier than 2—3 months after the trauma;
— aniridia can be managed with wearing of cosmetic colored contact lenses, surgical treatment of aniridia with iris implants bears a high risk of different complications;
— cyclodialysis is treated with conservative medical therapy (topical cycloplegics and corticosteroids), but when this fails, a wide range of laser and surgical procedures (transscleral photocoagulation, transconjunctival cryotherapy) have been reported to be effective;
— choroid rupture is treated by hemostatic, anti-inflammatory, resorptive therapy; in late terms it requires photocoagulation to prevent retinal detachment;
— commotio retinae (Berlin's edema) does not need treatment, it gradually resolves spontaneously in few days;
— retinal hemorrhages are treated with angioprotective and resorptive therapy, osmotherapy, vitamins, and corticosteroids;
— retinal tears require photocoagulation;
— retinal detachment is treated surgically;
— hemophthalmos is treated by hemostatic and resorptive therapy (Fibrinolysin, Gemaza, Dicynone, Lidaza, Aminocaproic acid), total hemophthalmos requires vitrectomy;
— scleral rupture requires precise suturing of the scleral edges with setting in or excising if the prolapsed structures are necrotized.

Prognosis. Final visual outcome of ocular contusion depends on prompt appropriate diagnosis and treatment, as well as severity of the condition. Mild contusions have a good prognosis. Severe blunt trauma leads to the development of complications that may result in a decrease or even loss of vision.

Complications. Corneal opacities, traumatic cataract, secondary glaucoma, retinal detachment, loss of vision, loss of the eye.

Prophylaxis. Most eye contusions can be prevented with safety glasses or other forms of eye protection.

Non-Penetrating Eyeball Injuries

Non-penetrating or closed globe injuries refer to superficial traumas that do not cause a full-thickness damage to the eye wall (cornea or sclera). As a rule, such injury does not create much risk for complications and visual impairment.

Common conditions associated with non-penetrating trauma are corneal erosion and superficial foreign body.

Corneal erosion, also referred to as corneal abrasion or scratched eye, is the damage of the corneal epithelium that is a very common presentation of ocular emergency care (about 10 % of cases). It can happen in an instant, e.g. a person may scratch the eye with a fingernail, damage it while applying make-up, cut with paper, scratch with a tree branch, or the eye may be scratched by an animal, damaged with a cigarette, injured with inverted eyelashes, there may be a blunt trauma, also a person might rub the eye with a foreign body, such as dust, sand, an eyelash or a contact lens, inside.

Corneal erosion is a quite painful condition as the cornea has many nerve endings just under the surface. Other symptoms may include tearing, sensitivity to light, blepharospasm, irritation, foreign body sensation, and blurry vision.

Early diagnosis and treatment will help to avoid all future complications and make recovery faster. Most corneal abrasions usually heal within 2 days. If left untreated, many corneal abrasions can cause a secondary infection, or even a corneal ulcer, which can leave permanent scarring. *(Read more about the etiology, clinical picture, signs, and methods of treatment for* ***corneal erosion*** *in the Chapter* ***Diseases of the Cornea, p. 257****).*

Superficial Foreign Bodies

Definition. Superficial foreign bodies refer to any objects that are adherent to or embedded in the corneal epithelium and/or the conjunctiva. It is a common presentation to the emergency department and accounts for approximately 35 % of all eye injuries seen.

Etiology. Usually, the cause is accidental trauma that may occur in windy weather or either at work or while doing household work (hammering, grinding, gardening, or DIY.

The most common foreign bodies in the conjunctiva and cornea include the following: dirt, sand, caterpillar hairs, small insects, plant particles that are frequently blown into the eye and adhere to the superficial cornea or conjunctiva. Other common foreign bodies are eyelashes, cosmetics or pieces of metal, glass, plastic, wood when working with tools.

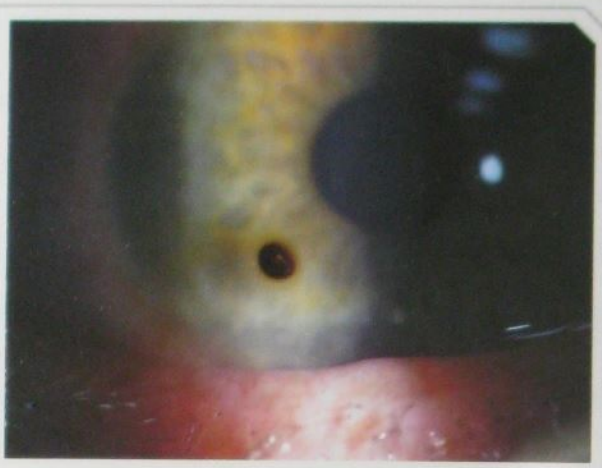


Fig. 16.8. Corneal metal foreign body with signs of infiltration and metallosis

Clinical Picture. The condition is usually unilateral. Foreign bodies may be single or multiple and are easily seen without magnification or barely detectable with slit lamp examination. A foreign body may be revealed on or in the cornea or on the conjunctival surface in the posterior third of the eyelid fornix (fig. 16.8).

The onset of pain occurs either immediately after the injury or within the first twenty-four hours. Typically, there is a sensation of something in the eye, pain, and photophobia. The pain is aggravated by blinking or moving the eye. Vision may be affected if the foreign body is in the visual axis.

The patients sometimes may be asymptomatic if the foreign body is below the epithelial or conjunctival surface. Over a period of a few days, the epithelium often grows over small corneal foreign bodies, with a resultant reduction in pain.

Complaints. Patients may complain of acute pain, foreign body sensation, continuous irritation, discomfort with blinking, blepharospasm, photophobia, tearing, red eye and blurry vision.

Signs of the superficial foreign body are normal or decreased VA, conjunctival or ciliary injection, a visible foreign body, a rust ring (if a metallic foreign body has been embedded for hours or days) or infiltration around the foreign body, an epithelial defect that stains with fluorescein, corneal or conjunctival edema.

Methods of Examination. Thorough trauma history, external ocular inspection, slit lamp exam, corneal staining, eversion of the upper eyelid, ultrasonography and CT scan may be indicated if there is any suspicion of a penetrating intraocular foreign body.

Differential Diagnosis. Bacterial keratitis, acute conjunctivitis, corneal erosion, recurrent erosion syndrome, intraocular foreign body.

Treatment. All foreign bodies of the cornea and conjunctiva should be removed as soon as possible or they may be complicated by posttraumatic keratitis or corneal ulcer.

— *A foreign body from the conjunctiva* can be removed in out-patient conditions even without topical anesthesia, but it is more comfortable if the eye is anesthetized. It is important to evert the upper eyelid to examine the superior fornix and tarsal plate. It is done with the help of cotton swab, a pad or a clean soft tissue (fig. 16.9). Foreign bodies laid superficially can also be irrigated with sterile water or saline solution.

NOTE

To reduce the risk of corneal penetration, ensure that the needle approaches the cornea tangentially.

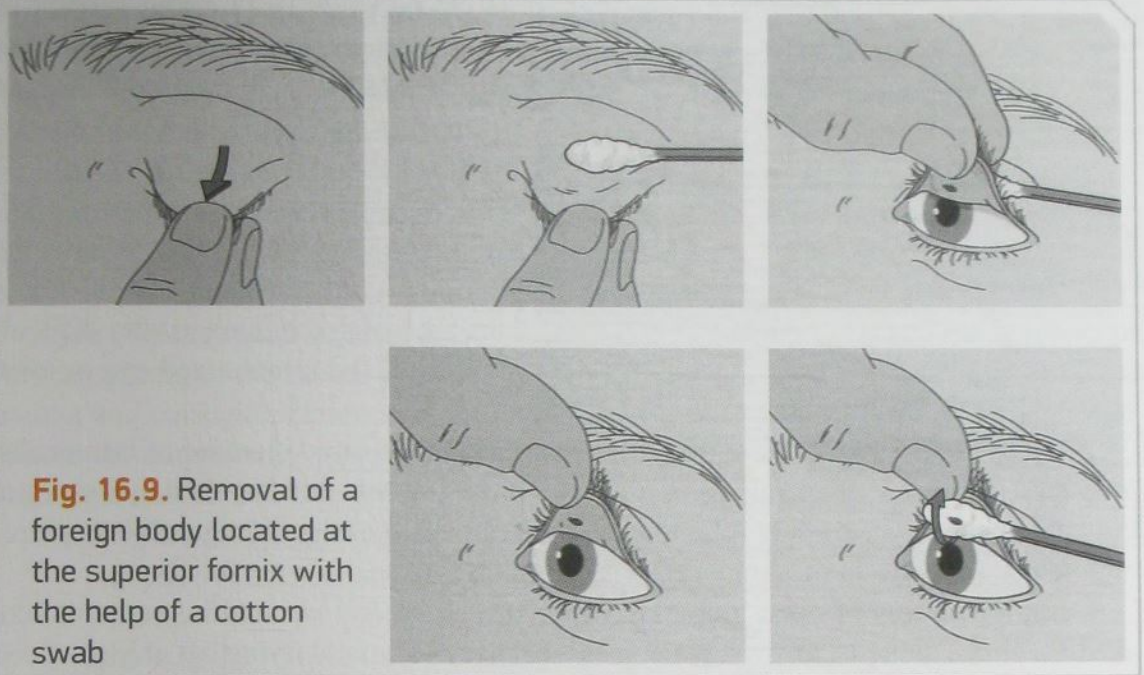


Fig. 16.9. Removal of a foreign body located at the superior fornix with the help of a cotton swab

— If the *foreign body is embedded in the corneal epithelium*, it is removed with the use of a sterile 25-gauge needle, a foreign body spud or a special bur after topical anesthesia (Proxymetacain 0.5 %, Tetracaine 0.5 %, Dicain 0.3 %).

— An *iron-containing foreign body* can be removed with a magnet. It should be removed under the control of a slit-lamp; for the safety of the manipulation the patient's head must be tightly pressed against the slit-lamp bar. If the rust ring remains after removal, the cornea should be cleaned from all little particles using a bur to prevent infection and improve epithelization.

NOTE

Never give a topical anesthetic to the patient for repeated use as this delays corneal healing and can lead to permanent scarring.

Steroids should be avoided while epithelial defect exists.

— *After foreign body removal the eye should be treated for abrasion* with a disinfectant solution (Sulfacetamide, Decamethoxin, Myramistin), antibiotic drops or ointments (Ciprofloxacin, Ofloxacin, Norfloxacin, Levofloxacin, Gentamicin, Tetracycline), and re-epithelization drugs (Carbomer, Dexpanthenol, Solcoseryl, Actovegin) for 5—7 days with an aseptic eye bandage for 24 hours.

— *Topical cycloplegics* can be considered for pain and photophobia.

Prognosis. Prognosis for vision is usually good unless a rust ring or scarring involves the visual axis. If infection develops, prognosis is more guarded.

Complications. Infrequent complications include infection and/or tissue necrosis, or scar formation. Sometimes a foreign body can extend through the entire cornea into the anterior chamber.

Prophylaxis. All superficial foreign bodies may be prevented by wearing protective eyewear (e.g., while doing sports, working at construction, in workshops, and other types of environment that have a high risk of particles or objects flying into the eyes).

Penetrating Eyeball Injuries

Definition. Penetrating eyeball injuries refer to open-globe injuries where the traumatic agent cuts all layers of the eye wall. In other words, it is any full-thickness injury to the eyeball with an entrance wound. This type of eye trauma is very difficult since it may cause a dramatic loss of visual functions of the traumatized eye or even seriously affect the other (pair) eye.

Etiology. Full-thickness eye injury may happen due to rapid increase of intraocular pressure (inside-out mechanism) in case of a blunt trauma, this is called eye rupture. Full-thickness injury of the globe may also be caused by sharp foreign objects piercing the eye (outside-in mechanism), it is called laceration.

Common causes of open-globe laceration are sharp and pointed instruments like needles, sticks, pencils, arrows, pens, glass, or pieces of metal flying off at high speed because of metal to metal contact (e.g., hammer and chisel) during the use of tools or vehicle accidents etc. Globe penetration may occur in gunshot and stab wounds.

Clinical Picture. According to the localization of the wound, open globe injuries are divided into corneal, corneo-scleral, and scleral injuries. Corneal laceration is a full-thickness wound limited to the cornea; corneo-scleral laceration is a wound within the limbus with the cornea and sclera involvement; and scleral laceration is a wound extended posteriorly. The edges of the wound can be linear, irregular, stabbed, gaping or adapted or with a tissue defect. If more than one wound is present, each must have been caused by a different agent.

Penetrating injuries may present with subconjuntival hemorrhage, corneal lacerations, hyphema, iris deformities. The iris may plug the wound, resulting in an irregular pupil. Penetrating injuries of the anterior segment may involve the anterior capsule of the lens, causing a localized or, more commonly, a diffuse lenticular opacity. The vast majority of such scleral and corneo-scleral wounds involve underlying structures, which may result in prolapse of the iris or choroid. Posterior wounds involve the retina and cause the development of vitreous hemorrhage, retinal hemorrhage, retinal tears and detachment (fig. 16.10, 16.11).

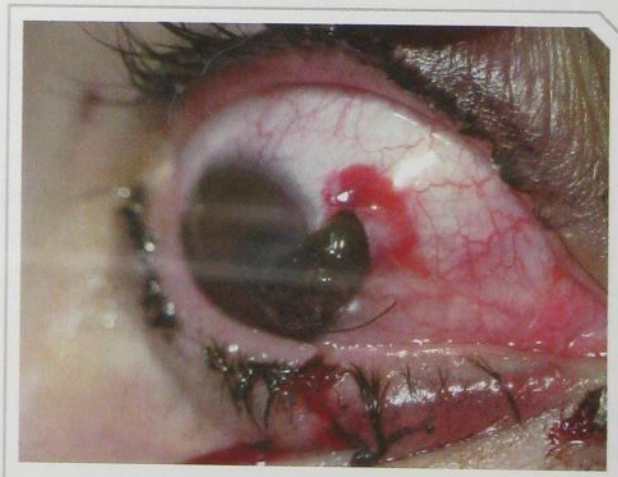


Fig. 16.10. Penetrating scleral injury with prolapse of the iris root

Complaints. Usually patients complain of sharp pain, irritation, tearing, foreign body sensation, blurred vision directly after the trauma. But if a foreign body is small and does not cause much damage to the eye structures, the course may be almost symptomless.

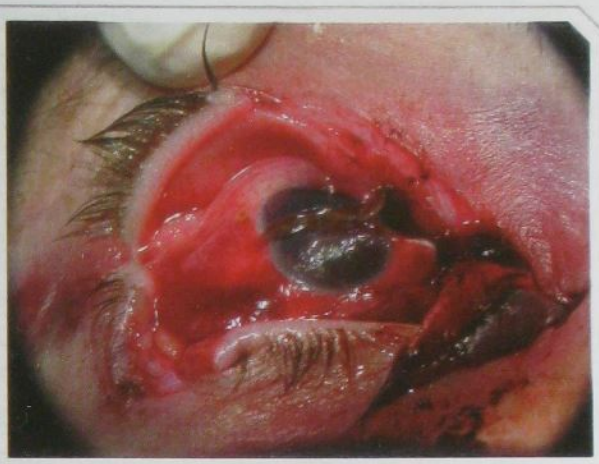


Fig. 16.11. Severe penetrating trauma of the eyeball and eyelid laceration

Signs of penetration injuries may vary according to the severity of the trauma and causative factors. To facilitate the diagnosing, signs are divided into absolute and relative:

1. *Absolute, or reliable signs*:
— presence of corneal or scleral wound;
— a hole in the iris;
— prolapse of the intraocular contents (iris, ciliary body, choroid, vitreous);
— leakage of the intraocular humor — positive Seidel test;
— an intraocular foreign body;
— an air bleb in the vitreous.
2. *Relative, or suggestive signs*:
— the anterior chamber is shallow, or absent, or too deep (shallow — in corneal lacerations, deep — in scleral);
— change of the pupil shape (drop-shaped and displaced toward the penetration wound);
— hypotony of the eye;
— a punctate lens capsule defect or opacification;
— focal iris-corneal adhesion;
— bleeding in the anterior chamber (hyphema) and vitreous body (hemophthalmos);
— marked conjunctival edema (chemosis) or subconjunctival hemorrhage.

Methods of Examination. An accurate anamnesis is important to establish the mechanism of trauma (time, circumstances, the type of traumatic object).

A detailed ocular examination includes VA test, pupillary evaluation, slit-lamp exam, ophthalmoscopy, diaphanoscopy. In cases of minimal penetrating injuries, a Seidel test is performed to reveal leaking of the corneal wound. A local anesthetic (Proxymetacain) may be needed.

NOTE!

If you suspect penetrating injury, avoid any procedures that might apply pressure to the eyeball (tonometry, gonioscopy, etc.).

If penetrating injury is apparent, do not apply diagnostic eyedrops (fluorescein, midriatics).

If absolute signs of penetrating injury are revealed, all other examinations should be delayed until the operating room.

If a retained intraocular foreign body is anticipated, additional methods of diagnostics are mandatory to perform for its localization: ultrasonography, CT scan with axial and coronal cuts, plain X-ray of the orbits in two (frontal and lateral) views, MRI.

Differential Diagnosis. Ocular contusion, corneal abrasion.

Treatment. Penetrating injuries must be treated as soon as possible in order to preserve vision. The most important goal is to prevent further damage.

First aid should be rendered by any medical personnel and consists of:

— *instillation of a topical anesthetic* (Proxymetacain 0.5 %, Tetracaine 0.5 %, Dicain 0.3 %), *disinfectant solutions* (Sulfacetamide 20 % or 30 %, Decamethoxin 0.2 %, Myramistin 0.01 %), and *antibiotic drops* (Ofloxacin, Tobramycin, Levofloxacin, Ciprofloxacin);

— *tetanus prophylaxis* if not current;

— *binocular bandage* (the injured eye — with a protective shield, the uninjured eye — with a pressure patching to prevent excessive movement of the injured eye);

— *systemic sedatives and analgesics* may be needed.

NOTE!

DO NOT try to remove any object, which is penetrating the eye as you can remove intraocular structures protruded through the wound. Removal should be delayed until the operating room.

DO NOT apply pressure when bandaging the injured eye.

After first aid administration, the patient must be transported in supine position (if possible) to the nearest specialized ophthalmic trauma department for surgical treatment.

Ultimate therapy is surgical repair in the operating room under the microscope with the use of microsurgical instruments under local or general anesthesia, with the primary goal to restore the integrity of the globe — optic-reconstructive surgery. Non-absorbable 8/0—10/0 running or interrupted sutures are used to appose the edges of the wound.

Lacerations of the *conjunctiva* longer than 3 mm should be sutured.

A little linear wound of the *cornea* without prolapse of the intraocular structures and humor leakage may seal by itself. Its treatment consists of topical antibiotics and eye bandage. Laceration of more than 2 mm should always be sutured.

Surgical repair of *complicated corneal (or corneo-scleral) injuries* includes several essential steps:

— excision of the anteriorly prolapsed vitreous, lens fragments, transcorneal foreign bodies;

— repositioning of the anteriorly prolapsed uvea, retina;

— stitching of limbal landmarks, watertight closure of the corneal edges of the wound;

— exposure of the scleral component with meticulous revision of the posterior part of laceration;

— excision of the posteriorly prolapsed vitreous;
— repositioning of the posteriorly prolapsed uvea, retina;
— stitching of the scleral wound;
— stitching of the conjunctiva.

In cases with a high risk for endophthalmitis, intraocular antibiotics may be administered during surgery. Depending on the extent of injury, systemic antibiotic treatment (i.v.) may be started as in endophthalmitis. Postoperative management usually consists of cycloplegic, steroid, and antibiotic drops.

A considerable damage to the intraocular structures may require secondary repair such as cataract extraction, glaucoma surgery, vitrectomy, retinal cryo- or photocoagulation.

Prognosis of penetrating eyeglobe injuries is always serious; it depends largely on the extent of damage, accompanying infections, presence of complications.

For example, wounds that are isolated to the cornea without damage to the anterior segment structures may close spontaneously with little visual morbidity, especially if they are off the visual axis. Larger or more complex wounds result in scarring, which itself may be visually disabling.

Complications of penetrating injury include corneal scarring, adhesions between the iris and the angle of the anterior chamber, post-traumatic glaucoma, cataract, retinal detachment, sympathetic ophthalmia, endophthalmitis, panophthalmitis, phthisis bulbi, and loss of the eye.

Prophylaxis. Penetrating eye injuries are serious and can be prevented by the use of proper protective eyewear at work or during sports activities. If an injury does occur, prompt first aid may greatly improve the chances of preserving vision.

Perforating Eyeball Injuries

Definition. Perforating eye injuries are the type of open-globe injuries characterized by the presence of an entrance and an exit wounds caused by the same traumatic object. They are injuries throughout the whole globe.

Etiology. A typical perforating injury is caused by high speed or sharp objects (e.g., BB or shotgun pellets, blades, nails, needles, glass, pens or pencils). Most of the injuries occur during chopping or cutting wood, hammering metals or nails, and carving stone. These are associated with professions such as farming, garage work, and carpentry in adults. Road traffic accidents may cause perforating lacerations of the eye globe with broken glass. Children mostly sustain accidental injuries by needles, scissors, pencils, etc. while playing with others.

Clinical Picture. Depending on the site of perforation, injuries can be divided into corneal, scleral or injuries that perforate across the limbus.

These ocular traumas can cause rupture of the globe, subconjunctival hemorrhage, corneal lacerations, hyphema, iris deformities, lens disruption, or posterior segment findings such as vitreous hemorrhage, retinal tears and retinal detachment. At presence of two openings a dramatic loss of inner structures may occur (posterior prolapse of the vitreous, choroid, and retina) with subsequent eye hypotony.

Complaints. Patients with perforating eye injuries typically complain of eye pain, redness, tearing, swelling, and decreased vision.

Signs. The absolute signs of perforating eye injuries are a foreign body behind the eye, entrance and exit wounds, hemorrhagic chemosis, proptosis due to bleeding into the retro-bulbar space, loss of the eye shape, prolapse of the crushed intraocular structures through the edges of the wound. Additional findings may include hyphema, collapsed anterior chamber, limitation of ocular movements, enophthalmos, vitreal hemorrhage, and chorioretinal injury.

Methods of Examination. Principles of ophthalmic examination are the same as in cases of penetration injury and include thorough trauma history, external ocular inspection, VA test, pupillary evaluation, slit-lamp exam, ophthalmoscopy, diaphanoscopy. If an intraocular foreign body is suspected, ultrasonography, CT scan, X-ray, or MRI are needed.

NOTE!

DO NOT apply any pressure to the eye surface.

DO NOT dilate the pupil.

Differential Diagnosis. Penetrating eye injury, eyeball rupture, acute red eye.

Treatment. First aid is the same as in penetrating injury — instillation of a topical anesthetic, disinfectant solutions, broad-spectrum antibiotics, tetanus immunization or prophylaxis, and patching of both eyes. After this the patient should be referred to a hospital for surgical treatment. Removal of foreign bodies and blood clots is contra-indicated. Surgical treatment consists of foreign body removal and optic reconstructive surgery.

Prognosis. Perforating eyeball injuries have poor visual and anatomic outcomes associated with proliferative vitreoretinopathy (PVR).

Complications. Multiple complications are known to develop after perforating injury, such as: corneal scarring, iridodialysis, iridocyclitis, post-traumatic glaucoma, uveitis, cataract, vitreous hemorrhage, retinal detachment, suppurative endophthalmitis, panophthalmitis, loss of the eye. Sympathetic ophthalmia is autoimmune inflammation of the other eye in response to eye injury; it may occur beginning from 2 weeks to even few years after the injury.

Prophylaxis. While not every perforating injury can be prevented, appropriate and adequate eye protection when performing visually threatening activities is the most effective method to prevent many accidents of ocular trauma.

Intraocular Foreign Bodies

Definition. Intraocular foreign bodies (IOFBs) refer to the presence in the eye of a foreign object from a penetrating injury.

Etiology. An intraocular foreign body can be associated with a penetrating or perforating injury and can involve the anterior chamber, crystalline lens, posterior chamber, and even the orbit if a posterior exit wound is present.

Hammering, using machine tools, shooting weapons, and being in proximity to explosion are among the most common mechanisms responsible for retained IOFB cases.

Other causes may be: road traffic accidents, different hobbies, playing sports or games with sharp objects. An IOFB may be any material such as metal, glass, stone, or even pencil lead, etc.

Clinical Picture. Patients with IOFB most often have eye pain and decreased vision, but, initially, if the foreign body is small and was introduced into the eye at high velocity, they may have no symptoms. In some cases IOFBs can protrude from the eye or be easily identified at ocular examination, but there are cases when IOFBs can be missed due to their size, nature, and location (fig. 16.12, 16.13).

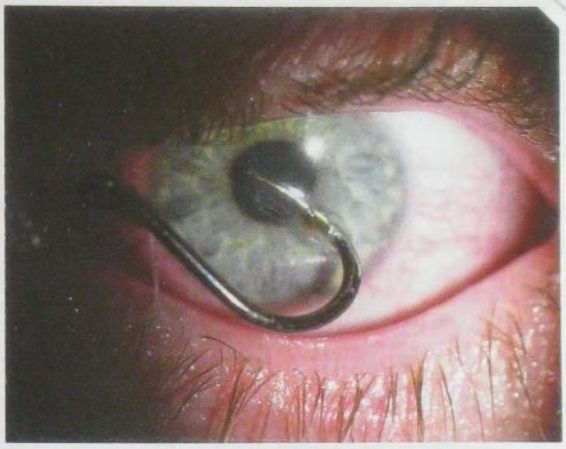


Fig. 16.12. Intraocular foreign body protruded from the cornea (fish hook)

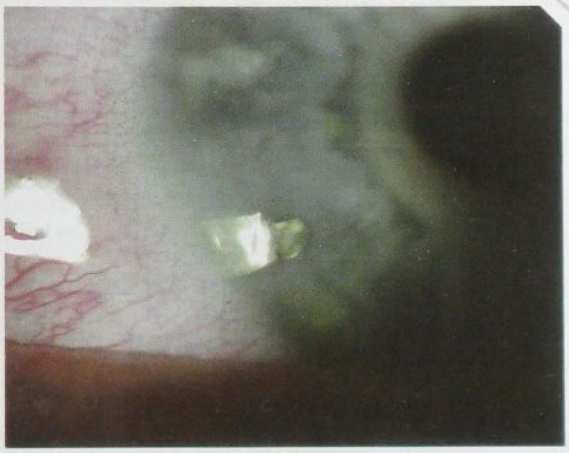


Fig. 16.13. Intraocular foreign body located in the anterior chamber

IOFBs can be classified according to the:

— anatomical zone of the entry and exit: as zone 1, 2 or 3 (according to the Ocular Trauma Classification System (OTCS);
— location: as the anterior or posterior segment;
— according to the nature: as a magnetic foreign body (copper, iron, etc.) or a non-magnetic foreign body (glass, plastic, stone, wood, etc.).

Intraocular foreign bodies that enter the eye may cause damage in two ways: 1) they may cause structural damage to the intra-ocular contents as they enter and pass through the eye; and 2) they may cause toxicity to the tissues as they degrade or oxidise if not removed urgently.

Complaints. The symptoms are quite variable depending on the speed of penetration and the intraocular structures affected. Patients complain of eye pain, redness or bloodshot eyes, burning or irritation in the eye, excessive tearing, light sensitivity, difficulty in eye opening, discharge of blood or fluid, blurred vision.

Signs. Ophthalmologic findings in IOFB include: decreased VA, a penetrating corneal or scleral injury, conjunctival chemosis, hyphema, localized cataract, iris injury, pupillary asymmetry, vitreous hemorrhage, commotio retinae, retinal tear, decreased IOP, aqueous humor leakage (positive Seidel test), visualized and localized IOFB within the eye.

Methods of Examination. Thorough history, including circumstances of the trauma, time of injury, possible materials involved in the injury. External ocular inspec-

tion, VA assessment, pupillary evaluation, slit-lamp exam, ophthalmoscopy, B-scan ultrasonography, CT scan, X-ray, MRI.

The use of CT is limited in case of small fragments < 0.7 mm in size, wooden foreign bodies, IOFBs in the scleral wall, or glass near the lens.

MRI can be used when a plastic or wooden IOFB is suspected, but it is not indicated in the case of metallic IOFBs as the magnetic forces can alter the position of a metallic IOFB and cause further injury to the eye.

Differential Diagnosis. Corneal abrasion, globe rupture, superficial foreign body, endophthalmitis.

Treatment. The final management of an IOFB injury consists of urgent repair of the entry site, removal of the IOFB and attention to the associated ocular damage.

First aid in retained IOFB includes:

— washing the wound with *disinfectant solutions* (Sulfacetamide, Decamethoxin, Myramistin);

— *instillation of broad-spectrum antibiotic drops* (Ofloxacin, Tobramycin, Levofloxacin, Ciprofloxacin);

— *tetanus prophylaxis* if not current;

— *a binocular aseptic bandage* to restrict eye movements in order to avoid further injury to the eye (if the IOFB is protruding from the eye, it has to be shielded by placing a plastic eye shield or a paper cup over it).

NOTE!

DO NOT apply any pressure to the eye surface.

DO NOT attempt to remove any protruding IOFBs.

DO NOT attempt to remove any dark-colored material on the ocular surface as that may be prolapsed intraocular contents.

After first aid administration, the patient must be immediately transported in supine position (if possible) to the nearest specialized ophthalmic trauma department for surgical removal of the IOFB.

The entry wound should be closed initially in order to provide stability to the eye. The IOFB should be removed in a manner that minimizes trauma upon extraction. Metal magnetic foreign bodies can be removed with a hand-held magnet, non-magnetic bodies are removed with the use of forceps. There are different approaches to foreign body removal:

— direct — through the wound site — is performed if a foreign body is incarcerated within the edges of the cornea or sclera;

— anterior — through the limbal incision — if the foreign body remains in the anterior segment of the eye: anterior chamber, iris, or stuck in the lens (in that case the lens may be removed at the same time);

— pars plana vitrectomy — through the plain part of the ciliary body (5 mm from the limbus) — if the foreign body is seen in the vitreous;

— transscleral — through the additional scleral incision as close to the foreign body as possible — if it has intraretinal localization.

At the time of surgery associated conditions are addressed and to avoid panophthalmitis intravitreal antibiotics are indicated.

Prognosis. Proper diagnosis, investigation, and timely and appropriate surgical and postoperative management are the keys to a successful outcome with an intraocular foreign body. But the prognosis of IOFB trauma is multifactorial and largely depends on the type of the IOFB, its size, location, associated ocular pathology, and the time of aid administration. In patients with smaller wound lengths (under 2 mm), IOFBs that are located in the anterior segment only have the best prognosis. Negative prognostic factors include a longer wound length (greater than 3.5 mm), posterior segment IOFBs, poor initial visual acuity, and the presence of complications arising from IOFBs (retinal detachment, endophthalmitis).

Complications. Corneal scar, cataract, secondary glaucoma, uveitis, vitreous hemorrhage, proliferative vitreoretinopathy, retinal detachment, endophthalmitis, panophthalmitis, phthisis bulbi, sympathetic ophthalmia. If a metallic IOFB is retained in the eye for a long time, it can lead to inflammatory reactions known as *metallosis*:

— *siderosis* usually occurs after 2 months to few years of the trauma due to the chemical reaction of iron ions with eye proteins, which leads to degenerative changes in the structures: rusty deposits and opacification of the lens, iris color change — initially to greenish, and later — to reddish-brown, retinal pigmentary degeneration, secondary glaucoma;

— *chalcosis* is produced by the alloy of copper that undergoes electrolytic dissociation and settles under the membranous structures of the eye. It represents as a golden brown ring at the periphery of the cornea under Descemet's membrane (Kayser—Fleischer ring); a sunflower cataract, golden deposits in the retina, vitreous fibrosis, secondary glaucoma, etc.

If IOFB is a piece of inert materials such as glass, plastic, porcelain, gold, silver or platinum, no reaction is produced.

Prophylaxis. Wearing appropriate protective eyewear (such as face shields, safety glasses, or goggles) is the best way to prevent intraocular foreign bodies and other types of eye injuries. Protecting the eyes is especially important when working with machinery that could cause chips of wood or metal to splinter, as well as doing chores around the house or yard, or playing sports.

4.2. Injuries of the Ocular Adnexa

The most frequent injuries of the ocular adnexa are conjunctival lacerations, eyelid hematoma and lacerations, and orbital trauma.

Eyelid Hematoma

Definition. Eyelid hematoma, or a black eye, or periorbital hematoma, refers to bruising around the eye as a result of a subcutaneous accumulation of blood due to bleeding from the eyelid vessels.

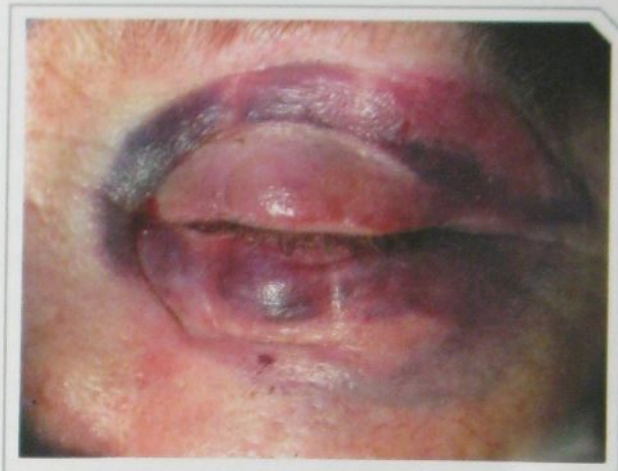


Fig. 16.14. Eyelid haematoma with subcutaneous emphysema after contusion trauma

Etiology. Eyelid hematoma commonly appears after a blunt trauma to the soft tissues of the periocular region because of good vascularization of the eyelids and loose attachment of the subcutaneous tissue. It also may follow affection of the other parts of the scull, such as the orbital cavity or nasal sinuses. Most black eyes happen by accident — during contact sports, at work, in a car crash or during home repair. The source of the injury is usually a blunt object — a fist, a baseball, a hammer, a rock, etc.

Other causes of a black eye include surgical procedures to the face, such as a facelift, jaw or nasal surgery, dental work, insect bites, allergic reaction, or cellulitis.

Bruising around both eyes, known as raccoon eyes, may indicate a skull fracture or another type of head injury. It requires urgent medical attention.

Clinical Picture. Eyelid hematoma appears as bright-red swelling of eyelids tender to palpation, often blood spreads to the subconjunctive space. Sometimes independent opening of the eyelids is not possible due to severe swelling or to mechanical damage to the levator of the upper lid muscle (traumatic ptosis). As a common bruise, eyelid hematoma discolorates with time to purple, black, green and yellow color and resolves within a week (fig. 16.14).

Complaints. Most often patients complain of pain, bruising, and swelling of the eyelid. Some may notice blurry vision, difficulty opening the eye, and headache.

Signs. Hematoma and swelling of the eyelids.

Methods of Examination. Medical history, external ocular inspection, ocular movements, palpation, VA assessment, pupillary evaluation, slit-lamp exam, ophthalmoscopy. An additional X-ray or CT-scan of the orbit is needed to exclude more serious disorders.

Differential Diagnosis. Globe contusion, superficial eyelid cellulitis, allergic reaction, orbital wall fracture, nasal fracture, basal skull fracture, intracranial hemorrhage.

Treatment. Normally eyelid hematoma is not a major problem, and usually hospitalization is not needed.

NOTE!

Most black eyes are relatively minor injuries, but they may indicate a more serious eye trauma or brain injury.

Symptoms of a serious injury are double vision, loss of sight, bleeding in the eye, persistent headache, loss of consciousness, nausea, vomiting, etc.

To relieve swelling you can apply a cold compress (such as an ice bag or a cool, damp cloth) for about 15 to 20 minutes at a time and can be reapplied every hour for the first day after the injury occurs. This will help constrict blood vessels and limit the amount of swelling.

NOTE!

Never put a raw steak or other raw meat on a black eye, as the bacteria on raw meat pose a high risk of infection.

For pain relief analgesics, such as Tylenol, may help (avoid Aspirin, which is a blood thinner and therefore can increase bleeding).

When the swelling reduces, apply a warm compress and resorbtive ointments (Solcoseryl, Actovegin) to help promote blood reabsorption.

Traumatic ptosis that does not reside within one month requires additional therapy: electrostimulation, massage, or surgical treatment.

For complicated eyelid hematoma, the patient may be referred to an appropriate specialist (otorhinolaryngologist, neurosurgeon, maxillofaical surgeon, etc.) and may have additional treatment recommendations.

Prognosis for an uncomplicated black eye is generally good. If eyelid hematoma is a sign of a more serious injury, complications may arise.

Complications. Pain, swelling, and discoloration may persist for a few days to weeks after the injury, however eyelid hematoma heals without any complications.

If a black eye was caused by a severe trauma, the eyeglobe complications may arise, such as traumatic uveitis and iritis, hyphema, glaucoma, retinal detachment.

Prophylaxis. Basic injury prevention may help to avoid a black eye: wearing appropriate protective eye gear or a face shield for any sport or work-related activity; wearing goggles when working in the yard or being involved in some hobbies; fastening seatbelts while driving; wearing helmets while riding a motorcycle or a bicycle, etc.

Eyelid Laceration

Definition. Eyelid laceration is a cut to the eyelid caused by trauma.

Etiology. Laceration can be caused by a direct blow with a hard blunt object (a stone, a wooden stick, falling down, etc.), by cutting with sharp objects (metal weapon, a kitchen knife, a piece of glass, especially during car accidents, etc.), by handlebar injuries, bites of dogs and other animals, etc.

Clinical Picture. Eyelid lacerations can be superficial (skin), partial-thickness (muscle and fat) or full-thickness. Lacerations may involve the tarsal plate or extend through the lid margin. If the medial corner of the upper and lower eyelid is affected, this can cause damage to the canaliculi of the lacrimal system and puncta displacement (fig. 16.15).

Ocular injury may accompany eyelid laceration in up to two-thirds of cases; about one quarter of patients with open globe injures have associated eyelid or periorbital laceration. In cases of deep eyelid laceration, corneal or scleral lacerations or IOFBs are suspected.

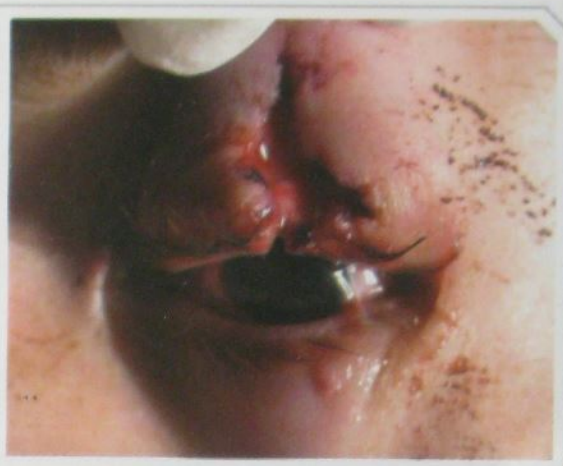


Fig. 16.15. Laceration of the upper eyelid with integrity damage of the intermarginal edge

Complaints. Periorbital pain, bleeding, swelling and sometimes impaired vision (due to swelling or drooping of the eyelid).

Signs. Visible partial or full-thickness defect in the eyelid.

Methods of Examination. History of trauma, external ocular inspection, pupillary reaction, ocular movements, VA test, focal, bifocal illumination, slit-lamp exam, ophthalmoscopy, tonometry. If a foreign body is suspected, CT-scan of the orbit or head may be performed.

Differential Diagnosis. Eyelid laceration is an obvious condition, but be sure not to miss associated ocular trauma or a foreign body.

Treatment. Eyelid lacerations should be repaired within 12 to 24 hours of the injury and proper primary repair can reduce subsequent complications. Wounds with signs of bad contamination or purulent inflammation are treated first with local and general antibiotic therapy, their surgical repair should be delayed.

Tetanus vaccine (if the last booster was received more than 5 years ago) and oral analgetics must be given. If laceration is due to an animal bite, the standard rabies protocol must be followed.

First, the wounds must be cleaned thoroughly of all the gravel, metal, glass or other particles and irrigated with saline solutions. At times it is necessary to debride some lid tissue.

Simple superficial (skin) horizontal lacerations that involve less than 1 mm do not require suturing and will heal spontaneously. After cleaning, with antibiotic ointment is applied onto the wound, and the wound is covered with an adhesive surgical tape or a sterile bandage. In cases when cuts are long, they can be repaired easily with sutures (stitches).

Deeper lacerations involving the tarsal plate or lacerations extending to the lid margin require surgical repair. The multiple layers of the lid must be apposed correctly to avoid poor lid function (fig. 16.16) according to the algorithm:

— the intermarginal border is sutured first with 3 stitches, the long ends of the posterior margin suture are pulled anteriorly and fixed under the anterior margin stitch;
— the posterior eyelid lamella (the tarso-conjunctive plate) is sutured with absorbable 5/0—7/0 sutures, the ends are cut short;
— the anterior eyelid lamella (the skin and orbicularis muscle) is sutured with interrupted and/or running non-absorbable 5/0—7/0 sutures;
— antibiotic ointment should be applied 2 times a day for 5—7 days;

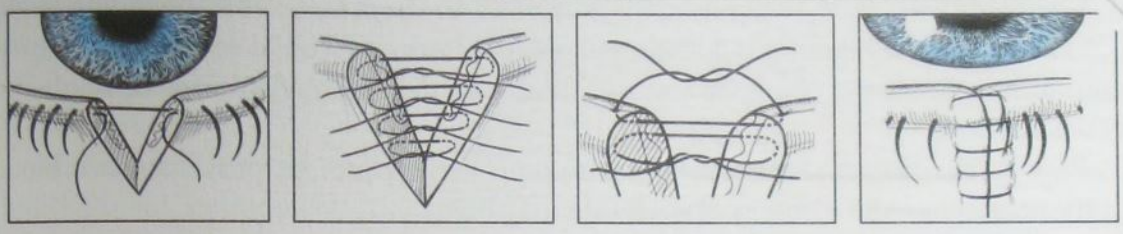


Fig. 16.16. Surgical reconstruction of deep eyelid laceration with thorough layers apposing

— sutures should be removed in 5—7 days, the intermarginal sutures are taken off no earlier than in 2 weeks.

Surgical restoration of lacerations through the canaliculus requires the use of silicone nasocanalicular stents or special probes. The silicone stent remains in situ for three to four months and is removed then (fig. 16.17).

Eyelid injuries resulting in large tissue loss require the use of grafts or z-plasty reconstruction that should be administered by an oculoplastic specialist.

Prognosis. The visual prognosis for lid lacerations is usually excellent unless there is accompanying globe rupture. With proper reconstruction of lid lacerations, the cosmetic result is usually quite good.

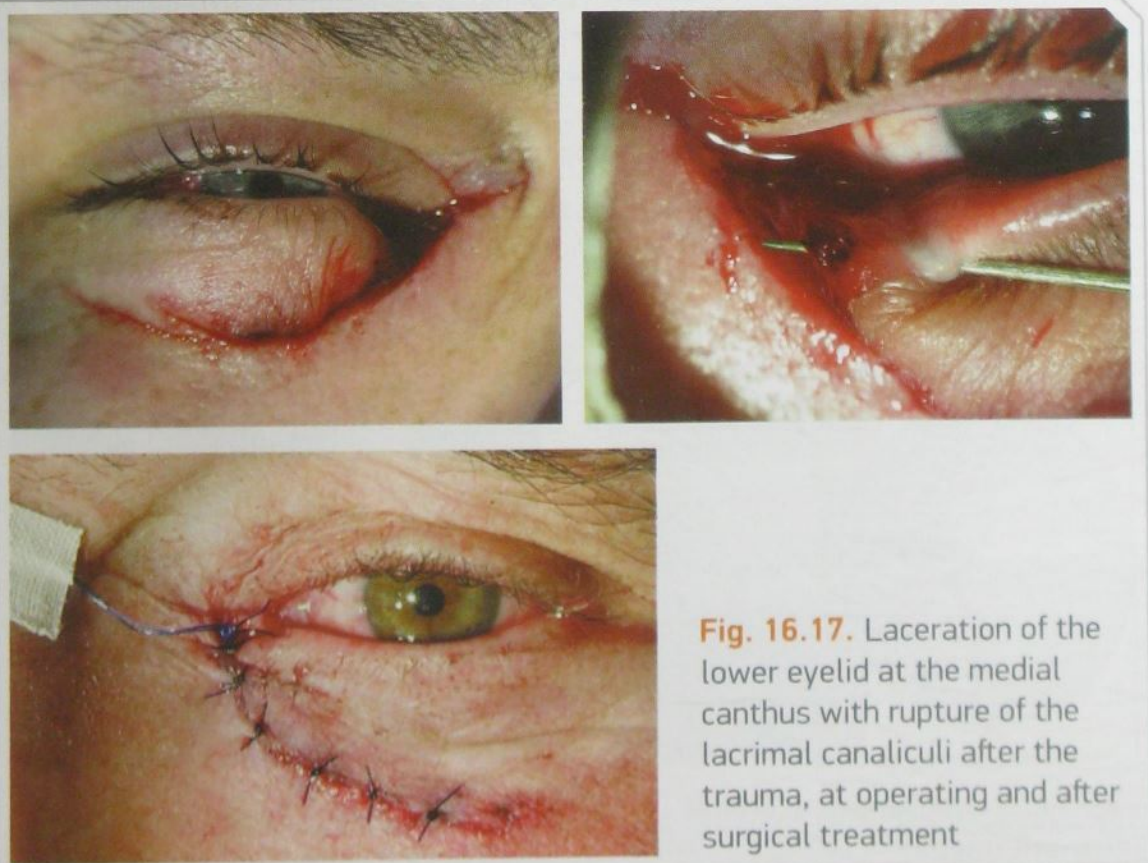


Fig. 16.17. Laceration of the lower eyelid at the medial canthus with rupture of the lacrimal canaliculi after the trauma, at operating and after surgical treatment

Complications. Early wound repair is usually successful, but complications such as cutaneous scars, lid margin notching, lagophthalmos, cicatricial ectropion or entropion, traumatic ptosis, epiphora and infection may develop and require oculoplastic reconstruction.

Prophylaxis. Most of the eyelid laceration cases cannot be prevented but basic injury prevention and using eye protection may help.

Orbital Blow-out Fracture

Orbital trauma is a traumatic injury to the orbital soft tissues (contusion, retrobulbar hemorrhage) or fracture of the orbital bones. The most common type of orbital trauma is an orbital blow-out fracture.

Definition. Orbital blow-out fracture is a traumatic break of one of the walls of the orbit with the orbital rim remaining intact. Most often, the orbital floor or medial wall of the orbit is affected.

Etiology. Orbital blow-out fracture is usually caused by blunt trauma and may result from car accidents, sports-related injuries, industrial accidents, falls, dog bites, assaults, and explosions. Typically, this type of injury results from the impact of a blunt object larger than the orbital aperture or eye socket.

The mechanism of orbital blow-out fractures is explained by a sudden increase of intraorbital pressure, which in its turn causes decompression by the fracture of one or more of the bounding walls of the orbit (fig. 16.18, fig. 16.19). It also causes damage of the orbital vessels, nerves, etc.

Clinical Picture. Orbital blow-out fractures are classified according to several characteristics:

— size — big or small;
— location — medial wall, orbital floor;

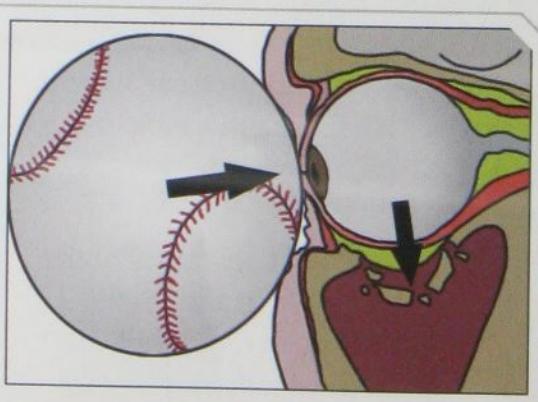


Fig. 16.18. Mechanism of orbital blow-out fracture

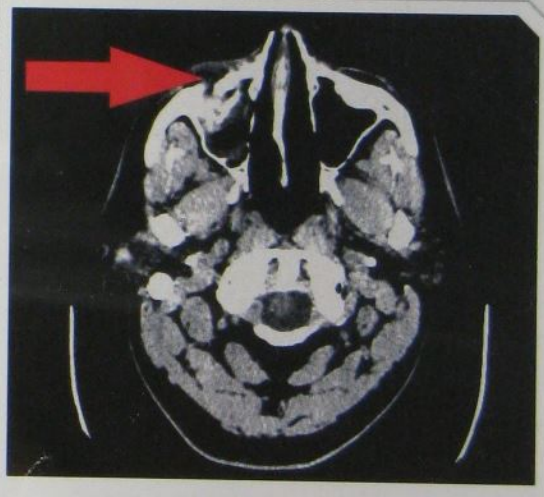


Fig. 16.19. Fracture of the orbital floor seen on MRI

— bone position — in place or displaced;
— entrapment of tissues or muscles in the fracture — entrapped or not;
— accompanying symptoms — double vision, pain, affecting eye position and movements.

Medial wall fracture may lead to rupture of the lacrimal canaliculi or medial canthal ligament, entrapment of the medial rectus muscle in bony fragments, periorbital emphysema.

Inferior wall (orbital floor) fracture may result in an injury of the optic nerve, inferior oblique and rectus muscles, infraorbital nerve.

Mild trauma may cause a fracture of minimal size with minimal or no double vision, minimal or no interference with eye movements. Severe trauma may cause facial bone fractures, injury to the eye itself, and injuries to the skull and brain.

Complaints. Symptoms usually depend on both the type and severity of the orbital fracture. Patients typically complain of bruising; tenderness and swelling around the eye; redness of the eye; facial numbness on the side of the trauma; nose bleeding (epistaxis); painful eye movement; decreased visual functions and double vision that disappears when one eye is covered.

Signs. Restriction of eyeball movements, subcutaneous periorbital emphysema, hypoesthesia in the infraorbital or supraorbital region, displacement of bony fragments at palpation, enophthalmos (may be masked due to the retrobulbar edema or hematoma), syndrome of superior orbital fissure (ophthalmoplegia, ptosis, midriasis, accommodative paresis, corneal sensitivity decrease).

Methods of Examination. History of trauma, external ocular inspection, palpation, pupillary reaction, eye movements, VA test, focal, bifocal illumination, slit-lamp exam, ophthalmoscopy, exophthalmometry, forced duction test, lacrimal patency tests, X-ray of the orbit and nasal sinuses, CT or MRI of the orbit; consultations with a neurosurgeon, ENT, a cranio-facial surgeon must be suggested.

Differential Diagnosis. Eyelid hematoma, orbital edema or retrobulbar hemorrhage without orbital bone fractures, skull fracture.

Treatment depends on the location and severity of the injury. For a small, uncomplicated blowout fracture that does not affect eye movement only conservative treatment is required that includes:

— an ice pack to the periorbital region for 24—48 hours to decrease pain and swelling;
— nasal vasoconstrictors for 2 to 3 days to minimize nosebleeding;
— systemic antibiotics within 10—14 days to prevent spread of infection from the sinuses into the orbit;
— systemic steroids to decrease swelling and scarring;
— instructing the patients not to blow the nose to prevent extreme pressure to the orbit from the sinuses;
— systemic analgesics to help control pain.

If the eye itself is not damaged but there is an eyelid or canalicular damage, this should be repaired within 3 days of the injury.

Surgical treatment of orbital blow-out fracture is performed not earlier than in 7—14 days after the trauma; that allows the initial edema and hemorrhage to decrease. It involves removing of the bone fragments, freeing the trapped eye muscles and soft tissues of the orbit and restoring the normal architecture of the eye socket with the use of an implant, a thin plastic sheet, or a bone graft to connect the broken parts and assist healing.

Prognosis. Isolated blow-out fractures, even those requiring surgical repair, have good prognosis. Untreated orbit fractures can result in permanent facial deformity of the face, enophthalmos, and diplopia.

Complications. Enophthalmos, double vision, paresis or fibrosis of an exrtraocular muscle, orbital cellulitis, exophthalmos, implant extrusion, facial deformity.

Prophylaxis. Prevention of an orbital blow-out fracture is only possible by preventing blunt trauma to the eye. To decrease the risk of this trauma it is necessary to use appropriate protective eyewear while working, playing sports, to fasten seat belts while driving or wear a helmet when cycling or driving a motorbike, etc.

4.3. Burns

The blinking reflex usually causes the eyelids to close quickly in response to a dangerous situation to protect the eyes from damage. However, irritating or harmful agents still sometimes get onto the surface of the eye causing burns.

NOTE!

Any burn injury to the eye is considered an ophthalmologic emergency and requires immediate and intensive treatment as it could lead to a permanent loss of vision.

A burn of the eye (ocular burn) can involve the sclera, conjunctiva, cornea, and eyelids, as well as the deeper structures inside the eye. The severity of eye burn depends on the cause of the injury, the duration of exposure to the agent, and the time elapsed before initiation of treatment. Ocular burns can be caused by chemical substances, thermal agents, or radiation. They occur in 6—40 % of eye trauma, and ¾ of all burns are chemical.

Chemical Burns

Definition. Chemical burns are burns of the eye that occur when solid, liquid or vaporous chemicals get into the eye.

Etiology. Almost any chemicals used at home and at work can cause ocular burn, but serious damage usually results from alkaline or acidic compounds.

Alkali burns generally result from exposure to compounds found in lime, whitewash, caustic potash, lye, liquid ammonia, cement, drainpipe cleaners, fertilizers, firework sparklers, and flares.

Acid burns are typically caused by compounds found in car batteries, refrigerants, cleaners (bleach), acetic acid (vinegar, glacial acetic acid) and industrial materials containing hydrochloric or hydrofluoric acid.

Alkali burns are reported more commonly than acid burns because of their frequent use in many household cleaning agents and building materials. Some of the more common chemical agents are listed in table 16.3.

Table 16.3

Common Chemical Agents and Their Sources*

Substance	Class	pH	Source
Sodium hydroxide	Alkali	14.0	Lye soaps, airbags, hair relaxer
Calcium hydroxide		12.4	Mortar, plaster, cement, whitewash
Ammonium hydroxide		11.6	Fertilizers, refrigerants, sparklers
Sodium hypochlorite		11.0	Bleaches, drain cleaners
Magnesium hydroxide		10.0	Oven & drain cleaners
Acetic acid	Acid	2.9	High vinegar concentrations
Hydrofluoric acid		2.1	Rust removers, glass, mineral, gasoline, silicone industries
Sulfurous acid		1.5	Bleach, refrigerants
Sulfuric acid		1.2	Industrial cleaners, car battery acid
Hydrochloric acid		1.1	Household and pool cleaners

* *Table adapted from Emergency Medicine Procedures*

Chemical burns may be caused by a chemical agent getting directly into the eye or when a person rubs the eye after working with hazardous chemical materials. They may be associated with periocular, facial, or body burns.

Clinical Picture. Alkali burns are more severe than acid burns. Alkali chemicals have a high pH and are lipophilic. They cause significant epithelial cell disruption and easily penetrate into the corneal stroma and the anterior chamber (in less than 1 min) resulting in saponification of fatty acids of cells and gelatinisation of tissues. This is referred to as liquefactive or colliquative necrosis.

Alkali chemicals spread wide and deep, exposing the iris, ciliary body, lens, and trabecular meshwork to further damage resulting in a danger of permanent loss of vision. They continue to cause damage long after the injury has taken place. The prognosis is worse than with acid burns.

Acids have lower pH than alkalis. They are superficial and unable to penetrate into the eye, but are still dangerous. They cause coagulation and denaturation of proteins preventing deeper penetration of a substance (coagulative necrosis). However, some acids penetrate deeply like alkalis and cause similarly severe injuries. Concentrated sulfuric acid (such as from an exploded car battery) draws water out of tissue and simultaneously develops intense heat that affects every

layer of the eye. Hydrofluoric acid and nitric acid have a penetrating effect similar to alkalis.

Typically, acid burns cause their maximum damage within the first few minutes to hours and are non-progressive. They may cause a significant direct damage to the cornea that also may result in vision loss.

Clinical presentation of ocular burns goes through four different stages:

1) acute (duration of 7—8 days);
2) marked trophic disorders (duration of 2—3 weeks);
3) compensatory vascularization (duration of 4—5 weeks);
4) scarring (following months).

The severity of acid burns as well as of thermal burns is determined in the first hours after the injury, and of alkaline burns — in a few days, due to the peculiarities of deep penetration of alkali into the tissues.

According to damage severity based on the corneal appearance and the extent of limbal and conjunctival ischemia, chemical burns are divided into four grades:

I grade (mild burn) — hyperemia of the eyelid skin and conjunctiva, epithelial damage of the cornea, mild corneal edema and no limbal or conjunctival ischemia. Insignificant reduction in vision acuity.

II grade (moderate burn) — blisters on the eyelid skin with eyelid edema, conjunctival chemosis, ischemia of the conjunctiva that affects less than one third of the limbus, corneal erosions, superficial dense opacification of the cornea, but the anterior segment structures are typically visible (fig. 16.20). Visual acuity is reduced to 0.2—0.3; corneal sensitivity is greatly reduced.

III grade (severe burn) — total epithelial loss, advanced chemosis with anemic conjunctiva, dense stromal haze ("dim glass"), blurring of the anterior chamber details, iritis, limbal ischemia of less than one half of the limbus, elevated intraocular pressure, signs of necrosis.

IV grade (very severe burn) — necrosis stage — deep necrosis of the conjunctiva and cornea, dense cornea opacification ("porcelain glass") that doesn't allow to see the anterior segment structures, 2nd and 3rd degree burns of the periocular tissues, limbal ischemia is greater than one half of the limbus, possibility of corneal perforation. Visual acuity is reduced to light perception.

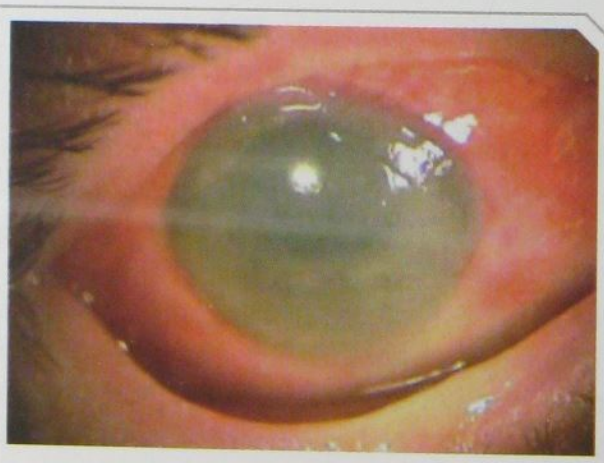


Fig. 16.20. Chemical burn of the eye II grade

Complaints. The most common patients' complaints are severe pain, eye redness, foreign body sensation, blurred vision, excessive tearing, photophobia, blepharospasm, inability to keep the eye open, swelling of the eyelids.

Signs. Clinical signs depend on the grade of the burn. A whole range of eye damage is usually seen after 3—4 days of the injury:

— burns to the eyelids and surrounding skin;
— conjunctival chemosis and hyperemia;
— limbal and conjunctival blanching;
— corneal epithelium defects of different range;
— corneal edema and opacification;
— raised IOP;
— corneal melting;
— formation of leukoma.

The intensity of corneal opacity is greater with acid burns, but alkalis cause deeper damage, involving the internal structures of the eyeglobe (iris, lens, ciliary body), initiating early iridocyclitis, later — cataract and secondary glaucoma.

Methods of Examination. History taking and physical examination must be delayed until the affected eye is irrigated and the pH of the ocular surface is neutralized. VA test, external ocular inspection, slit-lamp exam, ophthalmoscopy, tonometry, perimetry.

History of the trauma is vitally important to know the chemical causing the injury as antidote solutions are different.

Differential Diagnosis. Corneal foreign body, traumatic corneal abrasion.

Treatment. Chemical burn management consists of three phases: 1) immediate first aid; 2) midterm therapy (for several days to weeks immediately postburn); 3) treatment of chemical burn complications (after 21 days).

NOTE!

Chemical burns are a true ophthalmic emergency.

It's the only ocular condition where history taking and examination should be delayed.

Severe bilateral chemical burn is especially devastating and often results in complete visual disability.

Immediate first aid at the scene of the accident is crucial:

— irrigation with sterile isotonic saline or with any watery solution of neutral pH (plain water, mineral water, tea, or similar liquids) for at least 30 min. Milk should be avoided as it increases burn penetration by opening the epithelial barrier. If possible, carefully pull down the lower eyelid and evert the upper eyelid to irrigate the fornices. Any obvious foreign bodies should be also irrigated away or removed;
— topical anesthesia (Proxymetacain 0.5 %) may be used prior to irrigation to help decrease the pain and make it possible for the patient to open the eyes;
— for chemical neutralization in case of alkaline burns — weak acidic solutions (e.g. boric acid), ethylendiaminetetraacetic acid (EDTA) 1 % solution for lime can be used; acid burns are neutralized with weak alkaline solutions (e.g. sodium bicarbonate);
— if litmus paper is available, test pH in the lower eyelid fornix and continue irrigation until pH is neutralized to a range of 7.0—7.2;

— after irrigation, an antibiotic ointment (Ofloxacin, Tobramycin, Levofloxacin, Tetracycline) and a soft bandage should be applied.

Only after these actions have been taken the patient should be transported to an ophthalmologist or an eye clinic.

Midterm therapy includes:

— tetanus prophylaxis;
— topical antibiotic ointments (Ofloxacin, Tobramycin, Levofloxacin, Erythromycin, Gentamicin, Tetracycline) for infection prevention;
— midriatics, cycloplegics, and topical steroids to decrease local inflammatory reaction; control of exudation;
— antiglaucoma agents as prophylaxis against secondary glaucoma (Timolol);
— topical lubricants (Dexpanthenol, Actovegin, Solcoseryl) for promotion of ocular epithelial healing;
— therapeutic soft contact lenses with high water content to assist epithelial healing, which in its turn will inhibit enzyme release and stromal melting;
— oral and topical vitamin C to neutralize cytotoxic radicals;
— oral analgetics and sedatives;
— massage of the conjunctival fornices with glass plates twice a day for prevention of symblepharon formation.

For *treatment of chemical burn complications* surgical options are used such as:

— excision of necrotized tissues to decrease inflammation and help healing;
— peritomy of the conjunctiva;
— grafting of the autoperilimbal stem cells or amniotic membrane;
— transplantation of healthy mucous membrane (from the other eye or oral mucosa);
— corneal transplantation;
— symblepharon lysis to improve the motility of the globe and eyelids;
— plastic surgery of the eyelids;
— secondary cataract and/or glaucoma surgical treatment.

Prognosis is usually good in I—II-stage chemical burns with full recovery or some scarring. But it is guarded for III-grade and very serious for IV-grade burns, because they initiate a complex of secondary toxic and fibrotic reactions in the eye.

Complications. Corneal opacities, corneal scarring, cataract, secondary angle-closure glaucoma, symblepharon, lagophthalmos, entropion, ectropion, walleye (or leukoma), corneal perforation, atrophy of the eyeball, blindness.

Prophylaxis. Safety officials estimate that up to 90 % of chemical eye injuries can be avoided. The easiest way to prevent this injury is wearing appropriate safety goggles or a face shield when working with liquid or powder chemicals, or sprays both at work and at home. Keep all hazardous home products away from children.

Thermal Burns

Definition. Thermal burns are ocular burns that occurr due to exposure of the eye to a thermal factor.

Etiology. Thermal burns can be caused by boiling water, hot steam, splatters of hot grease or cooking oil, molten metal or plastic objects, a cigarette, explosions and open flame. Scald burns from hot liquids are the most common burns to children and older adults.

Direct injury to the globe is usually limited due to the reflex closure of the eyelids and a good Bell's phenomenon, therefore thermal injuries to the eye are often associated with periocular, facial, or entire body burns.

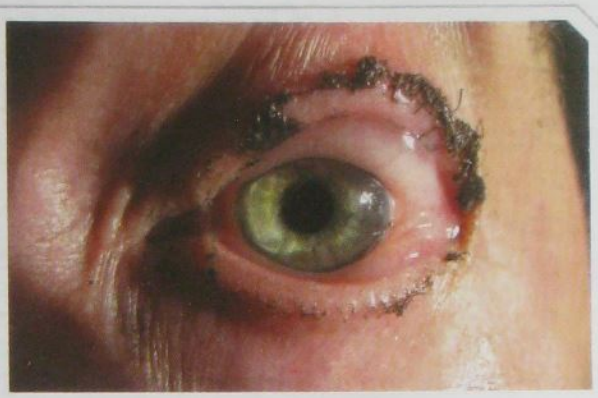


Fig. 16.21. Thermal burn of the eye (I-II grade) by petard explosion; eyelashes melting, conjunctival chemosis and corneal erosion

Clinical Picture. The clinical course of a thermal burn is usually less severe than that of a chemical injury. This is because burns cause superficial coagulation; however, thermal necrosis and penetration can occur.

Complaints. Severe pain, foreign body sensation, irritation, lacrimation, blepharospasm, photophobia, blurred vision.

Signs. According to the grade of the burn ophthalmologic findings may be as following (fig. 16.21):

I grade — hyperemia of the skin and conjunctiva, superficial corneal erosions;

II grade — eyelid skin vesicles, superficial conjunctival membranes, superficial opacity of the cornea;

III grade — necrosis of the skin and conjunctiva, opacification of the superficial and middle layers of the cornea;

IV grade — necrosis of the periocular skin, subcutaneous tissues, conjunctiva, sclera; opacification of all layers of the cornea.

Methods of Examination. After copious irrigation, a full ophthalmologic examination is required. VA test, external ocular inspection, slit-lamp exam with fluorescein staining, ophthalmoscopy, tonometry, perimetry.

Differential Diagnosis. Chemical burns, corneal foreign body, traumatic corneal abrasion, ulcerative keratitis.

Treatment. The goal of the thermal burn treatment is to reduce pain, prevent infection, promote wound healing and prevent complication development.

First aid consists of:

— irrigation with cold water for 20 minutes and removal of the thermal agent if present;

— topical anesthesia (Proxymetacain 0.5 %) may be used to help decrease the pain in order to open the eye;

— topical antibiotic ointments (Ofloxacin, Tobramycin, Levofloxacin, Tetracycline, Gentamicin, Tobramycin, Ciprofloxacin, Erythromycin, etc.);

— cool compresses to reduce pain;
— cover the eye with a sterile bandage or cloth without applying any pressure and send the patient to an ophthalmologist for the next treatment options.

The following treatment includes:
— update of tetanus toxoid;
— removal of necrotic tissue;
— instillation of antibiotic eye drops or ointments;
— topical mydriatics;
— instillations of NSAIDs — Diclofenac, Indometacine;
— instillations of lubricating drops and gels;
— oral analgesics;
— using a glass rod to prevent symblepharon.

In case of complications — their surgical treatment as with chemical burns.

Prognosis is usually good in I—II-stage of thermal burns with full recovery or some scarring, relatively good for III-grade and very serious for IV-grade burns.

Complications: eyelid contracture (entropion, ectropion), exposure keratitis, corneal ulcer, corneal opacification, corneal scarring, uveitis, glaucoma, symblepharon or ocular atrophy.

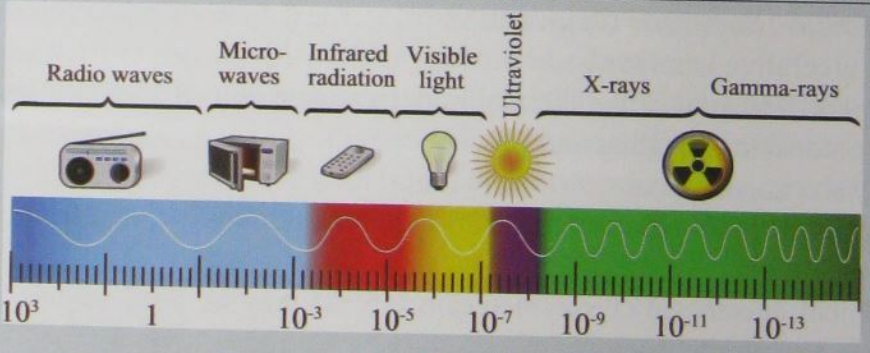
Prophylaxis — the use of appropriate protecting goggles when working at industries with a danger of thermal burn, careful handling of hot fluids at home, and keeping hot fluids out of children's reach.

Radiation Burns

Radiation burns of the eyes are caused by ultraviolet rays, infrared rays, X-rays, microwaves, laser beams, and gamma rays. Burns caused by ultraviolet rays occur most often.

Due to the transparency of the optical media of the eye, radiation burns affect not only the superficial structures of the eye (the eyelid skin, conjunctiva, cornea), but also the deeper structures (the lens and retina) (fig. 16.22).

EYE FACTS



Different types of electromagnetic radiation by their wavelength in the order of increasing frequency and decreasing wavelength (from: http://www.pbslearningmedia.org)

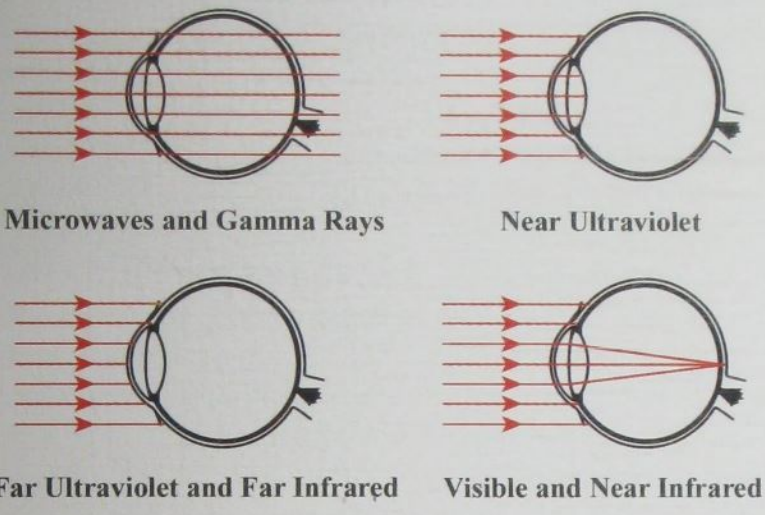


Fig. 16.22. Light-induced damage to the eye according to the wavelength (from: http://oregonstate.edu)

Ultraviolet Burns

Definition. Ultraviolet burns are radiation injuries to the eye caused by ultraviolet (UV) light.

Etiology. UV burns to the eye can be caused by exposure to UV radiation from direct sunlight, viewing of solar eclipses, or sunlight reflection off water, snow, or ice (snow blindness), and from artificial sources such as welder's arc (commonly known as flash burn, welder's flash, or an arc eye), electric sparks, suntanning beds, photographic flood lamps, or even halogen desk lamps.

Clinical Picture. More than 99 % of UV radiation is absorbed by the front of the eyes causing corneal damage.

Within a short time (for example just in a few minutes of welding without proper eye protection) intense ultraviolet light can lead to superficial punctate necrosis in the corneal epithelium that is known as photokeratitis, photo- or electroophthalmia, ultraviolet keratoconjunctivitis.

Chronic exposure to UV radiation may cause degenerative diseases of the conjunctiva (e.g., pinguecula, pterygium, squamous metaplasia, epidermoid carcinoma), may be responsible for cataract formation, be a predominant factor in the development of age-related macular degeneration, or may cause burn to the macula.

Complaints. Patients experience the onset of extreme pain, eye redness, irritation, a foreign body sensation, photophobia, blepharospasm, tearing, decreased visual acuity, and rarely temporary vision loss arised 6—12 hours after the exposure.

Signs. Eyelid edema, conjunctival hyperemia, punctate epithelial defects, diffuse corneal haze.

Methods of Examination. Medical history, external inspection, VA test, slit-lamp exam with fluorescein dye staining, ophthalmoscopy.

Differential Diagnosis. Dry eye syndrome, toxic keratopathy, acute conjunctivitis, corneal abrasion, contact lens-related disorder, superficial foreign body, chemical or thermal burns.

Treatment. The goal of UV keratitis therapy is to treat the pain associated with damage in the corneal epithelium and prevent infection.

First aid is to flush the eyes for several minutes with water or saline solution and to apply cool compresses.

NOTE!

Examination requires administration of a topical anesthetic that relieves pain and allows the patient to see clearly and open his or her eyes.

Medicamental treatment involves the following options:

— topical cycloplegics to relieve the pain from ciliary spasm (Atropine, Scopolamine);
— ophthalmic antibiotic ointments (e.g., Erythromycin, Gentamicin, or Tetracycline) to prevent infection in the damaged cornea;
— a therapeutic bandage contact lens with antibiotic eye drops (ointments dislodge the lens) to assist healing of the epithelial defect;
— topical lubricating eye drops (Hypromellose, Dexpanthenol, Solcoseryl);
— oral pain medication;
— sunglasses if the eyes are sensitive to light.

NOTE!

Topical anesthetics for UV keratitis treatment should never be used as they slow the healing of the cornea and can lead to ulcer formation.

Prognosis for visual recovery is excellent as UV keratitis is a transient condition; the recovery is within 1—2 days. Chronic UV exposure can lead to cataract development and macula burn, which may impair vision.

Complications. Secondary infection keratitis, loss of corneal sensation, vascularization, cataract, macular degeneration, decrease and loss of vision.

Prophylaxis. UV ocular burns can be prevented by wearing proper eye protection that blocks UV radiation, which includes sunglasses, snow goggles, welding helmets, or not looking directly at the sun when there is an eclipse.

Ionizing Radiation Burns

Definition. Ionizing radiation burns to the eye are ocular injuries caused by exposure to ionizing radiation.

Etiology. The sources of ionizing radiation include X-rays, gamma rays and particulate radiation (alpha, beta, gamma, and neutron radiation) produced from X-ray sets or radioactive substances.

People may be exposed to this type of radiation from different sources such as:

— medical exposure (both diagnostic and therapeutic) — X-rays, CT scans, PET (positron emission tomography), radiotherapy of tumors;

— occupational exposure — atomic reactors, mines, and radiology departments;
— non-occupational exposure — atomic bomb explosion and nuclear reactor accidents;
— natural exposure — cosmic rays.

Clinical Picture. Ionizing radiation has high energy that can cause ionization and formation of radicals in the cellular tissue resulting in characteristic types of tissue damage. Usually, this tissue damage manifests itself after a latency period, ranging from a few months up to years after exposure. Sudden exposure to severe radiation levels may cause immediate eye damage, including acute corneal erosions, perforation, and tissue death (radiation necrosis) in multiple layers of the eye, including the lens and retina.

The typical ocular presentations of injury include loss of eyelashes, eyelid pigmentation, blepharitis, dry eye syndrome, keratitis, necrosis of the conjunctiva and the cornea, transient iridocyclitis, radiation cataract and radiation retinopathy (microaneurysms, flame-shaped retinal hemorrhages, hard exudates, cotton wool spots, macular edema, vascular occlusion, retinal neovascularization, and proliferative retinopathy), optic nerve edema.

Complaints. Patients may complain of loss of eyelashes and eyebrows, ocular pain, red eye, irritation, foreign body sensation, discharge, photophobia, blepharospasm, decrease of vision, scotomas in the visual field.

Signs of ionizing radiation ocular burns depend on the type of radiation, the dose and duration of exposure and may include erythema of the eyelid skin, conjunctival hyperemia, chemosis, circumcorneal injection, watery or mucopurulent discharge, necrosis of the conjunctiva and sclera, corneal erosion, corneal necrosis, stromal edema, cataract, retinopathy, intraretinal hemorrhages, CRV thrombosis, cotton wool spots, telangiectasias (dilation of the small retinal vessels), papilledema.

Methods of Examination. Medical history, VA test, slit-lamp exam, ophthalmoscopy, funduscopy, OCT, perimetry, blood tests.

Differential Diagnosis. Other cases of acute red eye, ocular trauma due to other causative factors.

Treatment of ionizing radiation burns is symptomatic. Cycloplegics and topical antibiotic ointments or drops are used to reduce the pain of ciliary spasm, prevent synechia formation, and protect against infection. Tear substitutes and a therapeutic bandage contact lens with antibiotic eye drops (ointments dislodge the lens) may assist healing of the epithelial defect. Sometimes tarsorrhaphy may be needed.

Radiation cataract is treated surgically; radiation retinopathy — with panretinal photocoagulation with an argon laser.

Severe cases with conjunctival necrosis may require the use of contralateral autologous conjunctival flap (if the fellow eye is not affected); corneal perforation and/or radiation necrosis may require corneal transplantation (penetrating keratoplasty).

Prognosis for ionizing radiation burns is poor as poor healing is a hallmark for this type of injury.

Complications. Corneal scarring, corneal shrinkage, dry eye syndrome, loss of corneal sensation, secondary microbial keratitis, cataract, retinopathy, decrease of vision, loss of vision, high risk of cancer development.

Prophylaxis. The prophylaxis includes protection of the eye before exposure to radiation, which includes covering of the parts of the body not being treated or studied with lead screens or leaded glass. In the industrial field the staff working with radioactive material should wear appropriate protective clothing. The rooms where ionizing radiation is used must be shielded by thick concrete walls and doors with lead foil inside, have no windows and, in some cases, be arranged as a labyrinth (structural shielding).

Laser Radiation Injuries

Lasers are intense beams of light that can enter the globe of the eye. They are used therapeutically in ophthalmology to treat some ocular disorders in the anterior segment of the eye and the retina. The beam is focused directly onto the point of therapy that is exposed to the light for a specific amount of time.

Lasers are also used in industry and there are several classes of laser equipment: class 2 (laser pointers and measuring instruments), class 3B (CD and DVD writers), class 4 (most industrial, scientific, military, and medical lasers).

In spite of therapeutic effect, lasers may be dangerous to the eye (fig. 16.23). The unprotected eye is extremely sensitive to laser radiation and can be permanently damaged from direct or reflected laser beams. The extent of ocular damage is determined by laser irradiance, exposure duration, and beam size. These burns are mostly macular, with painless instantaneous and usually permanent loss of central vision.

Fig. 16.23. A European-style laser warning symbol

Protective eyewear (laser safety eyewear) in the form of goggles, glasses, and shields provides the principal means of protection against ocular injury, and must be worn at all times during laser operation or at work with laser equipment. Do not look directly at the laser beam and do not point a laser pointer at someone's eyes to prevent irreversible and untreatable macular burns.

Infrared Burns

Infrared (IR) radiation is widely presented in our life. It is found in the nature and in many industrial manufacturing processes such as steel mills, textiles, paper and glass manufacturing, or where lasers, arc lamps or electric radiant heaters are used. It is also used for medical diagnostic and therapeutic application, in mobile phones, remote controls for TV sets, modern weapons and astronomic devices, etc.

IR has a range of wavelengths, with "near infrared" being the closest in wavelength to visible light, and "far infrared" closer to the microwave region. Near infrared waves are short and not hot — in fact you cannot feel them — which makes them particularly dangerous to susceptible tissues, such as the skin and eyes. Far infrared waves are being experienced in the form of thermal energy and prolonged and intensive exposure to sources of this type of IR radiation can lead to ocular damage as well.

The main effect of IR radiation to the eyes is thermal. Exposure to intense IR radiation causes burns to the cornea and eyelid skin due to quick temperature rise in the tissues. Long-term exposure to IR radiation sources results in the development of complicated cataract and macular edema. People that work near intense IR radiation must use the appropriate eye googles or shields with filters or reflective coatings that can help to eliminate the problem.

Review:

1. Key Points

Ocular injuries according to the causative factors can be *industrial, car accidental, agricultural, domestic, children's, sports-related, military*; according to the localization — *injuries to the globe* or *ocular adnexa*; according to the mechanism of the injury — *mechanical (contact)* or *burns*; according to the extent of the mechanical trauma — *contusions (blunt trauma), non-penetrating, penetrating,* or *perforating injuries*; according to the cause of the burn — *chemical, thermal, radiation burns*; according to the grade of damage or visual outcomes — *mild, moderate, severe, very severe.*

Symptoms of ocular trauma may differ depending on the affected eye structure and severity of the injury. Sometimes it can be only cosmetic appearance, in cases of severe injuries — intense pain, photophobia, tearing, blepharospasm, foreign body sensation, loss of vision, and loss of the eye itself. The examination consists of thorough history of the trauma, external inspection, eye movements, pupil reaction to light, slit-lamp exam, eyelid eversion, ophthalmoscopy, perimetry, and investigative methods.

Almost all traumatic injuries are considered an ophthalmologic emergency and detailed trauma history, immediate and intensive treatment are essential as such injuries could lead to permanent loss of vision.

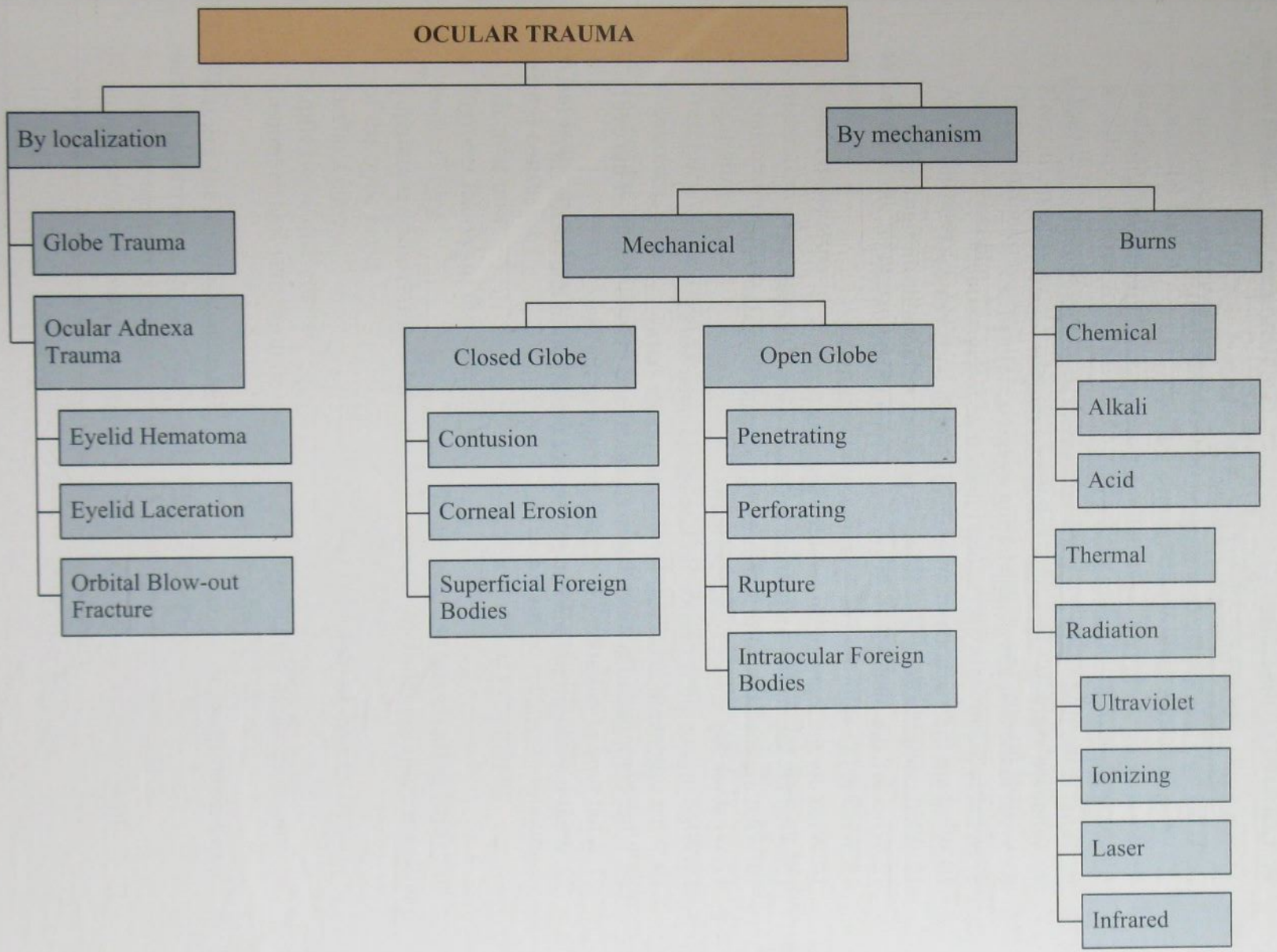
Closed eye injuries are traumas to the eye without the damage of the eye wall. It can be contusion, cornea erosion, a superficial foreign body. Contusions may be of different severity from simple black eye to severe intra-ocular disruption and globe rupture. The signs, methods of treatment, and prognosis are different according to severity.

Open globe injuries are full-thickness injuries to the eyeball with presence of entrance and exit wounds with or without an intraocular foreign body. Absolute signs of penetrating traumas are presence of a corneal or scleral wound; a hole in the iris; prolapse of the intraocular contents (the iris, ciliary body, choroid, vitreous); leakage of the intraocular humor — positive Seidel test; an intraocular foreign body; an air bleb in the vitreous. Relative signs are shallow anterior chamber; change of the pupil shape; hypotonia of the eye; lens capsule opacification; focal iris-corneal adhesion; hyphema; hemophthalmos; chemosis or subconjunctival hemorrhage.

The most frequent *injuries of the ocular adnexa* are conjunctival lacerations, eyelid hematoma and lacerations, and orbital blow-out traumas.

Ocular burns can be chemical, thermal, radiation. They can cause damage to the sclera, conjunctiva, cornea, and eyelids, as well as deeper structures inside the eye. The severity of eye burn depends on the cause of the injury, the duration of exposure to the agent, and the elapsed time before initiation of treatment. Alkali agents cause colliquative necrosis resulting in superficial ocular damage as well as damage in the anterior chamber due to penetration. Acidic agents cause coagulative necrosis resulting in corneal damage. Radiation burns affect different eye structures depending on the wavelength: UV — corneal defects; ionizing radiation — corneal radiation necrosis, cataract, retinopathy; laser — macula; infrared radiation — corneal burn, cataract, macular edema.

2. Diagrams


OCULAR TRAUMA
By localization
Globe Trauma
Ocular Adnexa Trauma
Eyelid Hematoma
Eyelid Laceration
Orbital Blow-out Fracture
By mechanism
Mechanical
Closed Globe
Contusion
Corneal Erosion
Superficial Foreign Bodies
Open Globe
Penetrating
Perforating
Rupture
Intraocular Foreign Bodies
Burns
Chemical
Alkali
Acid
Thermal
Radiation
Ultraviolet
Ionizing
Laser
Infrared

3. The Review Questions

A. Control Questions

1. What main types of ocular trauma do you know?
2. What ophthalmologic examination methods are used in ocular trauma?
3. What is the mechanism of blunt eye trauma?
4. What signs of clinical presentation of ocular contusion do you know?
5. What are the signs of ocular contusion?
6. What is the first aid in a superficial corneal or conjunctival foreign body?
7. What are absolute signs of penetrating eyeglobe injury?
8. What is the first aid in penetrating eyeglobe injury?
9. What is the first aid in retained intraocular foreign body?
10. What are the methods of intraocular foreign body removal?
11. What complications may follow a retained intraocular foreign body?
12. What are the methods of treatment of eyelid hematoma?
13. What is the algorithm of eyelid laceration suturing?
14. What are the signs of orbital blow-out fracture?
15. What are the treatment options in orbital blow-out fracture?
16. What are the clinical presentations of chemical ocular burns?
17. What are the grades of ocular burns?
18. What urgent aid is needed for chemical ocular burn?
19. What urgent aid is needed for thermal ocular burn?
20. What complications may follow chemical and thermal burns of the eye?
21. What type of radiation may cause damage to the eye?
22. What is photophthalmia and its clinical presentation and treatment?

B. Tests

1. **What corneal damage can occur in blunt eyeglobe trauma (contusion)?**
   A. Hemorrhage, rupture of the superficial layers
   B. Swelling, corneal ulcer
   C. Swelling, erosion, folds of Descemet's membrane
   D. Keratitis, precipitates
   E. Opacification in the form of adherent leukoma
   F. All of the above

2. **What does the term *iridodialysis* mean?**
   A. Pupil constriction
   B. Separation of the iris root
   C. Persistent pupillary membrane
   D. Pupil dilation
   E. Trembling of the iris at eye movement
   F. Tear of the iris sphincter

3. **What is the most frequent sign of retinal contusion?**
   A. Macular hole
   B. Pigment lesions of the "bony bodies" type
   C. Separation of the retina at the site of the optic nerve
   D. Berlin's edema
   E. Optic nerve disc edema
   F. Central retinal vein thrombosis

4. **What are the absolute signs of ocular penetrating injury?**
   A. Shallow anterior chamber
   B. Presence of a corneal or scleral wound
   C. Prolapse of the inner structures of the eye
   D. A hole in the iris
   E. An intraocular foreign body
   F. An air bleb in the vitreous

5. **What are the relative signs of a penetrating ocular injury?**
   A. Shallow anterior chamber
   B. Change of the pupil shape
   C. Prolapse of the inner structures of the eye
   D. Hypotony of the eye
   E. Hyphema
   F. Subconjunctival hemorrhage

6. **When an intraocular foreign body is suspected, all methods are used for diagnostics EXCEPT:**
   A. Biomicroscopy
   B. Review X-ray
   C. X-ray with the Komberg—Baltin localizer
   D. Ultrasound scanning
   E. Magnetic resonance imaging
   F. Computed tomography

7. **First aid in penetrating injury of the eye includes complex measures EXCEPT:**
   A. Instillation of disinfecting eyedrops
   B. Instillation of anesthetizing eyedrops
   C. Removal of the prolapsed eye structures
   D. Tetanus prophylaxis
   E. Systematic instillation of antibiotics
   F. Binocular bandage

8. What is sympathetic ophthalmia?

A. Acute iridocyclitis after a penetrating injury
B. Uveitis after eye contusion
C. Chronic uveitis of the intact eye after a penetrating injury of the other eye
D. Acute inflammation of the inner structures after an eye penetrating injury
E. Acute uveitis of the intact eye after contusion of the other eye
F. Acute inflammation of all structures of the eyeglobe

9. The methods of first aid for chemical ocular burns are:

A. Instillation of anesthetizing eyedrops
B. Irrigation with a sterile isotonic saline or water
C. Removal of chemical substance remains
D. Instillation of antibiotic ointments
E. Treatment for tetanus
F. Binocular bandage

10. What are the signs of I-grade ocular burns?

A. Hyperemia of the eyelid skin and conjunctiva
B. Superficial corneal erosions
C. Eyelid skin vesicles
D. Superficial corneal opacity
E. Necrosis of the eyelid skin and ischemia of the conjunctiva
F. Deep necrosis of the cornea

11. What are the signs of II-grade ocular burns?

A. Hyperemia of the eyelid skin and conjunctiva
B. Superficial corneal erosions
C. Eyelid skin vesicles
D. Superficial corneal opacity
E. Necrosis of the eyelid skin and ischemia of the conjunctiva
F. Deep necrosis of the cornea

12. What are the signs of III-grade ocular burns?

A. Hyperemia of the skin and conjunctiva
B. Superficial corneal erosions
C. Eyelid skin vesicles
D. Superficial corneal opacity
E. Necrosis of the eyelid skin and ischemia of the conjunctiva
F. Deep necrosis of the cornea

13. What are the signs of IV-grade ocular burns?

A. Hyperemia of the skin and conjunctiva
B. Superficial corneal erosions
C. Eyelid skin vesicles
D. Superficial corneal opacity
E. Necrosis of the eyelid skin and conjunctival ischemia
F. Deep necrosis of the cornea

14. What are the complications of ocular burns?

A. Corneal opacities

B. Cataract
C. Secondary angle-closure glaucoma
D. Eyelid contracture (entropion, ectropion)
E. Symblepharon
F. Atrophy of the eyeball

15. What is electroophthalmia?

A. Damage to the eye due to infrared waves
B. Damage to the eye due to X-ray waves
C. Damage to the eye due to ultraviolet waves
D. Damage to the eye due to ultrasound waves
E. Damage to the eye due to laser waves
F. Damage to the eye due to exposure to a thermal factor

C. Clinical Cases

Case 1

A 65-year-old patient presents with severe left eye pain, foreign body sensation, tearing, photophobia and blurry vision after being kicked in the left eye by his grandson 3 hours ago. Examination findings: Vis OD = 1.0; Vis OS = 0.4; does not improve with correction. IOP OD 18 mm Hg. IOP OS 16 mm Hg. Conjunctiva/sclera: OD — white and quiet; OS — conjunctival injection, no fluorescein staining. Cornea: OD — normal; OS — 3 by 2 mm corneal epithelial defect with fluorescein staining. Iris is not changed OU. Fundus examination shows no changes OU. What is the diagnosis and first treatment options?

Case 2

A 16-year-old patient complains of severe pain, redness, tearing, visual decrease in the left eye that he started to develop 5 days ago after a blunt ocular trauma. Examination findings: Vis OD = 0.1 cc sph-2.0 = 1.0. Vis OS = 0.05, does not improve with correction. OD — anterior segment of the eye without changes, the lens is decentered so that the inferior equator is visible in the pupil. No changes of the retina at fundus examination. OS — edema of the upper eyelid, congestive injection of the conjunctiva, the cornea is swollen and rough, its sensation is decreased. The anterior chamber is deep and filled with a spherical-shape transparent body. The other structures of the eye are not visible. IOP 50 mm Hg. The patient is very tall and has long extremities. What are the diagnosis and first treatment options?

Case 3

A 5-year-old girl presented to the emergency department after falling from the bed on a glass nightstand which shattered upon impact. The mother checked her and found no bruises or bleeding. The next morning the mother noticed the patient rubbing her right eye several times. Examination findings: the OD pupil is irregular,

peaked to the limbus. External examination reveals no foreign bodies. Slit-lamp exam: diffuse hyperemia of the conjunctiva, at 1 o'clock a 3 mm corneal laceration, the anterior chamber is shallow, a small amount of iris peaked to the wound. The lens and vitreous are clear, fundus exam shows no changes. What are the diagnosis and first aid options?

Case 4

A 42-year-old male patient presented to a clinic reporting a sore and red left eye shortly after being elbowed during a sports game. He reports blurry vision, tenderness over his left eye, eyebrow, and cheek bone. Examination: Vis OD = 1.0. Vis OS = 0.1, does not improve with correction. Confrontational visual fields constricted inferiorly OS, IOP: 19 mm OD, 28 mm OS. External: OS no lid lacerations. Slit-lamp exam: mild diffuse conjunctival hyperemia, no corneal epithelial defects, blood obscuring 1/3 of the anterior chamber. The iris is normal, round, no obvious neovascularization. The lens: no evidence of cataract or dislocation. The vitreous and fundus exam shows no blood and edema. What are the diagnosis and treatment options?

Case 5

A 31-year-old patient, who is an auto-mechanic, complains of burning pain in the left eye and surrounding skin that started after something exploded and sprayed onto his face while he was working with a car engine. He reported that his co-workers had performed a 10-minute irrigation of his eyes before the ambulance arrived. Examination: Vis OD = 1.0. Vis OS = 0.1, does not improve with correction. Slit-lamp exam: OD — the conjunctiva and sclera are not affected, the cornea is clear, the anterior chamber is deep, the other parts show no changes. OS — hyperemia and swelling of the eyelids, mixed injection and chemosis of the conjunctiva, corneal edema, epithelial defects with fluorescein, mild cell reaction in the anterior chamber. What are the diagnosis and first aid options?

Case 6

A 17-year-old male presented with double vision with both eyes open, pain when attempting to look up, nausea and vomiting after a blunt trauma to his left eye when fighting. Examination: Vis OU = 1.0. Slit-lamp exam: OS — minimal periorbital edema, small subconjunctival hemorrhages; IOP OD — 20 mm Hg, OS — 19 mm Hg. Exophthalmometry: OD — 19 mm, OS — 16 mm. Ocular motility: left hypotropia in primary gaze. OD: full motility, OS: significantly reduced supraduction, pain when looking up and restriction on forced supraduction OS compared with OD. Fundus OU — normal. CT — left inferior orbital floor fracture with entrapment of the intraorbital fat and left inferior rectus muscle. What are the diagnosis and treatment options?

Case 7

A 36-year-old male patient presented with right eye pain, tearing and blurry vision. He was pounding a metal object with a metal chisel without wearing safety glasses

and felt that something striked his right eye. The accident happened a day before he came to the hospital, his vision decreased severely. Examination: Vis: OD — hand movements, OS — 1.0; IOP OD — 16 mm Hg, OS — 17 mm Hg. OD slit-lamp exam: mild injection of the conjunctiva, full-thickness corneal laceration, iris defect, peaked pupil, dense cataract with disruption of the anterior lens capsule, no view of the funduds. Ultrasound of the right eye revealed mild vitreous hemorrhage, a highly reflective object lying posteriorly near the optic nerve with shadowing. What are the diagnosis and treatment options?

Case 8

A 24-year-old male patient presented with a history of a cement burn to his right eye. He was working on a building site and while mixing cement some of the cement powder entered his eye. He immediately washed out the cement with water and was brought to the hospital. He complained of severe ocular irritation, pain, and a decrease in visual acuity. Before examination residual cement particles were removed and his eye was irrigated until pH was 7. After first aid administration: Vis OD — 0.2, OS — 1.0; OD — edema of the eyelid skin, the conjunctiva is anemic, three to seven clock hours of limbal ischemia, entire stroma is opaque, iris details are not visible, IOP — 19 mm Hg. What are the diagnosis and the following treatment options?

Case 9

A 14-year-old female patient presented with severe ocular and facial pain, reduction in vision, tearing, photophobia, and redness in both eyes following an accidental splash of boiling water into her face. After immediate irrigation with cold water she was rushed to the hospital. On examination: Vis: OD — 0.2, OS — 0.1; facial scald, bilateral periorbital edema, worse on the left, blepharospasms, and diffuse conjunctival hyperemia without limbal ischemia; corneal ulcers stained with fluorescein, involving the whole right cornea and two-thirds of the left cornea on slit-lamp examination. The pupillary activity, the lenses, and the fundi appeared normal. What are the diagnosis and first aid options?

Case 10

A 35- year-old male boxer complains of considerable visual decrease, bruises around the left eye. He informs of being hit with a fist during a training 5 hours ago. Examination: Vis OD = 1.0. Vis OS = 0.04, does not improve with correction. Slit-lamp exam: OD — the conjunctiva and sclera are not affected, the cornea is clear, the anterior chamber is clear and of normal depth, the lens is located typically; no pathologic findings in the retina with ophthalmoscopy. OS — eyelid hyperemia, swelling. Diffuse redness over the conjunctiva; the cornea and anterior chamber are clear, the lens is located typically, and the vitreous is transparent. Ophthalmoscopic examination: the retina in the posterior region is milky-white, edematous, the optic nerve is of normal appearance. What is the full diagnosis? What is the cause of visual decrease? What is the prognosis for the patient?

Case 11

A 42-year-old construction worker complains of severe pain, burning, tearing, and visual decrease in both eyes that he started to experience last night after helping a friend with welding works. Examination: Vis OD = 0.5, does not improve with correction. Vis OS = 0.3, does not improve with correction. OU — eyelid edema, blepharospasm, mixed injection of the conjunctiva, the cornea is swollen and staining diffusely with fluorescein. The anterior chamber is clear, the iris is normal, the lens and vitreous body are transparent, and the retinal structures are not changed. What are the diagnosis and its treatment? How could the patient prevent the disease?

CHAPTER 17

Ocular Emergencies

OBJECTIVES

Upon completion of the chapter the students should:
- know the most common cases of ophthalmic emergencies;
- know the clinical picture of different types of ocular emergencies;
- differentiate ocular emergency conditions;
- be able to render first aid in cases of ocular emergencies.

Plan:

1. **Acute Angle-Closure Glaucoma**
2. **Acute Bacterial Conjunctivitis**
3. **Acute Dacryoadenitis**
4. **Acute Dacryocystitis**
5. **Acute Iridocyclitis**
6. **Adenoviral Conjunctivitis**
7. **Allergic Conjunctivitis**
8. **Central Retinal Artery Occlusion**
9. **Central Retinal Vein Occlusion**
10. **Chemical Burn**
11. **Corneal Abrasion**
12. **Corneal Ulcer Serpens**
13. **Endophthalmitis**
14. **Eyelid Hematoma**
15. **Eyelid Laceration**
16. **Herpes Simplex Keratitis**
17. **Hyphema**
18. **Neonatal Dacryocystitis**
19. **Optic Neuritis**
20. **Orbital Phlegmon**
21. **Orbital Blow-Out Fracture**
22. **Panophthalmitis**
23. **Penetrating Eyeball Injuries**
24. **Superficial Foreign Bodies**
25. **Thermal Burn**
26. **Ultraviolet Burn**
27. **Vitreous Hemorrhage**

Content:

Ocular emergencies are medical conditions that involve sudden threats to the visual system that if not promptly recognized and treated can result in permanent blindness or even loss of the eye. It is of great importance that every general practitioner and health care staff member could recognize the associated signs and symptoms of such emergencies and provide first aid before referring to an ophthalmologist for following treatment.

Common eye emergencies may present as an acute red eye, sudden visual loss or ocular trauma. Acute red eye can present as acute conjunctivitis, keratitis, iridocyclitis, and acute-angle closure glaucoma. Sudden visual loss can be due to retinal detachment, central retinal artery occlusion, retinal vein occlusion, vitreous hemorrhage, and optic neuritis. Ocular trauma can be due to mechanical injuries, chemical, thermal or ultraviolet burns. Other painful or painless ocular conditions such as acute dacryoadenitis and dacryocystitis, endophthalmitis and panophthalmitis have a potential risk of vision decrease and development of complications; orbital phlegmon can be life-threating.

Ocular emergencies may be broadly classified according to the urgency of referrals as:

— true emergencies (must be attended to within minutes) — chemical burns, central retinal artery occlusion, corneal foreign body;
— very urgent (must be attended to within a few hours) — acute-angle closure glaucoma, open globe injuries, orbital phlegmon, etc.;
— urgent (must be attended to within one day) — orbital fracture, blunt trauma, acute conjunctivitis, etc.

For example, chemical burns require immediate help even before history taking and also ocular examination as it can lead to loss of the eye. Most other ocular emergencies will require initial basic examination and primary management before referral to an ophthalmologist.

The following table presents a summary of common ocular emergencies and algorithms of first aid administration. Please refer to the given page number for detailed description of the etiology, clinical picture, examination methods, differential diagnosis, treatment options and prognosis of the stated conditions.

Table

Ocular Emergencies

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
1	Acute Angle-Closure Glaucoma	*Symptoms* — severe eye pain — red eye — haloes around lights — severe worsening of vision — blurry vision — headache — nausea, vomiting *Signs* — vision reduced to counting fingers — conjunctival and ciliary (mixed) injection — fixed dilated pupil — corneal edema — shallow anterior chamber — high IOP (up to 35—60 mm Hg) — edema of the optic nerve disc	Preliminary emergency treatment involves: Topical treatment: — *Pilocarpine 1 %* eye drops every 15 min for an hour, followed by instillation every hour. Further instillation rate is reduced to 6 times a day. — *Timolol 0.25—0.5 %* 2 times a day. — *Dorzolamide 2 %* 3 times per day or *Brinzolamide 1 %* 2 times per day. — *Dexamethasone sodium phosphate 0.1 %.* Urgent referral to an ophthalmologist for a thorough medical evaluation and further management that may include: Systemic treatment: — *Acetazolamide 500 mg* intravenously; it may be given orally but the onset of action is not rapid, 2—3 times per day. — *Mannitol 20 % 1—2 g/kg* intravenously, or *Glycerol 50 % 1—1.5 g/kg* orally. — Analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* Revulsive procedures are indicated along with drug therapy: hot foot baths, mustard plasters on the gastrocnemius muscles, salt laxatives and hirudotherapy (leeches to the temple). Laser or surgical treatment is indicated if the attack is not resolved within one day	393

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
2	Acute Bacterial Conjunctivitis	*Symptoms* — red eye — discomfort and foreign body sensation — mild photophobia — yellow or green purulent discharge — slight blurring of vision *Signs* — diffuse conjunctival hyperemia — bulbar conjunctival injection — lid edema — flakes of pus in the canthi and on the lid margins — the cilia stick together with yellow crusts	— Cleaning of the eye by irrigation with normal saline to remove the purulent discharge and crusts or stickiness. — Topical broad-spectrum antibiotics *(Ofloxacin 0.3 %, Levofloxacin 0.5 %, Tobramycin 0.3 %, and Gentamicin 0.3 %)* three to four times a day for five to seven days. Referral to an ophthalmologist for a following observation and treatment	226
3	Acute Dacryoadenitis	*Symptoms* — pain and swelling in the outer part of the upper eyelid — tenderness, warmth and pain increase upon pressing the swollen region — ocular motility restriction — pain on moving the eye *Signs* — conjunctival injection — chemosis — erythema of the eyelids — swelling of the lateral third of the upper lid (S-shaped lid) — proptosis — palpebral lobe of the lacrimal gland is seen when the upper lid is everted	— Disinfectant instillation *(Furacilin 1:1500, Sulfacetamide 30 %).* — Topical antibiotic ointments *(Tobramycin 0.3 %, Tetracycline 1 %).* — Topical NSAIDs *(Diclofenac 0.1 %).* — UHF-therapy. — Systemic antibiotics *(Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg).* — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* The patient must be referred to an ophthalmologist for a following observation and treatment	179

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
4	Acute Dacryocystitis	*Symptoms* — 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 expressed from the punctum	— Systemic antibiotics *(Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg).* — Topical antibiotics according to the specific pathogens detected *(Levofloxacin 0.5 %, Ofloxacin 0.3 %, Ciprofloxacin 0.3 %, Tobramycin 0.3 %).* — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* Referral to an ophthalmologist for a following observation and treatment that may include lacrimal pathway flushing with saline solution or furacilin	184
5	Acute Iridocyclitis	*Symptoms* — severe eye pain — red eye — blurry vision — significant sensitivity to light — tearing *Signs* — ciliary injection (circumlimbal flush) — constricted pupil (miosis) — discoloration of the iris — keratic precipitates — flare and cells in the anterior chamber — posterior synechiae	— Mydriatic eye drops *(Atropine 1 %* or *Tropicamide 1 %).* — Topical steroids *(Prednisolone acetate 1 %* or *Dexamethasone sodium phosphate 0.1 %)* every 2 hours while awake. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* Urgent referral to an ophthalmologist for a thorough medical evaluation and further management	320

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
6	Adenoviral Conjunctivitis	*Symptoms* — red eye — itching and burning — foreign body sensation — extreme sensitivity to light — watery discharge — decreased vision — swelling of the eyelids — sore throat, runny nose *Signs* — eyelid edema — pseudoptosis — conjunctival hyperemia — follicular conjunctivitis — chemosis — subconjunctival hemorrhage — palpable periauricular lymphadenopathy — respiratory infection	— Cold compresses. — Topical antiviral medications *(Interferon)* 6—8 times a day; ointment 2—3 times a day. — Lubricants *(Polyethylene Glycol 0.4 %, Propylene Glycol 0.3 %, Hypromellose 0.3 %, Carbomer 0.2 %)*. — Systemic antihistamine *(Mebhydrolin 0.05—0.1 g, Chloropyramine hydrochloride 25 mg, Loratadine 10 mg)* and vitamin medications. Referral to an ophthalmologist for a following observation and treatment if needed	232
7	Allergic Conjunctivitis	*Symptoms* — red eye — itching — severe burning — foreign body sensation — extreme sensitivity to light — watery discharge — heaviness of the lids *Signs* — conjunctival injection — giant "cobblestone" papillae on the upper tarsal conjunctiva — gelatinous nodules on the limbus — signs of keratitis	— Cold compresses. — Lubricants *(Polyethylene Glycol 0.4 %, Propylene Glycol 0.3 %, Hypromellose 0.3 %, Carbomer 0.2 %)*. — Topical antihistamines *(Cromoglicate 2 %, Lodoxamide 0.1 %, Cromoglycate 2 %, Olopatadine 0.1 %)*. — Topical NSAIDs *(Diclofenac 0.1 %, Indometacine 0.1%)*. — Systemic antihistamine *(Mebhydrolin 0.05—0.1 g, Chloropyramine hydrochloride 25 mg, Loratadine 10 mg)*. Referral to an ophthalmologist for a following observation and treatment if needed	236

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
8	Central Retinal Artery Occlusion	*Symptoms* — sudden painless loss of vision in one eye, or — fallout in the visual field in one eye *Signs* — relative afferent pupillary defect — pale retina with vessel attenuation — milky-white ischemic edema of the posterior pole — opaque fundus with a red fovea (a cherry-red spot) — pale optic disc	Preliminary emergency treatment involves: — Adoption of a supine posture. — Firm ocular massage intermittently through closed eyelids for at least 15 minutes. — Sublingual administration of *Nitroglycerin*. — *Acetazolamide 500 mg* intravenously and/or topical *Timolol 0.5 %*. — *Papaverine 40 mg* intramuscularly. — *Mannitol 20 %* or *Glycerol 50 %* intravenously. — Inhalation of 95 % oxygen and 5 % carbon dioxide mixture for 10 minutes or the patient is asked to breathe into a paper bag firmly applied around the mouth and nose. Urgent referral to an ophthalmologist for a thorough medical evaluation and further management that may include: paracentesis of the anterior chamber, panretinal photocoagulation, transluminal Nd:YAG laser embolysis, thrombolysis (extrapolating from the treatment of stroke and myocardial infarction), high dosage of intravenous steroids for patients with giant cell arteritis	415

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
9	Central Retinal Vein Occlusion	*Symptoms* — sudden painless decrease or loss of vision in one eye — "veil" in front of the eyes — floaters *Signs* — relative afferent pupillary defect — abnormal red reflex — tortuosity and dilatation of retinal veins — diffuse retinal hemorrhages — cotton wool spots — retinal edema — retinal neovascularization	Preliminary emergency treatment involves: — *Aminophylline 2.4 %* 5—10 ml in Sol. *Glucose 20 %* 20 ml intravenously (or *Aminophylline 24 %* 0.5—1.0 ml intramuscularly). — Solution *Magnesium Sulfate 25 %* 10 ml + *Papaverine 40 mg* + *Dibasol 2 %* 0.5 ml intramuscularly (in case of high blood pressure). — Anticoagulants intramuscularly: *heparin* 10 IU in 0.5—1 h after administration of *Aminophylline*. — *Heparin* 0.5 IU + *Fibrinolizin* 1 IU + *Dexamethasone 0.5 ml* parabulbar. — *Vitamins B, C, E.* — Leeches on the temple area, hot foot bath. Urgent referral to the ophthalmology department for a thorough medical evaluation and further management that includes treatment of the underlying medical condition and topical treatment: laser photocoagulation, intravitreal corticosteroids (*Triamcinolone Acetonide 0.1 %*), intravitreal anti-VEGF agents *(Ranibizumab 10 mg/ml, Bevacizumab 25 mg/ml, Aflibercept 40 mg/ml)*, focal laser therapy, pan-retinal photocoagulation and intraocular steroids	418

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
10	Chemical Burns	*Symptoms* — severe pain — eye redness — foreign body sensation — excessive tearing — photophobia — blepharospasm — blurred vision — swelling of the eyelids *Signs* — burns to eyelids and surrounding skin — conjunctival chemosis and hyperemia — conjunctival chemosis — limbal and conjunctival ischemia — corneal epithelium defects of different range — corneal edema — corneal opacification ("dim glass", "porcelain glass") — conjunctival and corneal necrosis — corneal melting	— *Irrigation* with sterile isotonic saline or with any watery solution of neutral pH for at least 30 minutes, until ocular surface pH is neutralized to a range of 7.0—7.4 and removal of chemical agents if present. — For alkaline burns — weak acidic solutions (e.g. boric acid), ethylendiamine tetra acetic acid (EDTA) 1 % solution for caustic lime can be used. For acid burns — weak alkaline solutions (e.g. sodium bicarbonate). — Antibiotic ointments *(Tetracycline 1 %, Levofloxacin 0.5 %, Ofloxacin 0.3 %, Ciprofloxacin 0.3 %, Tobramycin 0.3 %).* — Monocular soft bandage. — Tetanus prophylaxis if not current. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* Only after these actions have been taken the patient must be transported to an ophthalmologist or an eye clinic for the following treatment	498

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
11	Corneal Abrasion	*Symptoms* — sharp pain — foreign body sensation — tearing — photophobia — blepharospasm — blurry vision *Signs* — light conjunctival injection — epithelial defects seen with ultraviolet light after fluorescein staining	— Antibacterial drops or ointments *(Ofloxacin 0.3 %, Tobramycin 0.3 %, Levofloxacin 0.5 %, Tetracycline 1 %).* — Re-epithelization drugs *(Dexpanthenol 5 %, Solcoseryl, Carbomer 0.2 %, Hypromellose 0.3 %, Polyethylene Glycol 0.4 %, Propylene Glycol 0.3 %).* — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* Referral to an ophthalmologist for a following observation and treatment if needed	257
12	Corneal Ulcer Serpens	*Symptoms* — severe eye pain — red eye — photophobia — profuse tearing — blepharospasm — purulent discharge — significant decrease in vision *Signs* — mild lid edema — conjunctival hyperemia — chemosis — ciliary injection — a grey-white or yellow-grey disc-shaped ulcer on the cornea with one edge undermined the corneal stroma — hypopyon	— Topical broad-spectrum antibiotics *(Ofloxacin 0.3 %, Levofloxacin 0.5 %, Lomefloxacin 0.3 % and Gentamicin 0.3 %)* — As soon as the agent that causes the ulcer has been identified — topical antibiotics, anti-fungal agents *(Natamycin 5 %)*, or topical antivirals *(Acyclovir 3 %).* — Systemic antibiotics *(Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg).* — Cycloplegics *(Atropine sulfate 1 %, Tropicamide 1 %, Scopolamine 0.25 %).* — Re-epithelization drugs *(Carbomer 0.2 %, Dexpanthenol 5 %, Solcoseryl 20 %).* — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* — Systemic antihistamines *(Mebhydrolin 0.05—0.1 g, Chloropyramine hydrochloride 25 mg, Loratadine 10 mg).* — Vitamins. Referral to an ophthalmologist for the following observation and treatment if needed	261

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
13	Endophthalmitis	*Symptoms* — swelling of the eyelid — red eye — severe eye pain and irritation — extreme sensitivity to light — tearing — decreased vision — headache — nausea or vomiting *Signs* — decreased VA — ciliary injection — chemosis — corneal edema — keratic precipitates — cells and flare in the anterior chamber — hypopyon — decreased red reflex — vitreous cells — cotton wool spots — proptosis — decreased IOP	Urgent referral to an ophthalmology department for aggressive treatment. — Intravitreal antibiotics *(Vancomycin 1 mg/0.1 ml, Amikacin 0.4 mg/0.1 ml, Ceftazidime 2.25 mg/0.1 ml, Ceftazidime 2 mg/0.1 ml).* — Systemic broad-spectrum antibiotics *(Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg, Cefazolin 0.5—1.0 g, Tobramycin 40 mg/ml).* — Intraocular anti-inflammatory agents and topical steroid therapy *(Prednisolone acetate 1 %, Dexamethasone 0.1 %).* — Topical mydriatics *(Atropine sulfate 1 %, Tropicamide 1 %)* — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* *Vitrectomy* may be performed to remove infectious material from the inside of the eye if infection is severe. *Evisceration* might be performed when the eye cannot be saved and the patient is completely blind with no perception of light. Intravitreal and systemic *Amphotericin B* in case of fungal endophthalmitis	326

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
14	Eyelid Hematoma	*Symptoms* — bruising and swelling of the eyelid — painful eyelid — difficulty in opening the eye *Signs* — hematoma of the eyelids — swelling of the eyelids	— Cold compresses for about 15 to 20 minutes at a time reapplied every hour for the first day. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* — Resorptive ointments *(Actovegin 5 %, Heparin 1 g)*	491
15	Eyelid Laceration	*Symptoms* — laceration of the eyelid (margin, skin or total) — bleeding — periorbital pain *Signs* — visible partial or full-thickness laceration of the eyelid	— Cleaning of the wound of all foreign particles and irrigating with saline solutions. — Antibiotic ointments *(Tetracycline 1 %, Gentamicin 0.3 %).* — Tetanus prophylaxis if not current. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* Cover the eye with a sterile bandage or cloth and send the patient to an ophthalmologist for surgical repair	493

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
16	Herpes Simplex Keratitis	*Symptoms* — severe pain in and around the eye — open sores/blisters around the eye — swelling of the eyelids — red eye — irritation and itching — foreign body sensation — excessive tearing — sensitivity to light — blurring of vision — haloes around light sources *Signs* — eyelid edema — corneal injection — reduced corneal sensitivity — photophobia — decreased VA — dendritic epithelial defects with terminal bulbs that stain with fluorescein	Urgent referral to an ophthalmologist for aggressive treatment: — 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. For immunocompromised patients — intravenous *Acyclovir* (5 mg/kg IV every 8 hours for 7 to 14 days). — 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))	263

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
17	Hyphema	*Symptoms* — history of trauma — bleeding in the eye — eye pain — photosensitivity — blurred or cloudy vision *Signs* — blood in the anterior chamber — raised IOP	— Strict bed rest with head elevated by 30°—45°. — Initially limit any eye movement with a binocular eye bandage. — Cold compresses. — Haemostatic therapy *(Etamsylate 250—500 mg)*. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)*. Referral to an ophthalmologist for anterior chamber paracentesis and blood draining if no effect after 4—5 days	477
18	Neonatal Dacryocystitis	*Symptoms* — redness and swelling at the inner corner of the eye — tearing — 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	— Massage of the lacrimal passages. — Instillation of disinfectants and antibiotic drops. Referral to an ophthalmologist if the blockage is not resolved after several weeks to months for following treatment that may include forceful lacrimal irrigation, surgical probing and other surgical methods	188

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
19	Optic Neuritis	*Symptoms* — sudden painless loss of central vision in one eye — reduced perception of color — pain at eye movement — headache *Signs* — relative afferent pupillary defect — central or paracentral scotoma — optic disc hyperemia with blurred disc margin — decreased color vision	Referral to an ophthalmologist and neurologist for complete evaluation and treatment of suspected optic neuritis causes. Initial treatment includes: — *Methylprednisolone* 250 mg intravenously for 3 days followed by oral *Prednisolone (1 mg/kg/day)* for 11 days. — Vitamins B_1, B_6, B_{12}, C in high doses	437
20	Orbital Phlegmon	*Symptoms* — painful swelling of the eyelids — bulging of the eye — eye pain — decreased vision — double vision — painful or difficult eye movements — headache — fever *Signs* — infiltration and hyperemia of the eyelids — immobility of the eyelids — exophthalmos — full ophthalmoplegia — chemosis — decrease of vision	Admission to the hospital for close observation and consultation by an ophthalmologist and an ENT specialist. Initial treatment includes: — Intravenous broad-spectrum antibiotics in high doses up to 10 days *(Ceftriaxone 1 g, Cefuroxime 750 mg, Gentamicin 40 mg/ml)*. — Topical NSAIDs *(Diclofenac 0.1 %, Indometacin 1 ml)*. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)*. — Systemic antihistamine *(Mebhydrolin 0.05—0.1 g, Chloropyramine hydrochloride 25 mg, Loratadine 10 mg)*. Surgery to drain the orbital and sinus infection if the condition does not improve after 2 or 3 days of intravenous antibiotic therapy	204

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
21	Orbital Blow-Out Fracture	*Symptoms* — bruising — tenderness and swelling around the eye — red eye — facial numbness on the side of the trauma — nose bleeding — restriction and pain at eye movements — double vision *Signs* — restriction of eyeball movements — subcutaneous periorbital emphysema — hypoesthesia in the infraorbital or supraorbital region — displacement of bony fragments at palpation — enophthalmos — syndrome of superior orbital fissure (ophthalmoplegia, ptosis, mydriasis, accommodative paresis, corneal sensitivity decrease)	Preliminary treatment involves: — Ice pack to the periorbital region for 24—48 hours. — Nasal vasoconstrictors for 2 to 3 days. — Instructing the patients not to blow the nose. — Tetanus prophylaxis if not current. Referral to an ophthalmology department for a thorough medical evaluation, management, and surgical treatment. — Systemic antibiotics (*Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg)* within 10—14 days. — Systemic steroids *(Dexamethasone 0.5 mg)*. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)*. — Surgical treatment	496

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
22	Panophthalmitis	*Symptoms* — rapid swelling of the eyelids — red eye — severe ocular pain — excessive tearing — purulent discharge — painful and limited ocular movement — complete loss of vision — headache — fever — vomiting *Signs* — eyelid edema — proptosis — ciliary and conjunctival injection — chemosis — corneal edema and haziness — massive pus in the anterior chamber and vitreous — high IOP — perception of light is absent	Urgent referral to an ophthalmology department for aggressive treatment. — Intravitreal antibiotics *(Vancomycin 1 mg/0.1 ml, Amikacin 0.4 mg/0.1 ml, Ceftazidime 2.25 mg/0.1 ml, Ceftazidime 2 mg/0.1 ml).* — Systemic broad-spectrum antibiotics *(Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg, Cefazolin 0.5—1.0 g, Tobramycin 40 mg/ml).* — Topical, subconjunctival injection and systemic administration of corticosteroids *(Prednisolone acetate 1 %, Dexamethasone 0.1 %).* — Topical mydriatics *(Atropine sulfate 1 %, Tropicamide 1 %).* — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg).* — Vitrectomy may be performed to remove infectious material from the inside of the eye. — The eye is eviscerated if it cannot be saved and the patient is completely blind with no perception of light	327

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
23	Penetrating Eyeball Injuries	*Symptoms* — sharp pain — irritation — foreign body sensation — tearing — photophobia — blepharospasm — blurry vision *Signs* Absolute, or reliable signs: — presence of corneal or scleral wound — hole in the iris — prolapse of the intraocular contents (iris, ciliary body, choroid, vitreous) — leakage of the aqueous humor — intraocular foreign body — air bleb in the vitreous Relative, or suggestive signs: — the anterior chamber is shallow, or absent, or too deep (shallow — in corneal lacerations, deep — in scleral) — change of the pupil shape (drop-shaped and displaced toward the penetration wound) — hypotony of the eye — punctate lens capsule defect or opacification — focal iris-corneal adhesion — bleeding in the anterior chamber (hyphema) and vitreous body (hemophthalmos) — marked conjunctival edema (chemosis) or subconjunctival hemorrhage	Initial treatment options involve: — Topical anesthetics *(Proxymetacain 0.5 %, Tetracaine 0.5 %, Dikain 0.3 %).* — Disinfectant solutions *(Sulfacetamide 30 %, Myramistin 0.01 %).* — Antibiotic drops *(Ofloxacin 0.3 %, Tobramycin 0.3 %, Levofloxacin 0.5 %).* — Tetanus prophylaxis if not current. — Binocular bandage (the injured eye — with a protective shield, uninjured eye — with a pressure patching to prevent excessive movement of the injured eye). — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)* may be needed. After first aid administration, the patient must be transported in supine position (if possible) to the nearest specialized ophthalmic trauma department for surgical treatment. ! DO NOT attempt to remove any dark colored material from the ocular surface as that may be prolapsed intraocular contents	484

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
24	Superficial Foreign Bodies	*Symptoms* — acute pain — foreign body sensation — tearing — photophobia — blepharospasm — blurry vision — pain upon blinking and eye movement *Signs* — normal or decreased VA — conjunctival or ciliary injection — visible foreign body — epithelial defect that stains with fluorescein — corneal or conjunctival edema	— Topical anesthesia *(Proxymetacain 0.5 %, Tetracaine 0.5 %, Dikain 0.3 %)* and obligatory upper eyelid eversion. — Removal of the superficial foreign body with the help of a cotton swab, a pad or a clean soft tissue afterwards. — Removal of the foreign body embedded in the corneal epithelium with the use of a sterile 25-gauge needle, a foreign body spud or a special bur after topical anesthesia. — Disinfectant solutions *(Sulfacetamide 30 %, Myramistin 0.01 %)*. — Antibiotic drops or ointments *(Ciprofloxacin 0.35 %, Ofloxacin 0.3 %, Norfloxacin 0.3%, Levofloxacin 0.5 %, Gentamicin 0.3 %, Tetracycline 1 %)*. — Re-epithelization drugs *(Carbomer 0.2 %, Dexpanthenol 5 %, Solcoseryl 20 %)*. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)*	481

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
25	Thermal Burn	*Symptoms* — severe pain — foreign body sensation — irritation — excessive tearing — photophobia — blepharospasm — blurred vision *Signs* — hyperemia of the skin and conjunctiva — superficial corneal erosions — eyelid skin vesicles — superficial conjunctival membranes — necrosis of the skin and conjunctiva — corneal opacity of different layers	First aid administration involves: — Irrigation with cold water for 20 minutes and removal of the thermal agent if present. — Topical anesthesia *(Proxymetacain 0.5 %, Tetracaine 0.5 %, Dikain 0.3 %)*. — Antibiotic ointments *(Ofloxacin 0.3 %, Tobramycin 0.3 %, Levofloxacin 0.5 %, Tetracycline 1 %, Gentamicin 0.3 %, Ciprofloxacin 0.35 %, etc.)*. — Tetanus prophylaxis if not current. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)*. In cases of second—fourth-stage burns the patient must be referred to an ophthalmology department for the next treatment options	502
26	Ultraviolet Burn	*Symptoms* — extreme eye pain — red eye — irritation — foreign body sensation — tearing — photophobia — blepharospasm — decreased vision — and rarely — temporary vision loss 6—12 hours after the exposure *Signs* — eyelid edema — conjunctival hyperemia — punctate epithelial defects with fluorescein staining — diffuse corneal haze	— Flushing of the eyes for several minutes with water or saline solution and to apply cool compresses. — Mydriatics *(Atropine sulfate 1 %, Scopolamine 0.25 %, Tropicamide 1 %)*. — Antibiotic ointments *(Gentamicin 0.3 %, Tetracycline 1 %)*. — Lubricating eye drops *(Hypromellose 0.3 %, Dexpanthenol 5 %, Solcoseryl 20 %)*. — Systemic analgesics *(Paracetamol 500 mg, Ibuprofen 200—400 mg)*	505

No	Ocular Emergencies	Clinical Picture	First Aid Algorithm	Detailed Description (Page)
27	Vitreous Hemorrhage	*Symptoms* — sudden development of floaters, or — loss of vision *Signs* — black shadows against the red reflex — poor red reflex — ophthalmoscopy shows presence of blood in the vitreous cavity	— Strict bed rest with head elevated by 30°—45°. — Limitation of any eye movement with binocular eye bandage initially. — Haemostatic and resorptive therapy *(Hyaluronidase 64 IU, Prourokinase 5000 IU, Etamsylate 500 mg, Aminocaproic acid 250 mg/ml).* Once the blood settles down — referral to an ophthalmologist for fundus examination. Vitrectomy must be performed if there is no visual improvement and when vision is reduced to only perception of light or hand movements	461

Review:

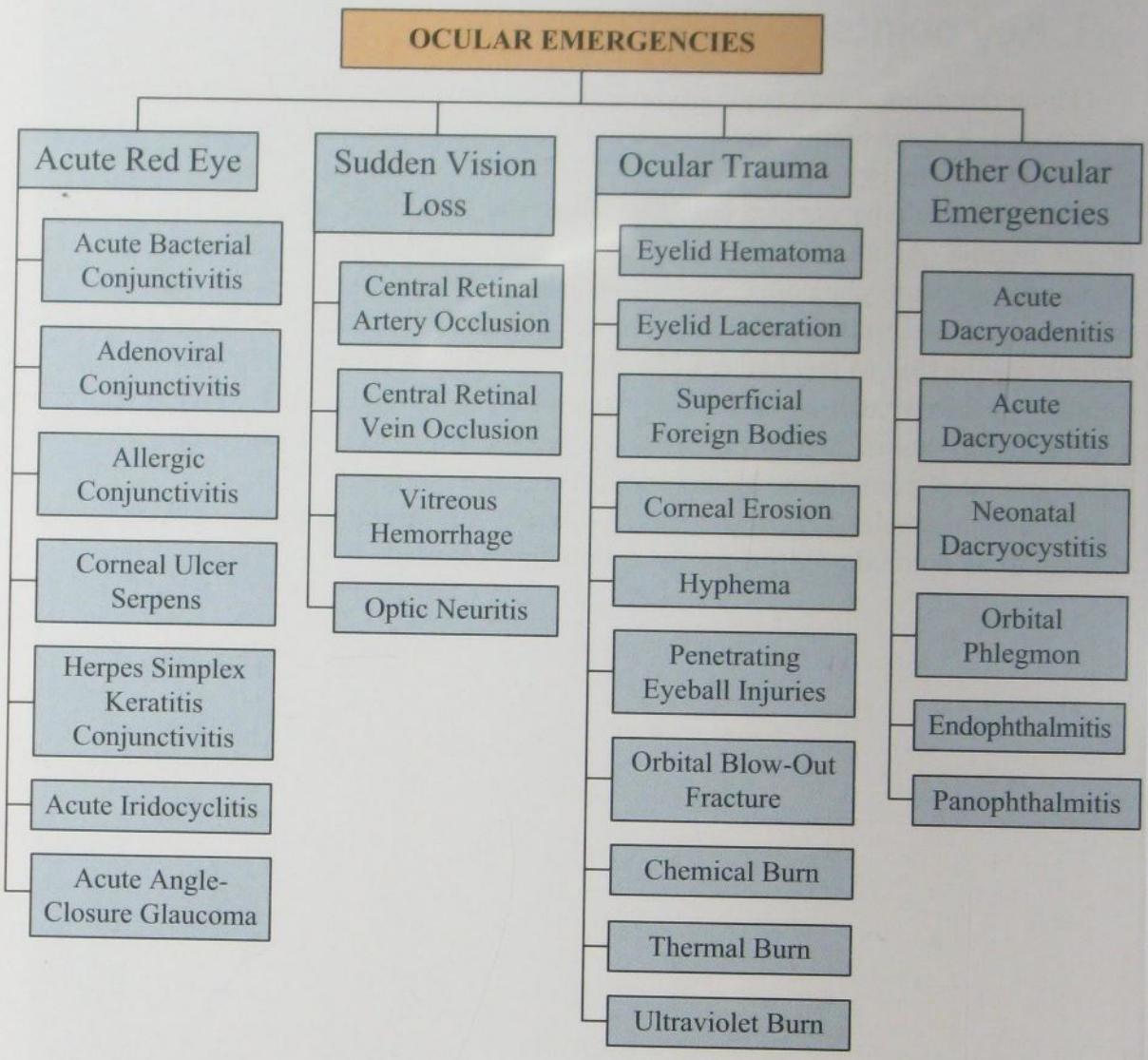
1. Key points

Ocular emergencies are medical conditions that involve sudden threats to the visual system and, if not promptly recognized and treated, can result in permanent blindness or even loss of the eye.

Common eye emergencies comprise acute red eye, sudden visual loss and acute ocular trauma. Acute red eye can present as conjunctivitis, keratitis, uveitis, and acute-angle closure glaucoma. Sudden visual loss can be due to central retinal artery occlusion, retinal vein occlusion, optic neuritis, and vitreous hemorrhage. Ocular trauma can be due to mechanical, chemical, thermal or combination injuries. Other ocular conditions such as acute dacryoadenitis and dacryocystitis, endophthalmitis and panophthalmitis, orbital phlegmon have a potential risk of vision decrease and development of complications, and require urgent treatment.

Most emergencies require ocular assessment and first aid administration before referral to an ophthalmologist for the following treatment.

2. Diagrams


OCULAR EMERGENCIES
Acute Red Eye
Acute Bacterial Conjunctivitis
Adenoviral Conjunctivitis
Allergic Conjunctivitis
Corneal Ulcer Serpens
Herpes Simplex Keratitis Conjunctivitis
Acute Iridocyclitis
Acute Angle-Closure Glaucoma
Sudden Vision Loss
Central Retinal Artery Occlusion
Central Retinal Vein Occlusion
Vitreous Hemorrhage
Optic Neuritis
Ocular Trauma
Eyelid Hematoma
Eyelid Laceration
Superficial Foreign Bodies
Corneal Erosion
Hyphema
Penetrating Eyeball Injuries
Orbital Blow-Out Fracture
Chemical Burn
Thermal Burn
Ultraviolet Burn
Other Ocular Emergencies
Acute Dacryoadenitis
Acute Dacryocystitis
Neonatal Dacryocystitis
Orbital Phlegmon
Endophthalmitis
Panophthalmitis

OCULAR EMERGENCIES
according to referral urgency

True Emergency (immediately)	Very Urgent (within a few hours)	Urgent (within one day)
— Superficial foreign bodies — Chemical burns — Central retinal artery occlusion	— Corneal erosion — Acute iridocyclitis — Acute angle-closure glaucoma — Optic neuritis — Penetrating eyeball injuries — Thermal burns — Ultraviolet burns	— Eyelid hematoma — Eyelid laceration — Acute bacterial conjunctivitis — Adenoviral conjunctivitis — Allergic conjunctivitis — Acute dacryoadenitis — Acute dacryocystitis — Neonatal dacryocystitis — Corneal ulcer serpens — Herpes simplex keratitis — Orbital phlegmon — Hyphema — Endophthalmitis — Panophthalmitis — Central retinal vein occlusion — Vitreous hemorrhage — Orbital blow-out fracture

3. The Review Questions

A. Control Questions

1. What are ocular emergencies and what common eye emergencies do you know?
2. What are clinical presentation and initial treatment options of acute angle-closure glaucoma?
3. What are the clinical presentation and initial treatment options of acute iridocyclitis glaucoma?
4. What are the clinical presentation and initial treatment options of acute conjunctivitis
5. What are the clinical presentation and initial treatment options of acute dacryoadenitis?
6. What are the clinical presentation and initial treatment options of acute dacryocystitis?
7. What are the clinical presentation and initial treatment options of central retinal artery occlusion?
8. What are the clinical presentation and initial treatment options of central retinal vein occlusion?
9. What are the clinical presentation and initial treatment options of chemical burns?
10. What are the clinical presentation and initial treatment options of corneal erosions?
11. What are the clinical presentation and initial treatment options of intraocular foreign bodies?
12. What are the clinical presentation and initial treatment options of orbital phlegmon?
13. What are the clinical presentation and initial treatment options of neonatal dacryocystitis?
14. What are the clinical presentation and initial treatment options of retinal detachment?
15. What are the clinical presentation and initial treatment options of hyphema?

B. Tests

1. Which of the following are the symptoms of angle closure glaucoma?

A. Headache and nausea
B. Severe ocular pain
C. Haloes around lights
D. Diplopia
E. Tearing
F. Blurry vision

2. Which of the following is NOT a sign of angle-closure glaucoma?

A. Mixed injection
B. Constricted pupil
C. Fixed dilated pupil
D. Cloudy cornea
E. Shallow anterior chamber
F. High IOP

3. Which of the following are the typical signs of acute iridocyclitis?

A. Circumlimbal flush
B. Constricted pupil
C. Dilated fixed pupil
D. Keratic precipitates
E. Shallow anterior chamber
F. Posterior synechiae

4. **Which of the following are NOT useful in the treatment of acute iridocyclitis?**
   A. Myotic eye drops (Pilocarpine)
   B. Mydriatic eye drops (Atropine, Tropicamide)
   C. Topical steroids
   D. Systemic analgesics
   E. Topical broad-spectrum antibiotics
   F. Warm compresses

5. **What are NOT typical exam findings in acute conjunctivitis?**
   A. Eyelid erythema
   B. Red conjunctiva
   C. Subepithelial corneal infiltrates
   D. Anterior chamber cells
   E. Mucus in the canthus
   F. Posterior synechiae

6. **What are the typical complaints of patients with acute dacryoadenitis?**
   A. Pain and swelling in the outer part of the upper eyelid
   B. Pain and swelling around the lacrimal sac
   C. Ocular motility restriction
   D. Pain on moving the eye
   E. Excessive tearing
   F. Pus discharge from the lacrimal puncta

7. **What are the typical symptoms in a patient with orbital phlegmon?**
   A. Painful swelling of the eyelids
   B. Painful or difficult eye movements
   C. Headache and fever
   D. Decreased and double vision
   E. Exophthalmos
   F. Enophthalmos

8. **What are the typical signs of central retinal artery occlusion?**
   A. Sudden painless loss of vision
   B. Relative afferent pupillary defect
   C. Milky-white ischemic edema of the posterior pole
   D. Loss of the red reflex
   E. Opaque fundus with a red fovea (a cherry-red spot)
   F. Cotton wool spots

9. **What are the typical symptoms in a patient suspected of having a central retinal vein occlusion?**
   A. Hazy vision in one eye, floaters
   B. Eye pain and redness
   C. Dilated fixed pupil
   D. Double vision
   E. Corneal haze and decrease visual field
   F. Tearing

10. **Which symptoms would typically be present in a patient who has a corneal abrasion?**
    A. Pain
    B. Redness
    C. Tearing
    D. Photophobia
    E. Flashes
    F. Blurry vision

11. **What is the most appropriate next step in patient management after VA and slit lamp exam with fluorescein staining that reveals normal vision and multiple vertically oriented, nearly parallel, fine corneal abrasions?**
    A. Apply an eye patch
    B. Apply antibiotic ointment
    C. Instill cycloplegic drops
    D. Evert the upper eyelid
    E. Irrigate with normal saline solution
    F. Instill anesthetic drops

12. What are absolute signs of penetrating eyeball injuries?

A. Presence of a corneal or scleral wound
B. Hole in the iris
C. Prolapse of the intraocular contents
D. Hyphema
E. Intraocular foreign body
F. Leakage of the aqueous humor

13. What should be done first when initially evaluating a person with chemical substance exposure into the eye?

A. Get a good history to help determine what kind of substance got into the eye
B. Check the vision, pressure, and perform a slit lamp exam to determine the extent of damage done by the substance
C. Begin immediate ocular irrigation
D. Instill anesthetic drops so that the patient is more comfortable and able to undergo an ocular evaluation
E. Getting the patient some oral pain medications so that the patient is more comfortable and able to undergo an ocular evaluation
F. Apply binocular bandage and send to the ophthalmology department in supine position

C. Clinical Cases

Case 1

A 50-year-old woman complains of a left-sided headache and severe pain in the left eye associated with nausea and vomiting and haloes around light sources. On examination: Vis OD — 0.5, OS — 0.1; OD appears normal, OS — conjunctival injection, mid-dilated non-reactive pupil, corneal edema, shallow anterior chamber, on palpation the eye is stone hard. What are the diagnosis and initial treatment options?

Case 2

A 25-year-old man presented with right eye pain and redness with associated photophobia, excessive tearing, and reduced vision. On examination there were no significant findings in his left eye. In the right eye: Vis — 0.5; circumcorneal injection, keratic precipitates, flare and cells in the anterior chamber, miosis and posterior synechiae. The anterior chamber angle is open, IOP — 18 mm Hg. The fundus OU appeared normal. What is the diagnosis and initial treatment options?

Case 3

A 38-year-old woman presented with complaints of red irritated eyes, foreign body sensation, tearing, swelling of the eyelids, slight blurry vision that is more evident in the left eye than in the right one. Six weeks prior to any ocular symptoms, the patient had an upper respiratory infection, which had subsided spontaneously. Examination findings: eyelid edema, conjunctival injection, follicles on the palpebral conjunctiva, mild chemosis, crusting on the lashes OS > OD, palpable pre-auricular lymphadenopathy. What are the diagnosis and initial treatment options?

Case 4

A 53-year-old woman presented with complaints of painful red left eye, which started 2 days ago after she had a mild trauma caused by a tree branch. She also had complaints of yellowish-white mucus discharge, photophobia, tearing, and blurred vision in her left eye. Examination findings: OS — lid edema, the palpebral fissure is narrowed, conjunctival hyperemia and chemosis, mixed injection, yellow-grey 1.75 mm × 1.75 mm lesion located inferiorly nasally on the cornea with one edge undermined the corneal stroma, hypopyon. What are the diagnosis and initial treatment options?

Case 5

A 24-year-old man presented with complaints of swelling and tenderness of the upper eyelid, eye pain, redness, photosensitivity, tearing, and limited ocular movement in the left eye for two days associated with fever and upper respiratory infection. Examination findings: OD — normal, OS — erythema of the eyelid, edema of the lateral one-third of the upper eyelid, conjunctival injection, chemosis, mucopurulent discharge, eversion on the upper lid — the lacrimal gland is seen. What are the diagnosis and initial treatment options?

Case 6

A mother with a child of 2 months old referred with complaints of tearing, redness, swelling, and purulent discharge in the inner corner of the child's right eye that appeared over one month ago. Examination findings: OD — epiphora, edema, and redness in the region of the medial epicanthus, mucopurulent discharge from the puncta on pressing the region of the lacrimal sac. OS — without pathology. What are the diagnosis and initial treatment options?

Case 7

A 36-year-old woman presented with complaints of severe headache, fever, nasal discharge. She also notices pain in the right eye that worsens upon eye movement, swelling of the eyelids and bulging of the eye, double vision with two eyes open. From the medical history: the patient suffers from chronic sinusitis. Examination findings: Vis OU — 1.0; OD — edema and hyperemia of the eyelids, the palpebral fissure is narrowed, exophthalmos, eye motility restriction; the anterior structures and fundus are normal. What is the diagnosis and initial treatment options?

Case 8

An 81-year-old man presented with complaints of sudden painless loss of vision in the left eye. He had a history of coronary artery disease and recently underwent endovascular stenting of the left carotid with transient right hemiparesis (resolved). Examination findings: Vis OD — 0.5, OS — hand motions; RAPD OS; mild nuclear sclerosis cataracts OU; OD fundus normal; OS — pale retina with vessel attenuation, milky-white cloudy edematous retina with a red fovea (a cherry-red spot), the optic disc is pale. What are the diagnosis and initial treatment options?

Case 9

A 58-year-old woman complains of decreased vision in the left eye. She first noticed it when she closed the right eye while washing off her make-up a week ago. Examination findings: Vis OD — 0.8, OS — 0.1; trace RAPD OS, anterior structures are normal OU; the fundus of OS — edematous optic disc with blurred margins, scattered intraretinal hemorrhages in all 4 quadrants, cotton wool spots, macular oedema with loss of foveal light reflex, the vessels are tortuous. What are the diagnosis and initial treatment options?

Case 10

A 10-year-old boy has presented with a red eye shortly after a blunt trauma to his right eye by an elbow during football game. He reports tenderness in his right eye, eyebrow, and cheekbone, photosensitivity and blurred vision. Examination findings: Vis OD — 0.5, OS — 1.0; mild conjunctival injection and edema of the upper eyelid of OD; IOP OD — 23 mmHg, OS — 15 mm Hg; sluggish pupil reaction in OD, normal pupil reaction in OS; extraocular movements are full in OU; the cornea with no epithelial defects; the anterior chamber of OD — blood of 1.5 mm high, OS — deep and quiet, no sign of a ruptured globe or an intraorbital pathology. What are the diagnosis and initial treatment options?

Case 11

A 32-year-old man presented with right eye pain immediately after he was pounding a metal object with a metal chisel. This was followed by photophobia, tearing, difficulty in eye opening, and blurred vision. The accident happened one day before the referral. Examination findings: Vis OD — hand motions, OS — 1.0; OD — conjunctival injection, penetrating corneal wound, a dense cataract with disruption of the anterior lens capsule, no view of the fundus due to cataract. OS — normal. What are the diagnosis and initial treatment options?

Case 12

A 24-year-old man presented with an acute ocular burn caused by a splash of acetic acid into his right eye. He complained of severe pain, constant irritation, excessive tearing, blepharospasm, and decreased vision. What are diagnosis and your initial management options?

Case 13

A 20-year-old man presented with a 3-day-old penetrating injury to the right eye with a tree branch. He complained of severe eye redness, pain and irritation, sensitivity to light, tearing, and decreased vision in the affected eye. Examination findings: Vis OD — counting fingers at 2 m, OS — 1.0; OD — eyelid edema, mixed injection, chemosis, entry wound 3 mm from the limbus at 9:00, keratic precipitates, grade 4 cells and flare in the anterior chamber, 1 mm hypopyon, the fundus showed severe vitritis. OS — without pathology. What are the diagnosis and initial treatment options?

CHAPTER 18

Ocular Manifestations of Systemic Diseases

OBJECTIVES

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

— list systemic diseases that most commonly affect the eyes;
— know the ocular manifestation of systemic diseases;
— know the most common changes on the eye fundus in systemic diseases;
— know the basic diagnostic methods of ocular fundus examination;
— evaluate and manage the patients with ocular pathology in systemic diseases;
— know the main principles of eye treatment in systemic diseases.

Plan:

1. SYSTEMIC OPHTHALMOPATHY
2. SYMPTOMS OF RETINAL DISORDERS IN SYSTEMIC DISEASES
3. EXAMINATION METHODS
4. THE EYE AND SYSTEMIC DISEASES

4.1. Ocular Manifestation of Cardiovascular Diseases

- The Eye and Systemic Hypertension
  - Hypertensive Retinopathy
- The Eye and Kidney Diseases
  - Renal Retinopathy
- Toxemia of Pregnancy
- The Eye and Atherosclerosis

4.2. Ocular Manifestation of Hematological Diseases

- The Eye and Anemias
- The Eye and Leukemia

4.3. Ocular Manifestation of Endocrine Diseases

- The Eye and Diabetes
  - Diabetic Retinopathy
- The Eye and Thyroid Diseases

4.4. The Eye and Multiple Sclerosis

4.5. Ocular Manifestation of Infectious Diseases

- The Eye and Acquired Immune Deficiency Syndrome
  - HIV retinopathy
- The Eye and Cytomegalovirus
- The Eye and Herpes
- The Eye and Syphilis
- The Eye and Toxoplasmosis
- The Eye and Tuberculosis

4.6. Ocular Manifestation of Nutritional Deficiencies

- Vitamin A deficiency
- Vitamin B_1 deficiency
- Vitamin B_2 deficiency
- Vitamin C deficiency
- Vitamin D deficiency

1. Systemic Ophthalmopathy

Systemic diseases, which involve certain organs or the whole body, may affect the eye too. Occasionally ocular findings may be the first indication of an underlying systemic disease leading to diagnosis, especially vascular, as the eye is the only organ where a microcirculatory system can be directly visualized and investigated with such precision. The health of the blood vessels in the eye often indicates the condition of the blood vessels (arteries and veins) throughout the body. Patients with known systemic illnesses need to have their eyes specifically checked for severity of the disease indication and monitoring of ocular complications.

There are many systemic diseases known to cause ocular or visual changes. They are:

— cardiovascular diseases,
— hematological diseases,
— endocrine diseases,
— multiple sclerosis,
— infectious diseases,
— connective tissue disorders,
— autoimmune disorders,
— neoplastic diseases,
— nutritional deficiencies.

Signs of a systemic disease may be evident on the outer surface of the eye (eyelids, conjunctiva, cornea and sclera), in the middle of the eye (uvea), and at the back of the eye (retina).

2. Symptoms of Retinal Disorders in Systemic Diseases

— Blurred vision.
— Reduced visual acuity.
— Floaters.
— Flashing lights.
— Visual field defects.
— Loss of color vision.

3. Examination Methods

- External examination.
- Visual acuity.
- Slit-lamp exam.
- Ophthalmoscopy, direct and indirect.
- Color fundus photography.
- Perimetry.
- Campimetry.
- Light and dark adaptation.
- Color vision testing.
- Ultrasonography.
- Fluorescein angiography.
- Optical coherence tomography.
- Magnetic resonance imaging.
- Computer tomography.
- Methods of electrophysiological examination of the retina and the optic nerve.

4. The Eye and Systemic Diseases

The retina is the only portion of the central nervous system visible from the outside. Likewise, the fundus is the only location where blood vessels can be visualized. So, viewing the fundus is a great way to evaluate the patient's overall vasculature. The fundoscopic exam can discover a pathological process otherwise invisible; examples are plentiful and include recognizing endocarditis, CMV in an HIV-infected patient, and being able to stage both hypertension and diabetes.

4.1. Ocular Manifestation of Cardiovascular Diseases

The Eye and Systemic Hypertension

Systemic hypertension has a profound effect on the retinal, choroidal, and optic nerve circulation. A variety of retinal vascular changes can be seen in hypertensive patients; these depend on the severity and duration of hypertension. Severe hypertension causes diminishing of the blood flow through the retina and choroid, resulting in generative changes in the retina and loss of vision. Retinal vascular abnormalities are useful first indicators for cerebrovascular disease and stroke.

Hypertensive Retinopathy

Hypertensive retinopathy is the most common ocular presentation of systemic hypertension. Other ocular manifestations include central retinal vein occlusion, central retinal artery occlusion, ischemic choroidal infarcts, ischemic optic neuropathy with a poor prognosis.

Definition. Hypertensive retinopathy is the damage to the retina and retinal circulation due to high blood pressure.

Etiology. Elevated blood pressure, defined as systolic greater than 140 mm Hg and diastolic greater than 90 mm Hg, causes a variety of blood vessels and retina abnormalities with the, including thickening and narrowing of the arteries, which decreases the amount of blood that can flow through them. As a result, the blood supply to the retina is reduced leading to retinal damage.

As hypertensive retinopathy progresses, blockages of the retinal blood vessels and leaking from them may develop. These changes lead to a gradual loss of vision, particularly if they affect the macula. Even mild hypertension may damage the retinal blood vessels if it goes untreated for years. Sudden, severe high blood pressure may cause swelling of the optic nerve.

Clinical Picture. Hypertensive retinopathy can be classified into different stages according to characteristic retinal changes that include retinal arteriolar narrowing, arterio-venous nicking, retinal hemorrhages, cotton wool spots and microaneurysms as well as optic disc and macular edema, in severe cases (fig. 18.1). These signs develop due to acute and chronic elevations in blood pressure.

There are several classification systems. The first classification of hypertensive retinopathy, consisting of four grades, was proposed in 1939 by Keith, Wagener and Barker, based on the level of severity of retinal findings. Nowadays, a simplified three-grade classification is used, it was proposed by Wong and Mitchell (2004) based on the strength of the reported associations between hypertensive retinopathy signs and cardiovascular risks (table 18.1). This classification allows clinicians to evaluate the risk of cardiovascular morbidity and mortality (stroke, heart failure, renal failure, etc.) according to the funduscopic picture of patients with systemic hypertension and suggest an appropriate management plan and preventive strategies for them.

Table 18.1

Classification of Hypertensive Retinopathy by Wong and Mitchell

Grade	Stage	Ophthalmoscopic Signs	Systemic Association
I	Mild retinopathy (fig. 18.2)	One or more of the following fundoscopic signs: — generalized arteriolar narrowing[1]; — focal arteriolar narrowing; — arteriovenous nicking[2]; — arteriolar wall opacity ("copper" or "silver" wiring[3])	Weak* associations with a risk of stroke, coronary heart disease and cardiovascular mortality
II.	Moderate retinopathy (fig. 18.3)	One or more of the following fundoscopic signs: — hemorrhages (blot-, dot- or flame-shaped); — microaneurysms; — cotton wool spots[4]; — hard exudates	Strong** association with a risk of stroke, congestive heart failure, renal dysfunction, and cardiovascular mortality
III	Malignant retinopathy (fig. 18.4)	Signs of moderate retinopathy with optic disc swelling	Associated with mortality and renal failure

Note: [1] — arteriolar constriction with a decrease in the ratio between the width of the retinal arterioles and the retinal venules (the normal ratio is 2:3);
[2] — arteriovenous nicking — the thickening artery compresses the veins so that the veins appear nipped at the arteriovenous crossing; this is also called A—V nipping (Gunn's sign);
[3] — "copper" wire reflex — the transparent arterial wall becomes thick and reflects light, the reflex looks wider and burnish copper-colored; "silver" wire reflex — marked thickening of the arterial walls causes all the light to reflect and the artery looks brilliant white;
[4] — cotton wool spots — oxygen-deprived areas due to retinal artery microinfarcts;
* — risk and odds ratio greater than 1 but less than 2;
** — risk and odds ratio of 2 or greater.

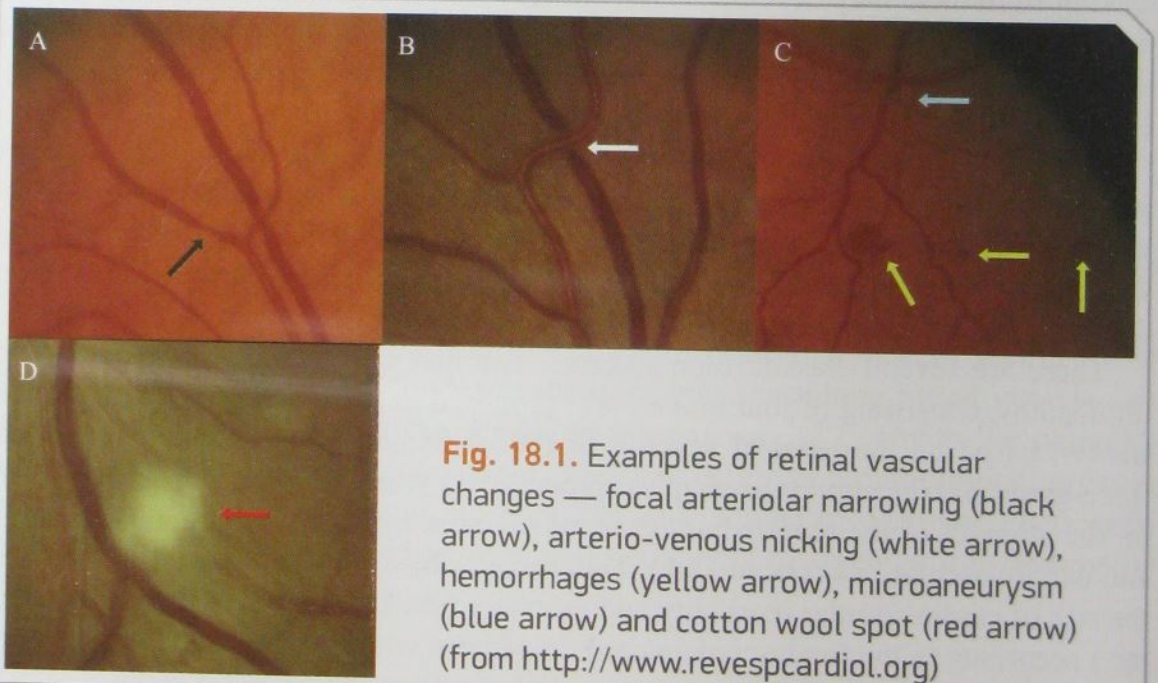


Fig. 18.1. Examples of retinal vascular changes — focal arteriolar narrowing (black arrow), arterio-venous nicking (white arrow), hemorrhages (yellow arrow), microaneurysm (blue arrow) and cotton wool spot (red arrow) (from http://www.revespcardiol.org)

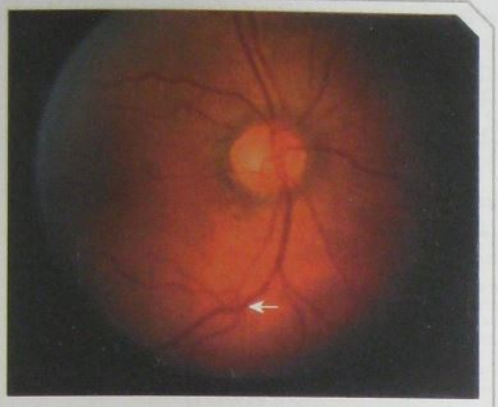


Fig. 18.2. Mild hypertensive retinopathy with the presence of generalized retinal arteriolar narrowing and arteriovenous nicking (white arrow) (Grosso A., Veglio F., Porta M., Grignolo F. M., Wong T. Y. Hypertensive Retinopathy Revisited: Some Answers, More Questions // Br J Ophthalmol. — 2005. — Vol. 89 (12). — P. 1646—1654).

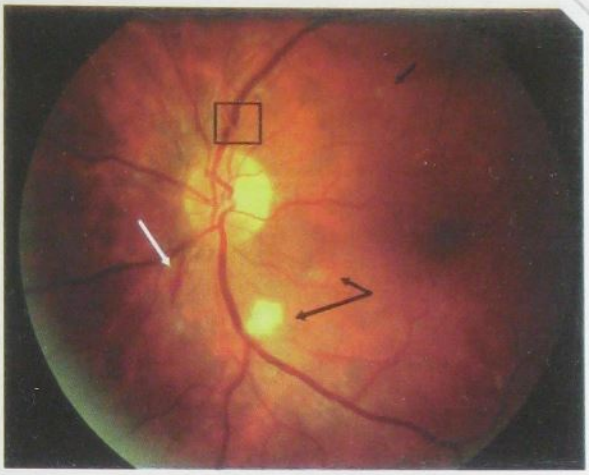


Fig. 18.3. Moderate hypertensive retinopathy with the presence of arteriovenous nicking (box), cotton wool spots (black arrows), and retinal hemorrhage (white arrow) (from http://www.eyerounds.org)

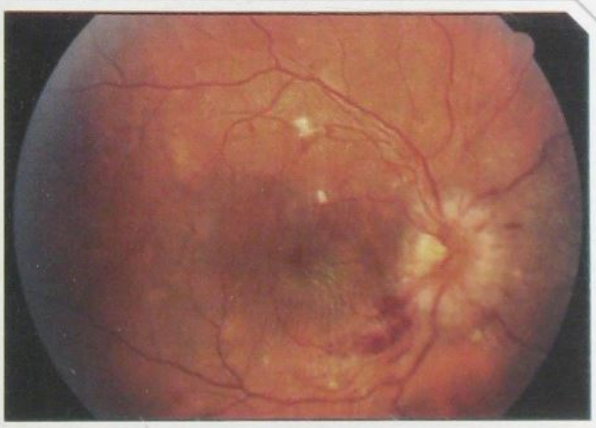


Fig. 18.4. Malignant hypertensive retinopathy with the presence of optic disc swelling, multiple cotton wool spots, retinal hemorrhages and macular exudates (star-shaped) and (from http://clinicalgate.com/hypertension-5/)

Malignant hypertensive retinal changes are likely findings in patients with hypertensive crisis, which is an abrupt elevation in blood pressure with a systolic blood pressure of greater than 210 mm Hg and a diastolic blood pressure of more than 120 mm Hg. The hallmark of malignant hypertension is optic disc swelling, which carries a poor prognosis if left untreated. The patient with malignant hypertension is at risk of developing heart and renal failure and hypertensive encephalopathy. Blood pressure must be controlled immediately, but in a controlled, careful fashion, since a sudden drop in tissue perfusion can result in infarction of the optic disc and subsequent blindness.

Complaints. Patients with hypertensive retinopathy usually do not experience any symptoms, which is why most cases are diagnosed after a routine eye exam. Some patients may experience blurred vision. As the disease progresses and in acute cases

they complain of reduced vision, red eye (subconjunctival hemorrhages), eye pain, headaches, visual field defects, diplopia.

Signs of hypertensive retinopathy according to the stage include:

— arteriolar constriction (generalized or focal);
— tortuosity of the retinal blood vessels;
— changes of the arteriovenous crossings (arteriovenous nicking);
— arteriolar wall opacity ("copper" or "silver" wiring);
— retinal hemorrhages (blot, dot or flame shaped);
— microaneurysms;
— cotton-wool spots;
— hard exudates (lipids, or fats, that have leaked from the blood vessels): this usually occurs in a star configuration around the macula, which can indicate swelling of the macula and cause vision loss;
— retinal edema, which can lead to retinal detachment and vision loss;
— optic disk edema.

Methods of Examination. Medical history analysis, systemic blood pressure measurement, slit-lamp examination, ophthalmoscopy, fundoscopy, OCT of the retina, fluorescein angiography.

Differential Diagnosis. Diabetic retinopathy, retinal venous obstruction, hyperviscosity syndromes, congenital hereditary retinal arterial tortuosity, radiation retinopathy, ocular ischemic syndrome.

Treatment for hypertensive retinopathy is directed at the lowering and control of blood pressure. With this aim anti-hypertensive drugs are required — angiotensin-converting enzyme inhibitors (ACE inhibitors), calcium channel blockers, diuretics, and β-adrenergic blockers. Other less commonly used medications include α-adrenergic blockers, direct vasodilators, and central α_2-adrenergic agonists.

When vision loss occurs, treatment of retinal edema with laser or intravitreal injection of corticosteroids or antivascular endothelial growth factor drugs (Ranibizumab, Bevacizumab) may be useful.

Treatment of hypertension of first stages results in improvement of ocular changes. However, in severe cases eye damages are irreversible, which causes permanent vision problems.

Prognosis is generally good for individuals with mild retinopathy; vision can easily be preserved with control of systemic blood pressure. Patients with moderate and malignant retinopathy are at a significantly increased risk of vision loss depending on the extent of damage to either the macula or the optic nerve. Patients with uncontrolled hypertension and malignant retinopathy have a generally poor prognosis for survival due to stroke, heart failure, kidney failure, or heart attack.

Complications. Optic neuropathy, central vein or artery occlusions, retinal detachment, vitreous hemorrhages, ischemic optic neuropathy, optic nerve atrophy. All the above complications can lead to vision loss.

Prophylaxis. Routine blood pressure monitoring and treatment, maintaining healthy lifestyle will prevent hypertensive retinopathy from developing.

The Eye and Kidney Diseases

Kidney diseases have a major influence on the body's vascular system and the eye's as well, causing hypertension and specific pathology of the retina. It is why in patients with kidney diseases the fundus examination can help to establish a diagnosis, reveal subclinical complications and severity of the underlying condition, and even have a prognostic significance.

Renal Retinopathy

Definition. Renal retinopathy is a damage to the retinal vasculature associated with diseases of the kidneys.

Etiology. Renal retinopathy develops in cases of chronic diffuse glomerulonephritis associated with albuminuria and systemic hypertension, and rarely in acute nephritis.

Clinical Picture. The ophthalmologic picture of retinal retinopathy differs according to the duration and severity of the underlying disease. After short duration of chronic diffuse glomerulonephritis the retinal blood vessels are involved in the pathological process and the ophthalmoscopic picture is characterized by narrowing of the retinal arteries, irregularity and tortuosity of the retinal vessels. With long-term course of renal disease the retinal vessels become sclerotic and the retina itself becomes involved: marked diffuse retinal edema, swelling of the optic disc, scattering of numerous flame-shaped hemorrhages over the fundus, appearance of cotton-wool spots and hard exudates, which present star-shaped in the macula, which is typical for this pathology. The clinical picture is almost the same as in hypertensive retinopathy, but more severe and has a bad prognosis for the life of the patient.

Complaints. Patients often complain of blurred vision, reduced visual acuity, distortion of objects, floaters.

Signs. Externally, the eyes most commonly remain calm, sometimes conjunctival edema, conjunctival vascular ischemic angiopathy, corneal and conjunctival calcifications may be noticed. The major changes are found on the fundus; they are:

— constriction of the retinal arterioles;
— irregularity and tortuosity of the retinal blood vessels;
— vascular sclerosis;
— generalized retinal edema;
— edema of the optic disc with indistinct margins;
— cotton-wool spots;
— retinal hemorrhages;
— hard star-shaped exudates on the macula.

Methods of Examination. Medical history analysis, systemic blood pressure measurement, blood and urine tests, slit-lamp examination, ophthalmoscopy, fundoscopy, OCT of the retina, fluorescein angiography.

Differential Diagnosis. Hypertensive retinopathy, diabetic retinopathy.

Treatment is mainly directed at the underlying disease. Topically medicaments that strengthen the vascular walls, improve trophic processes, resolve hemorrhages, and antioxidants (ATP, vitamins A, C, B complex) are applied.

Prognosis. Visual functions can be restored in some way under successful treatment of the underlying disease and topical treatment of ocular disorders but appearance of ophthalmoscopic findings in chronic diffuse glomerulonephritis is a bad prognostic sign for the patient's life.

Complications. Partial or complete detachment of the retina that leads to scotomas or loss of vision. Retinal detachment is a formidable diagnostic sign, as it usually appears a few weeks before the patient's death.

Prophylaxis. Regular eye examination for monitoring of the progression of the underlying kidney disease and its adequate treatment.

Toxemia of Pregnancy

Toxemia of pregnancy, also known as pregnancy-induced hypertension, or pre-eclampsia is a multisystem disorder that is characterized by sudden onset of hypertension in combination with generalized edema and/or proteinuria. It affects about 5—7 % of all pregnancies and occurs most often in young women with a first pregnancy or women older than 35, and in those patients who have diabetes or who are carrying more than one baby (twins, triplets, etc.). The onset of this disorder is usually in the third trimester of pregnancy.

Ocular involvement is common in the majority of cases of pregnancy-induced hypertension. Most often patients complain of blurring of vision, however, other symptoms may be noticed such as photopsia, scotomas, diplopia. Ophthalmoscopic picture reveals retinal changes similar to those from hypertensive retinopathy according to the severity of the condition — narrowing of the nasal arterioles followed by generalized narrowing; spasm of the retinal arterioles, cotton-wool spots, hemorrhages, retinal edema, exudates that may be associated with a macular star; rarely it may be complicated by exudative retinal detachment, optic disc edema, acute ischemic optic neuropathy, optic atrophy.

Toxemia of pregnancy can lead to serious complications not only for vision but even for life of the mother and the baby if not treated quickly. However, it cannot be completely cured until delivery. It only can be controlled by special diets, anti-hypertensive medications, and limitation of activity. In some severe cases of not responding to treatment, early delivery of the baby is needed to ensure the survival of the mother. The issue of pre-term birth is decided individually on the basis of general condition, the term of pregnancy, the nature and dynamics of changes in the fundus. Retinal changes usually regress with a decrease in blood pressure and may disappear completely after the baby is born.

The Eye and Atherosclerosis

Atherosclerotic retinopathy, or **retinal atherosclerosis** is a damage of the retinal vasculature caused by atherosclerosis (sometimes called "hardening" or "clogging" of

the arteries). In this condition, the inner walls of the arteries in the retina as well as in the whole body become less flexible and thicken as a result of inflammatory deposits (called plaque) of fatty cells, cholesterol, calcium, and other substances found in the blood. Over time, the plaque hardens, which causes narrowing of the arteries and restriction or blockage of blood supply to the retina resulting in its damage. Atherosclerosis can affect the arteries in the heart, legs, brain, kidneys, and other organs, and also lead to serious problems, including heart attack or stroke.

Atherosclerotic retinopathy is mostly asymptomatic at early stages until plaque severely narrows or totally blocks an artery causing blurring of vision, visual field scotomas, and can lead to permanent vision loss. Sometimes, one of the first signs of atherosclerosis will be sudden visual loss in one eye lasting just for seconds or minutes. Funduscopic picture reveals thickening of the arteriolar walls, changes in the arteriolar reflex ("copper" or "silver" wiring sign), retinal hemorrhages, cotton-wool spots, development of optic disc edema, and subatrophy.

Ocular symptoms and signs may be the first indications of systemic atherosclerosis. They warn of the risk of cardiac and cerebral complications of thrombosis and thromboembolism.

Atherosclerosis cannot be stopped and current treatment methods cannot reverse it, but there are medicines and other treatments that can slow down its progress to reduce the risk of vision loss, morbidity, and mortality. They are drugs to lower cholesterol, nutrition and dietary supplements, healthy lifestyle, medicines for improving cerebral and retinal circulation, and optic nerve protection.

4.2. Ocular Manifestation of Hematological Diseases

The Eye and Anemias

Anemic retinopathy is retinal damage caused by anemias — a group of diseases characterized by severe lowering of the level of healthy red blood cells or hemoglobin. Anemia can develop as a result of nutritional problems such as iron deficiency, vitamin deficiency or folate deficiency. Other causes of anemia include blood loss, inadequate production of red blood cells (aplasic anemia) or increased destruction of red blood cells (hemolytic anemia). Anemic retinopathy may also be secondary due to other systemic diseases such as cancer, infection, autoimmune disorders or pregnancy. Retinopathy in patients with anemia develops when the hemoglobin (Hb) level falls by 50 % and is consistently present when Hb is lower than 35 % (6 g/dL).

Most cases of anemic retinopathy are asymptomatic; rarely, blurring of vision may be the only complaint, but due to irreversible damage of the macula or optic nerve head loss of vision may occur. Common ocular findings include conjunctival pallor

and hemorrhages. Retinal changes include hemorrhages that are present at all levels of the retina and choroid, Roth's spot hemorrhages (white centered hemorrhages), exudates, cotton wool spots, retinal edema. In severe cases of anemia, the general fundus and optic nerve disc are pale, the veins are dilated and tortuous, hemorrhages are multiple, edema of the optic nerve disc and adjacent retina is present.

In most cases, only treatment of the underlying etiology is needed, and retinopathy generally resolves on its own.

The Eye and Leukemia

Leukemic retinopathy is a damage to the retina associated with leukemia, which is defined as a neoplastic blood disorder characterized by overproduction of abnormal white blood cells. Leukemia is classified according to the type of white blood cell that is affected — myeloid or lymphoid, and according to the onset — acute or chronic. Ocular findings are more common in the acute form, but both acute and chronic leukemia can have ocular signs and has been reported to occur in up to 90 % of all leukemia patients. Ocular involvement can be due to direct infiltration of leukemic cells or secondary to associated blood disorders such as hyperviscosity, anemia or thrombocytopenia.

The most common subjective symptom of leukemic retinopathy is blurring of vision. Ophthalmoscopic signs include pale and yellow-orange fundus color, scattered retinal hemorrhages (dot-shaped, flame-shaped, intraretinal, subretinal, or subhyaloid), cotton wool spots, Roth's spots, retinal vein tortuosity, peripheral retinal microaneurysms and neovascularization, neovascularization and edema of the optic disc with blurring of its margin (fig. 18.5).

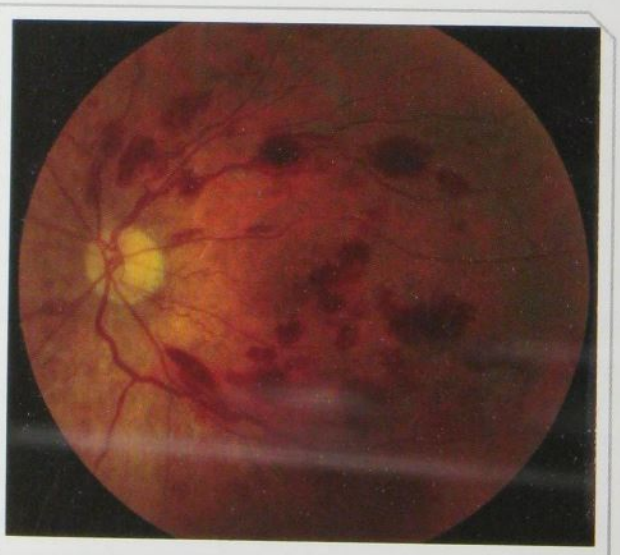


Fig. 18.5. Leukemic retinopathy with the presence of retinal vein tortuosity, central and paracentral superficial hemorrhages, some with a white center (Roth spots) (from http://www.eyerounds.org)

Leukemic infiltration can also lead to other ocular presentations in the anterior segment and the orbit as subconjunctival hemorrhages, chemosis, spontaneous hyphema, thickening of the iris, change in its color, sterile corneal ulcer, glaucoma, exophthalmos.

Leukemic retinopathy usually requires systemic treatment with the use of chemotherapy, immunotherapy, and radiotherapy, but the presence of this condition is mostly a poor prognostic indicator.

4.3. Ocular Manifestation of Endocrine Diseases

The Eye and Diabetes

Diabetes mellitus is a chronic metabolic disorder characterized by an increase in glycemia (levels of sugar in the blood) caused by insufficient production of insulin by the pancreas or inadequate sensitivity of cells to insulin, or both. Over time, high blood glucose level damages nerves and blood vessels, leading to microvascular and macrovascular complications such as heart disease, stroke, kidney disease, blindness, dental disease, and amputations. The two main types of diabetes are type 1 (insulin-dependent) diabetes and type 2 (non-insulin-dependent) diabetes. Gestational diabetes is a third form of diabetes that develops only during pregnancy. Other types of diabetes are caused by diseases and injuries of the pancreas, certain chemical toxins and medications, infections, and other conditions.

Diabetes may affect any structure of the eye, and 30 % of patients have ocular problems when diabetes is present. The eyelids can be affected by styes and lid cellulitis due to the increased susceptibility to infection. Senile cataract occurs earlier in persons who have diabetes and progress more rapidly than in most elderly people. Microvascular disease can affect the vascular supply to the extraocular muscles, which can lead to a muscle palsy that usually manifests as diplopia. The sixth and third nerve palsies are the most commonly affected. Rarely the iris may be affected by diabetes, with new blood vessel formation on it, and, if these block the drainage of aqueous fluid, glaucoma can be caused. Thus, chronic open-angle glaucoma is more common in diabetic than in non-diabetic patients.

Diabetic Retinopathy

Diabetic retinopathy is the most common form of eye problem affecting people with diabetes and a leading cause of visual loss worldwide, and is the principal cause of impaired vision in patients between 25 and 74 years of age.

Definition. Diabetic retinopathy (DR) is a chronic progressive, potentially sight-threatening disease of the retinal blood vessels associated with diabetes.

Etiology. Diabetic retinopathy is caused by prolonged high blood sugar levels. Over time, high sugar levels damage the blood vessels of the retina, which reduces the retina's blood supply leading to poor delivery of oxygen. The end result is hypoxic damage to the retina, which may swell or grow new compensatory blood vessels (neovascularization) that bleed easily, create scar tissue, pull on the retina leading to retinal detachment that can cause loss of vision.

Clinical Picture. There are two main clinical stages of diabetic retinopathy: they are non-proliferative or proliferative, named for the absence or presence of abnormal new blood vessels growing in the retina (fig. 18.6).

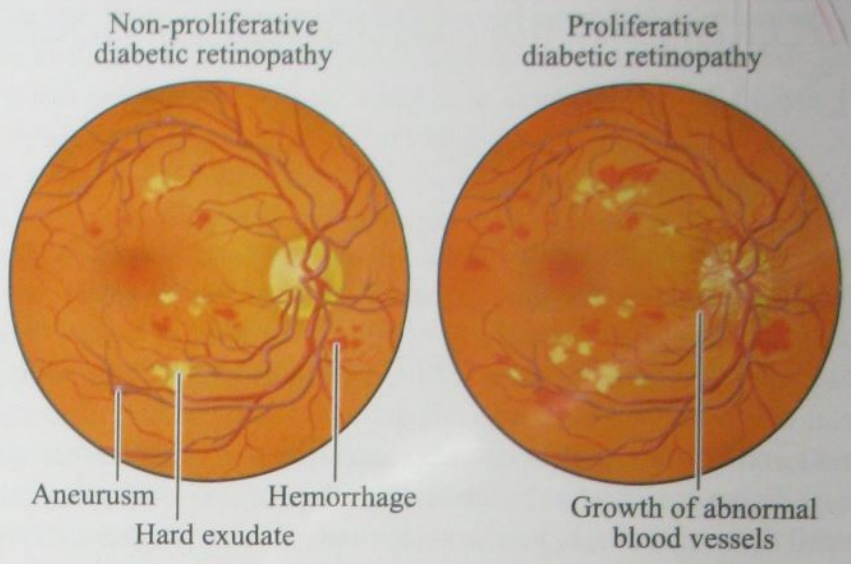


Fig. 18.6. Schematic picture of non-proliferative and proliferative diabetic retinopathy (http://kirkwoodeyecenter.com/services/diabetic-eye-care/)

Nonproliferative diabetic retinopathy (NPDR), also called *background retinopathy,* is the early stage of the disease that is characterized by weakening of the small retinal blood vessels and increasing of capillary permeability.

The severity of NPDR is graded as mild, moderate, severe, or very severe. In *mild disease*, the affected vessels develop tiny bulges called microaneurysms that may burst and leak blood and fluid into the retina, which leads to appearance of dot and blot hemorrhages and hard exudates (accumulations of lipids leaked from blood vessels) (fig. 18.7). These changes usually do not produce noticeable symptoms. However, if deposition of fluid involves the macula region, it causes macular edema that may eventually lead to vision loss (fig. 18.8).

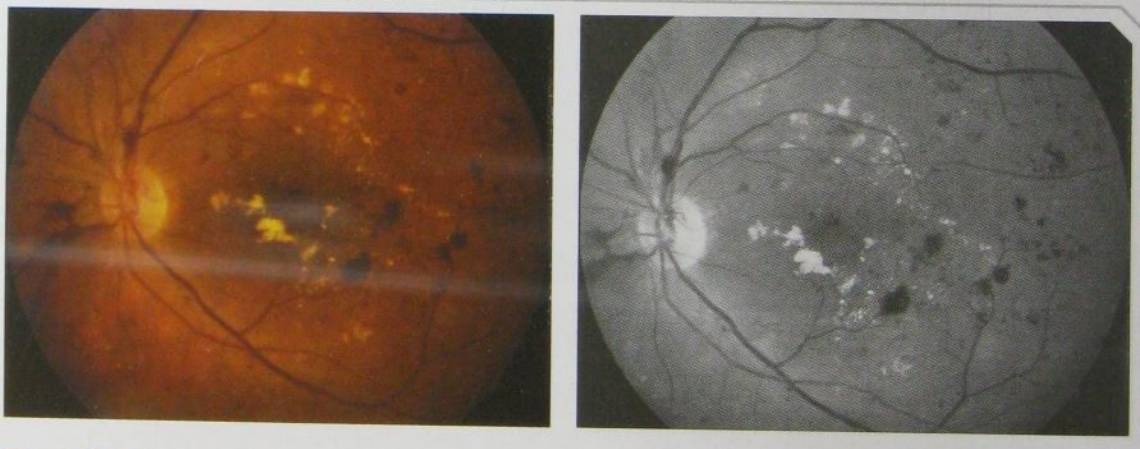
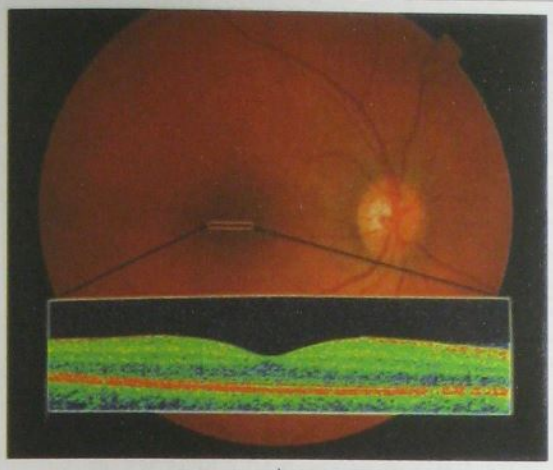
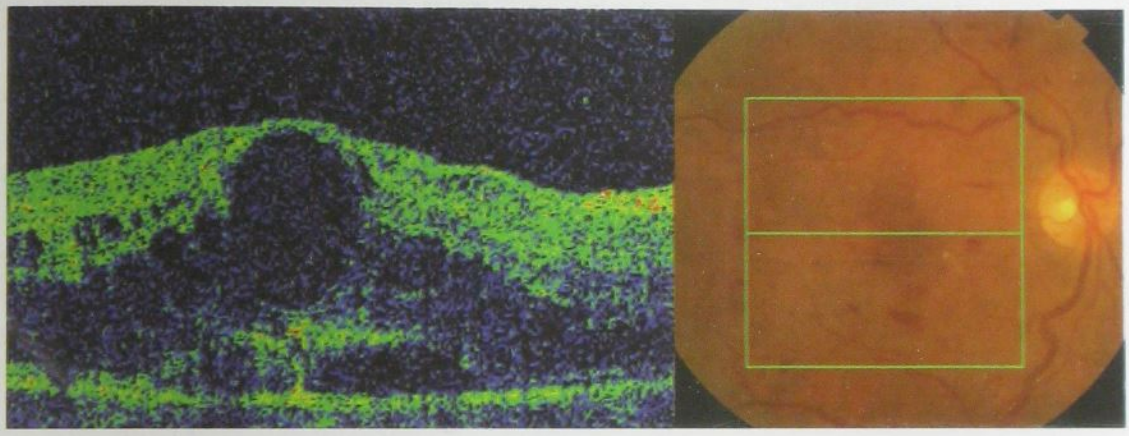


Fig. 18.7. Nonproliferative diabetic retinopathy with the presence of multiple microaneurysms, retinal hemorrhages and hard exudates (ophthalmoscopy and fluorescence angiography)



A



B

Fig. 18.8. OCT image of a healthy macula (A) (from http://northshore-eye.com/eye-health-services) and (B) diabetic macular edema (from http://www.goodhopeeyeclinic.org.uk)

In *moderate NPDR*, ophthalmoscopic findings are associated with cotton-wool spots resulted from obstruction of the affected vessels that cause infarction of the nerve fibre layer (retinal ischemia), abnormally dilated and tortuous retinal veins, and intraretinal microvascular abnormalities (IRMA) that represent areas of formation of new intraretinal vessels, which are flat and do not protrude into the vitreous cavity. *Severe NPDR* is diagnosed according the "4-2-1 rule" — hemorrhages and microaneurysms are present in 4 quadrants; or venous dilating is present in 2 or more quadrants, or prominent IRMA are present in 1 or more quadrants. *In very severe stage* of NPDR, two of these features are present.

Proliferative diabetic retinopathy (PDR) is a late stage of the disease characterized by neovascularization. The lack of oxygen in the retina causes new blood vessels to grow along the retina and may extend into the vitreous. Newly formed vessels are abnormal and extremely fragile. If an eye with proliferative diabetic retinopathy is not treated, these fragile new blood vessels may bleed into the vitreous causing a vitreous hemorrhage resulting in blurred vision and floaters (fig. 18.9). Fibrous tissue which accompanies the new blood vessels may contract and cause a tractional retinal detachment leading to blind spots, flashing lights, and sudden loss of vision. The new blood

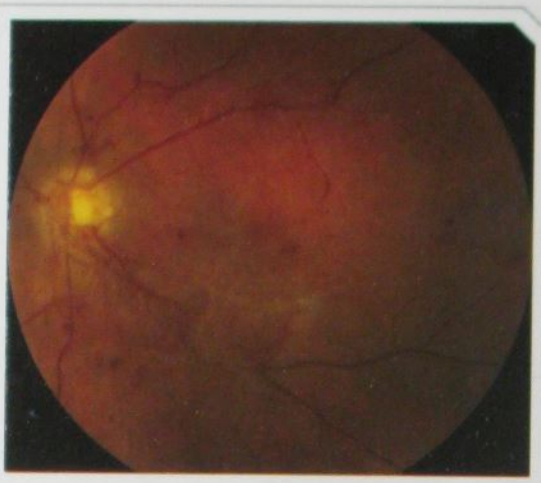


Fig. 18.9. Proliferative diabetic retinopathy with extensive neovascularizations (from http://www.eyerounds.org)

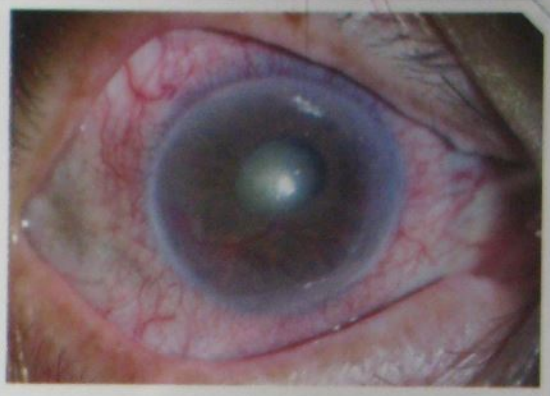


Fig. 18.10. Neovascular glaucoma with iris rubeosis and cataract in diabetes

vessels may also grow into the angle of the anterior chamber of the eye and cause neovascular glaucoma (fig. 18.10). If left untreated, the patient eventually becomes completely blind.

Complaints. Diabetic retinopathy starts silently and usually has no symptoms in the initial stages until the disease reaches advanced stages — after several years in most cases. Patients notice:

- blurred or distorted vision;
- distortion;
- spots or floaters in the field of vision;
- flashing lights;
- difficulty reading or seeing detailed work;
- difficulty distinguishing colors;
- sudden loss of vision in one or both eyes.

Signs. Funduscopic signs of different stages of DR are the following:

- *mild nonproliferative retinopathy:*
  - few scattered microaneurysms;
- *moderate nonproliferative retinopathy:*
  - multiple microaneurysms;
  - intraretinal hemorrhages (dot and blot);
  - retinal edema with hard exudates;
  - cotton-wool spots;
- *severe nonproliferative retinopathy:*
  - the above lesions, usually with exacerbation, plus:
  - venous beading and loops;
  - intraretinal microvascular anomalies;
- *proliferative diabetic retinopathy:*
  - neovascularization of the retina, optic disc or iris;

- vitreous hemorrhages;
- preretinal hemorrhages;
- tractional retinal detachment.

Methods of Examination. Visual acuity test, tonometry, ophthalmoscopy, funduscopy, color fundus photography, fluorescein angiography, OCT.

Differential Diagnosis. Branch retinal vein occlusion, central retinal vein occlusion, hypertensive retinopathy, ocular ischemic syndrome.

Treatment usually does not cure diabetic retinopathy, nor does it usually restore normal vision, but it may slow the progression of vision loss. The first step is to maintain blood glucose and blood pressure. Other treatment options depend on the stage and severity of the disease and may include:

— *Laser (or focal) photocoagulation* — application of laser burns to leaking blood vessels in a small area of the retina, usually near the macula, in order to seal them off, prevent from further leakage and reduce macular edema that can lead to vision loss.

— *Laser panretinal (or scatter) photocoagulation* — application of multiple scattered laser burns to the new abnormal blood vessels across the peripheral retina in order to destroy them and stop from growing, preventing further complications that may lead to severe visual impairment (fig. 18.11, 18.12).

— *Intravitreal steroid injections* — injections or implantation of long-acting steroids (Kenalog, Triamcinolone) into the vitreous to reduce the amount of fluid leaking and to decrease macular edema caused by diabetic maculopathy.

— *Intravitreal anti-VEGF drugs injection* — injections of anti-VEGF drugs (Bevacizumab, Ranibizumab, Aflibercept) into the vitreous to

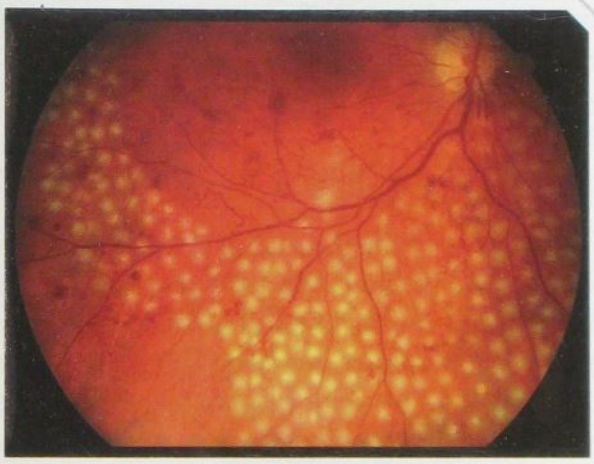


Fig. 18.11. Laser panretinal photocoagulation (from http://imagebank.asrs.org)

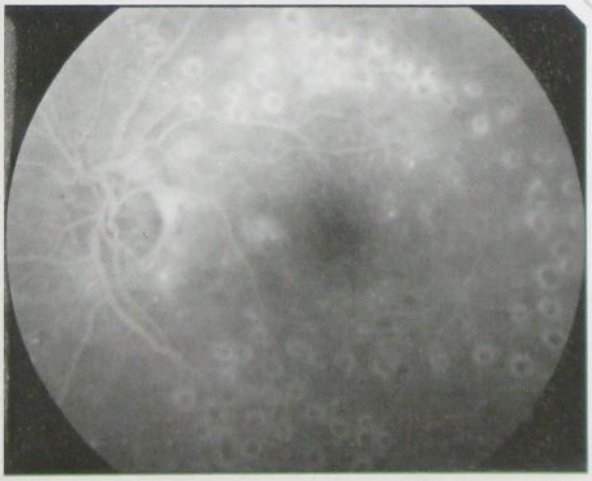


Fig. 18.12. Laser panretinal photocoagulation at fluoresceine angiography

block a particular protein called vascular endothelial growth factor (VEGF) that stimulates abnormal blood vessels to grow. These drugs, blocking VEGF, reverse abnormal blood vessel growth and decrease fluid in the retina.

— *Vitrectomy* — surgical removal of the blood-filled vitreous and replacement with a clear saline solution.

Prognosis depends on the type and severity of retinopathy. Patients with the initial stage of the disease do not develop vision loss; in patients with proliferative diabetic retinopathy the visual prognosis is worse. If left untreated, 50 % of patients with proliferative DR will lose their vision within two years and 90 % — in 10 years.

Complications. Untreated diabetic retinopathy can cause severe eye complications including vitreous hemorrhage, retinal detachment, neovascular glaucoma, blindness.

Prophylaxis. Diabetic retinopathy cannot be prevented however, regular eye exams, optimum control of blood sugar and blood pressure, early timely detection and treatment can help prevent severe vision loss but cannot be completely eliminated.

The Eye and Thyroid Diseases

Diseases of the thyroid gland can affect different parts of the eye and surrounding tissues, which is known as thyroid-associated ophthalmopathy. Although the condition is seen in all types of thyroid disorders, it occurs more often in patients with hyperthyroidism. According to the classification of A. F. Brovkin et al. (1985), three forms of thyroid-associated ocular pathology are distinguished — endocrine exophthalmos, edematous exophthalmos, and endocrine myopathy.

Thyroid disease can cause multiple eye problems. One of the most common is inflammation and swelling of the orbital tissues and extraocular muscles, which eventually cause bulging of the eyes and retraction of the eyelids. As the eyelids cannot be closed properly due to proptosis and failure of the lids, exposure of the conjunctiva and cornea may lead to excessive dryness of the eye and corneal ulcers. Compression of the orbital veins causes the conjunctival vessels to engorge, which results in conjunctival edema; in severe cases the conjunctiva may hang over the lower lid.

Swelling of the extraocular muscles can cause their weakness and impair ocular movement with consequent diplopia. In the early stages the movements most affected are elevation and abduction, but as the condition progresses all movements may become restricted.

Compression of the optic nerve by swollen tissues around the eye can cause optic atrophy, and compression of the central retinal veins and other veins — dilatation of the retinal veins with retinal edema and papilledema that may progress to optic atrophy and irreversible visual loss.

Patients with thyroid-associated ophthalmopathy usually complain of protrusion of the eye, swollen eyelids, eye irritation, red painful eye, tearing, light sensitivity, pain on eye movements, double vision, decrease of vision.

According to the severity of the disease ocular signs can include exophthalmos, lid retraction, lid lag, lagophthalmos, conjunctival injection, and/or chemosis, orbital fat prolapse, keratopathy, restrictive myopathy, optic neuropathy.

Endocrine exophthalmos, or **thyroid eye disease**, or **Graves' ophthalmopathy** is immune-mediated orbitopathy associated with thyroid gland disorders. This condition is not a disease, it is a sign or symptom of systemic disorder.

Thyroid eye disease is characterized by inflammation and swelling of the orbital tissues and the extraocular muscles that result in exophthalmos or proptosis (protrusion of the eyes), eyelid retraction, and restriction of eye movement. This causes a fixed stare appearance, inability to close the eyes, redness of the eyes, foreign body sensation, tearing, photophobia, dry eyes, pain in or behind the eye and double or decreased vision. Endocrine exophthalmos requires systemic and topical treatment.

*Read more about the etiology, clinical picture, and methods of treatment for **endocrine exophthalmos** in the Chapter **Diseases of the Orbit (p. 206)**.*

4.4. The Eye and Multiple Sclerosis

Multiple sclerosis (MS) is an autoimmune degenerative disease of the central nervous system (CNS) that is characterized by progressive inflammation and demyelination of the axons. In multiple sclerosis the myelin sheath, which is a protective membrane that wraps around the axon of a nerve cell, is damaged with inflammation and scarring that results in slowing or loss of neurologic signal transmission.

MS presents with a multitude of signs and symptoms depending on the location and severity of the lesion including physical, mental, and sometimes psychiatric problems. Visual disturbances are usually among the first symptoms of multiple sclerosis, among them optic neuritis, diplopia (double vision), and nystagmus are the most common conditions.

Optic neuritis is often one of the earliest symptoms of MS. It usually occurs because of inflammation or damage of the optic nerve. This can cause blurred vision, loss of color vision, blind spots in the center of the visual field, complete or partial blindness, and pain behind the eyes particularly during eye movement. It normally affects only one eye at a time.

Optic neuritis usually clears by itself within four to twelve weeks and people generally make a good recovery. However, in some cases there may be a permanent reduction in vision as the continued episodes cause scarring on the optic nerve. Medical treatment of optic neuritis may shorten recovery time but has no impact on the end result.

*Read more about the etiology, clinical picture, and methods of treatment for **optic neuritis** caused by multiple sclerosis in the Chapter **Diseases of the Optic Nerve (p. 437).***

4.5. Ocular Manifestation of Infectious Diseases

The Eye and Acquired Immune Deficiency Syndrome

Acquired immune deficiency syndrome (AIDS) is a serious (often fatal) infectious disease that suppresses the normal function of the immune system caused by the human immunodeficiency virus (HIV). The virus destroys specific cells of the immune system that are responsible for the proper response to infections (T cells) and the body's ability to fight infections. As a result people with AIDS become particularly vulnerable to opportunistic infections from the bacteria that other people normally fight off.

The virus is transmitted from one person to another through sexual contact, blood transfusions or contaminated needle sharing. An infected pregnant woman can pass HIV to her fetus through their shared blood circulation, delivering the baby during childbirth, and through breastfeeding. Once a person is infected by the virus, it stays inside the body for the whole life.

Characteristically a person infected with HIV initially experiences no symptoms for a variable period of time. This may be followed by the development of persistent generalized swelling of the lymph nodes (AIDS-related lymphadenopathy). Eventually most patients infected with HIV experience a syndrome of symptoms that includes excessive fatigue, weight loss, and/or skin rashes. The later stages of HIV infection are characterized by a progressive depression of T cells and repeated infections that can even occur during a course of antibiotic therapy for another infection (superinfections).

Ocular complications affect about 75 % of patients with AIDS and can involve almost any structure of the eye. Adnexal manifestations may include blepharitis, herpes zoster ophthalmicus, molluscum contagiosum, Kaposi sarcoma, squamous cell carcinoma or conjunctival microvasculopathy, among others. Anterior segment findings include dry eyes, infectious and fungus keratitis, microsporidiosis and iridocyclitis, and other complications. Posterior segment changes include an HIV-associated retinopathy/vasculopathy and a number of opportunistic infections of the retina or choroid, and neuro-ophthalmologic abnormalities.

HIV retinopathy

Definition. HIV retinopathy, also called AIDS-associated retinopathy is a non-infectious microvascular disorder of the retina seen in patients with AIDS. It is one of the most common retinal manifestations of HIV disease and is found in up to 70 % of AIDS patients.

Etiology. AIDS-associated retinopathy is caused by HIV-induced increase of plasma viscosity, HIV-related immune complex-mediated damage to endothelial cells and direct infection of the retinal vascular endothelium by HIV.

Clinical Picture. HIV retinopathy is a benign and transient condition, although some patients may develop vision defects associated with it.

Complaints. It is usually asymptomatic and transient, but as the condition progresses, common presenting complaints may include:

— blurred vision;
— floaters;
— flashing lights;
— visual field defect;
— decreased visual acuity;
— changes in color vision.

Signs. The most commonly ophthalmoscopic findings considered as early signs of HIV infection are:

— cotton-wool spots;
— retinal microaneurysms;
— intraretinal hemorrhages;
— Roth spots (white-centered hemorrhages).

Methods of Examination. VA, slit-lamp exam, ophthalmoscopy, funduscopy, color fundus photography, perimetry, fluorescein angiography, serological test for HIV.

Differential Diagnosis. Diabetic retinopathy, hypertensive retinopathy, ocular ischemic syndrome, vasculitis, CMV retinitis.

Treatment. Because of benign character HIV retinopathy does not require any specific therapy, but it can be a symptom of HIV infection progression that requires initiation of antiretroviral therapy (ART). ART cannot cure AIDS but helps to manage symptoms and improve the quality and length of life for people infected with HIV.

Prognosis for vision is very good, spontaneous resolution within 1—2 months with ART.

Complications. Retinal vein and artery occlusions, an increased risk for opportunistic infections (cytomegalovirus retinitis).

Prophylaxis. Periodic fundus screening examinations are indicated in the presence of known HIV infection to differentiate this condition from a more serious one. Early and proper ART can salvage vision and improve the quality of life.

The Eye and Cytomegalovirus

Cytomegalovirus (CMV) is a very common virus belonging to the herpes family, which also includes the viruses that cause herpes, chicken pox, and mononucleosis. The virus is widespread, between 50—80 % of the world's population become infected with CMV by the time they are 40 years of age. Usually CMV is a mild disease that does not cause any serious problems in healthy children and adults. Most people get flu-like symptoms if they develop any symptoms at all. Then the virus usually becomes inactive (latent or dormant), however, it can reactivate in situations with immune suppressions and be serious and even fatal to people with HIV and AIDS,

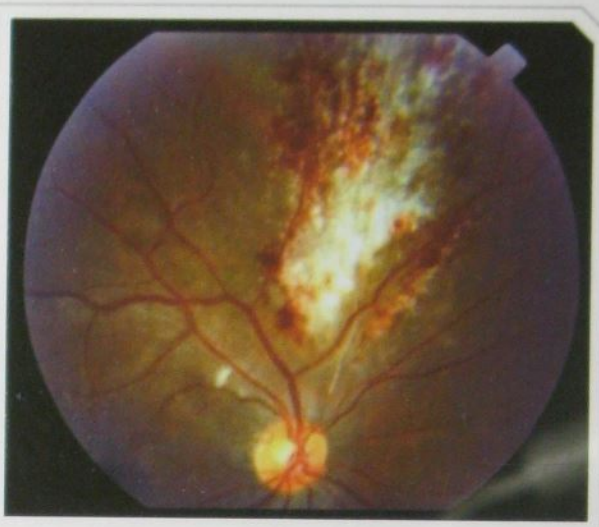


Fig. 18.13. Cytomegalovirus retinitis with a typical "pizza pie appearance" on the fundus picture (from https://openi.nlm.nih.gov)

newborns, the elderly, people taking chemotherapy for cancer, recipients of organ transplants.

Reactivated CMV infection can infect many organs in the body causing pneumonia, ulcers, hepatitis, intestinal obstruction, colitis, encephalitis, which may cause seizures or coma. One of the most common ocular complications is CMV retinitis.

CMV retinitis is a serious sight-threatening inflammation of the retina that develops from CMV infection. Common symptoms of the disease include blurred vision, floaters, blind spots, flashes, and sudden loss of peripheral vision. Some people with CMV retinitis may have no symptoms. Slit-lamp examination reveals mild vitreous inflammation. Ophthalmoscopic findings reveal peripheral scattered yellow-white areas of retinal necrosis and hemorrhages, often described as "pizza pie appearance" (fig. 18.13).

Treatment is directed at retaining of present vision and limiting retinal damage. CMV retinitis is treated with anti-viral drugs (Acyclovir, Ganciclovir, or Foscarnet) that can be administered orally, intravenously, injected directly into the eye or through an intravitreal implant (small capsules of medications surgically inserted into the eye). These drugs can slow down the progression of the disease, but they cannot cure it.

Prompt care and treatment is absolutely essential to control the disease. If left untreated, CMV retinitis can cause detachment of the retina and lead to irreversible blindness in just two to six months.

The Eye and Herpes

Herpes simplex is a common recurrent infection of the skin or mucous membrane caused by the herpes simplex virus (HSV) and characterized by formation of small vesicles in clusters. There are two major types of HSV. Type 1 is the most common and mainly associated with facial infections (mouth, lips, and eyes). It is referred to as "cold sores" or "fever blisters", because recurrences are often caused by a febrile illness, such as cold. Type 2 is a sexually transmitted form of herpes infecting the genitals. Both types of herpes simplex can spread to the eye and cause infection, but type 1 is the most frequent cause of eye infections, which can be transferred to the eye by touching an active lesion (a cold sore or blister) and then the eye.

Although HSV-1 infection can occur at any age, most people get their first infection in early childhood with flu-like symptoms. After the first infection, the virus remains dormant in the nerve cells in the brain or spinal cord and is present for life. The virus can be triggered to become active again by physical or emotional stress, sun exposure, upper respiratory tract infections, hormonal changes, immunosuppression, trauma or surgery. It results in painful clear blisters on a red base, usually on the face or lips, which crust and heal within a few days.

HSV may affect any part of the eye causing a simple infection or a condition that can possibly result in blindness. It typically infects the conjunctiva, eyelids, and surrounding skin causing conjunctivitis, blepharitis, and dermatitis respectively. Another sign of ocular herpes is inflammation of the cornea, or herpetic simplex keratitis. It can be superficial, involving the epithelium (epithelial keratitis) of the cornea, and usually heals without scarring, or it can involve deeper layers of the cornea (stromal keratitis) and lead to scars of the cornea, loss of vision, and sometimes even blindness. Less common but severe forms of ocular herpes are iridocyclitis and retinitis.

Herpetic simplex keratitis is an inflammation of the cornea caused by the virus of herpes simplex. It is the most common cause of corneal blindness in developed countries and normally affects only one eye at a time. Patients with HSV keratitis may complain of eye pain, redness, tearing, foreign body sensation, photophobia, blurred vision. Treatment of herpetic simplex keratitis depends on the severity of the condition and may include oral or topical antiviral medications, steroid eye drops. In case of severe corneal scarring with vision loss corneal transplantation may restore vision.

Read more about the etiology, clinical picture, and methods of treatment for ***herpetic simplex keratitis*** *in the Chapter* ***Diseases of the Cornea (p. 263).***

The Eye and Syphilis

Syphilis is a sexually transmitted chronic systemic infection caused by the spirochete Treponema pallidum. If left untreated, the disease progresses through four stages, with a potential to cause significant morbidity to any major organ of the body.

Involvement of the eye is uncommon, being more frequent at the second or third stage of the disease and is often associated with delayed diagnosis and delayed treatment, which may result in irreversible visual loss and structural damage. The eyes are affected in approximately 10 % cases and the most common presentation of syphilis in the eye is uveitis. Other ocular manifestations include interstitial keratitis, scleritis, retinal vasculitis, neuroretinitis, and optic neuropathies.

Syphilitic uveitis is an inflammation of the uvea that occurs in patients with syphilis. Due to the varying degrees of presentation patients may complain of blurred vision, floating spots, light sensitivity, double vision, eye pain, redness, and foreign body sensation. The preferred treatment for all stages of syphilitic uveitis remains parenteral penicillin G for 10—14 days. Topical, or periocular, and systemic corticosteroids (depends on the type and severity of uveitis) are adjunctive therapy to reduce

ocular inflammation related to syphilis. With proper diagnosis and prompt antibiotic treatment, the majority of syphilis cases can result in a cure.

Read more about the etiology, clinical picture and methods of treatment for ***syphilitic interstitial keratitis*** *in the Chapter* ***Diseases of the Cornea (p. 276)*** *and* ***syphilitic uveitis*** *in the Chapter* ***Diseases of the Uvea (p. 333).***

The Eye and Toxoplasmosis

Toxoplasmosis is an infection caused by the single-celled parasite Toxoplasma gondii, one of the world's most common parasites. Members of the cat family are its definitive hosts, other species, including mammals, birds, and reptiles, may serve as intermediate hosts. The parasite can be found in the host's tissues, such as muscles and body fluids (saliva, milk, urine, and feces). Humans can acquire toxoplasmosis through food (e.g., raw or undercooked meat, unpasteurized milk, contaminated water, unwashed fruit and vegetables), by coming into contact with infected cat litterboxes or sandboxes, or soil contaminated with cats' feces, and rarely dust in the air. Congenital toxoplasmosis is transmitted from the mother to the fetus across the placenta during pregnancy.

Toxoplasmosis may cause flu-like symptoms in some people, but most people affected never develop signs and symptoms, because a strong immune system keeps the parasite in an inactive state. For infants born to infected mothers and for people with weakened immune systems, toxoplasmosis can cause extremely serious complications, including irreversible vision loss and neurological problems.

Toxoplasmosis can affect any organ in the body: the lymph nodes, lungs, liver, skeletal and heart muscles, but the eyes and the brain are most commonly involved. The most common part of the eye to become affected is the retina and choroid (retinochoroiditis), which may lead to partial loss of vision or to blindness.

Toxoplasmic retinochoroiditis, or ocular toxoplasmosis, is an infection of the back of the eye caused by Toxoplasma gondii. It is a recurrent condition. The inactive parasite may later reactivate in individuals with compromised immune systems causing blurred vision, floaters, decreased visual acuity, sometimes photophobia, redness of the eye, and tearing. It can affect one or both eyes.

The goal of treatment is to arrest the multiplication of the parasite during the active period of retinochoroiditis and to minimize damage to the retina and optic nerve. However, treatment does not cure the condition but forces it back into a nonactive state.

Read more about the etiology, clinical picture, and methods of treatment for ***ocular toxoplasmosis*** *in the Chapter* ***Diseases of the Uvea (p. 331).***

The Eye and Tuberculosis

Tuberculosis (TB) is an infection caused by the bacillus Mycobacterium tuberculosis, which can cause disease in multiple organs throughout the body, including the

eye. Ocular tuberculosis is met rather frequently among the non-pulmonary forms of the disease and can affect any structure of the eye except the lens. It has a chronic course with exacerbations and remissions.

According to the pathogenesis ocular tuberculosis can be of three types:

— *primary ocular TB* is a condition of ocular involvement without evident systemic lesions. These are cases where the eye has been the initial portal of infection entry;

— *secondary ocular TB* is defined as ocular infection resulting from hematogenous spread from the lungs or contiguous spread from adjacent structures such as the sinus or the cranial cavity;

— *immune-mediated ocular TB* develops due to hypersensitivity to Mycobacterium tuberculosis antigens from a distant focus.

Nearly one-third of the world's population is latently infected with TB, and more than 9 million new cases are diagnosed each year, 95 % of them in developing countries. Ocular TB may not be associated with clinical evidence of pulmonary TB; up to 60 % of patients with evidence of extrapulmonary TB may not have diagnosed pulmonary TB. Ocular TB may be an initial presentation of extrapulmonary dissemination of infection. Keratitis and posterior uveitis are the most common presentations of intraocular TB.

Read more about the etiology, clinical picture, and methods of treatment for ***tuberculous interstitial keratitis*** *in the Chapter* ***Diseases of the Cornea (p. 278)*** *and* ***tuberculous uveitis*** *in the Chapter* ***Diseases of the Uvea (p. 334).***

Examples of ocular TB manifestations are the following:

Eyelids

— Lupus of the skin over the lids, which starts as a minute nodule and develops to lupus (soft brownish TB nodules with ulceration and scarring).

— Eyelid abscess or chalazion-like mass with spontaneous drainage of abscess.

Lacrimal Apparatus

— Asymptomatic dacryoadenitis or dacryocystitis.

— Presentation may be clinically indistinguishable from bacterial infection.

Conjunctiva

— Mucoid discharge, eyelid edema, chemosis.

— Large ulcerating follicles.

— Small millet-seed ulcers, pedunculated polyps, tuberculoma of the conjunctiva.

Sclera

— Diffuse or nodular episcleritis with localized, sharp subconjunctival nodules.

— Diffuse or nodular scleritis with deep nodules.

Cornea

— Interstitial keratitis with ciliary congestion, stromal haziness, deep vascularization.

— Sclerokeratitis.

— Keratoconjunctivitis.

Orbit

— Osteomyelitis.
— Orbital cellulitis or tuberculoma in the orbit.
— Endophthalmitis and panophthalmitis.

Uvea

— *Anterior uveitis:*
  • *non*-granulomatous anterior uveitis with ciliary congestion, fine keratic precipitates, aqueous cells and flare, hypopyon and posterior synechiae;
  • g*ranulomatous anterior uveitis* with ciliary congestion, aqueous cells and flare, mutton-fat keratic precipitates, iris nodules, broad posterior synechiae.
— *Choroiditis* with exudates in the vitreous, patches of yellowish white nodules, retinal edema, hemorrhages, and macular edema:
  • *serpi*ginous-like choroiditis (SLC) with multifocal lesions that progress in a serpiginoid pattern, individual lesions heal centrally and show centrifugal spread in ameboid fashion, mild vitreous inflammation;
  • choro*idal tubercles* are characterized by yellowish lesions with poorly defined borders and typically elevated in the center; they are commonly situated in posterior pole, if are present near or at macula VA may be diminished; inflammatory cells and subretinal fluid may be present.

Retina

— Exudative retinitis.
— Tuberculous periphlebitis (Eales disease) with vitreous hemorrhages, proliferative vitreoretinopathy, tractional retinal detachment.

4.6. Ocular Manifestation of Nutritional Deficiencies

Vitamins and minerals play an important role in maintaining the eye health as well as vision in general. Most of them cannot be made by the body, and can only be obtained from food and supplements, so it is why they are an essential part of the diet. Fortunately, eye problems from nutritional deficiency are rarely found in the developed countries.

Vitamin and mineral deficiency symptoms arise when there is a lack of any vitamins. They usually develop slowly and can initially be so mild as to be undetectable. When symptoms become obvious, health may have been affected for some time. Eye symptoms are among the early outward warning signs of vitamin and mineral deficiencies.

Vitamin A deficiency

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. Vitamin A deficiency also compromises the immune system, increasing the chance of death from malaria, measles, and diarrhea.

Ocular signs and symptoms of vitamin A deficiency are bilateral, although may be of different degree and include the following:

— poor adaptation to darkness — night blindness (nyctalopia);
— dryness of the conjunctiva and cornea, conjunctival xerosis;
— thinning and ultimately ulceration of the cornea — keratomalacia;
— appearance of slightly elevated white foamy lesions on the bulbar conjunctiva near the limbus, at the three o'clock or nine o'clock positions, more commonly on the temporal side — Bitot's spots;
— corneal perforation;
— blindness due to structural damage to the retina;
— increasing the risk of blindness in children with measles.

Vitamin B_1 deficiency

Vitamin B_1 deficiency or hypovitaminosis B_1 can lead to neurological and cardiovascular pathology, beriberi, cause skin sensitivity, digestive disorders, and progressive muscle weakness.

Ocular symtoms of vitamin B_1 deficiency may include:

— dry eye syndrome;
— epithelial keratitis;
— retrobulbar neuritis;
— optic nerve atrophy;
— dichromatism;
— visual field scotomas;
— paresis or paralysis of the oculomotor nerves (ophthalmoplegia);
— nystagmus.

Vitamin B_2 deficiency

Vitamin B_2 deficiency or hypovitaminosis B_2 primarily affects the mucous membranes and leads to the classic triad of the oral-oculo-genital syndrome. The oral lesions cosist of numerous microdamages of the lips with cracking at the corners accompanied by severe pain when opening the mouth, tongue inflammation, angular stomatitis, dermatitis, etc. Prolonged deficits may manifest as cerebral insufficiency with general weakness, dizziness, trembling, and delayed mental response.

Ocular symptoms and signs differ according to the severity of the condition and may include:

— itching and irritation of the eyes;
— lacrimation;
— light sensitivity;
— blurring of vision;
— blepharoconjunctivitis;
— keratitis;
— peripheral corneal neovascularization;
— cataract;
— retinal hemorrhage.

Vitamin C deficiency

Vitamin C is a powerful antioxidant and is essential for proper functioning of many different systems of the body, including the eyes. Chronic, severe deficiency of vitamin C results in scurvy, which is characterized by hemorrhagic manifestations and abnormal bone and dentine formation.

Typical ocular presentations of vitamin C deficiency are:

— hemorrhages in various parts of the eye — lid, conjunctiva, anterior chamber, vitreous cavity, or retina;
— corneal ulcer;
— neuroparalytic keratitis;
— paralysis of the ocular muscles;
— delayed healing of eye damages.

Vitamin D deficiency

Vitamin D deficiency results in rickets in young children; irritability, weakness, and softening of the bones in adults. Vitamin D may have a direct effect on the eyes; it is vital for maintaining overall good health.

The main ocular signs of vitamin D deficiency are:

— phlyctenular keratitis;
— blepharospasm;
— cortical cataract;
— orbital osteomalacia;
— age-related macular degeneration;
— retinopathy;
— optic nerve athrophy.

Review:

1. Key Points

The human body is a complex multifunctional system and changes or diseases in one or another system or organ may affect the eyes too. They may involve any structure of the eye, including the eyelids, conjunctiva, cornea, sclera, uvea, and retina. Occasionally the ocular findings may be the first indication of an underlying systemic disease, as nowhere else in the body a microcirculatory system can be directly visualized, investigated with such precision, or the neural tissue so easily examined. As a result, examination of the eye provides invaluable information for the diagnosis and monitoring of systemic diseases, the effectiveness of their treatment and prognosis. At the same time, early diagnosis of the eye and vision changes is also a key to successful prevention of visual impairment and blindness.

The main symptoms of retinal disorders in systemic diseases are blurred vision, reduced visual acuity, floaters, flashing lights, visusal field defects. *The main examination methods* are VA, slit-lamp exa, ophthalmoscopy, color fundus photography, perimetry, fluorescein angiography, optical coherence tomography.

Among cardiovascular diseases the most common conditions that can cause retinopathies are hypertension, kidney diseases, toxemia of pregnancy, atherosclerosis. *Hypertensive retinopathy* according to the characteristic retinal changes can be classified into different stages (mild, moderate, malignant) that allows to evaluate the risk of cardiovascular morbidity and mortality. Prognosis for vision is good with treatment of the underlying problem, but in cases of malignant retinopathy — poor not only for vision but also for life. *Renal retinopathy* has the same clinical picture as hypertensive retinopathy but has a bad prognostic sign for the life of the patient.

Retinopathy can be caused by hematological diseases such as anemia and leukemia or endocrine diseases such as diabetes, thyroid gland disorders. *Diabetic retinopathy* is the most common form of eye problems affecting people with diabetes and a leading cause of visual loss worldwide. It can be of two clinical stages — nonproliferative or proliferative, named so for the absence or presence of abnormal new blood vessels growing in the retina. Left untreated, diabetic retinopathy results in complete blindness. Treatment options include focal photocoagulation, panretinal photocoagulation, intravitreal steroid and/or anti-VEGF drugs injections, vitrectomy.

Systemic infectious diseases may have ocular complications: they are AIDS, CMV, herpes simplex, syphilis, toxoplasmosis, tuberculosis. Eye problems result from nutritional deficiencies. Treatment of the underlying disease and causative factors are the main options for releaving ocular disorders and restoring visual functions.

2. The Review Questions

A. Control Questions

1. What methods of fundus examination do you know?
2. What systemic diseases may cause ocular disorders?
3. What are hypertensive retinopathy and its classification?
4. What ophthalmoscopic signs of hypertensive retinopathy do you know?
5. What ophthalmoscopic picture is characteristic for renal retinopathy?
6. What are diabetic retinopathy and its main clinical stages?
7. What ophthalmoscopic signs are characteristic for different stages of diabetic retinopathy?
8. What are treatment methods of diabetic retinopathy?
9. What are the ocular manifestation of AIDS, its clinical picture, treatment methods, and prognosis?
10. What ocular manifestations of tuberculosis do you know?

B. Tests

1. The main examination methods of retinal disorders in systemic diseases are:

A. VA test
B. Slit-lamp exam
C. Ophthalmoscopy
D. Tonometry
E. Perimetry
F. Fluorescein angiography

2. What are the main ophthalmoscopic signs of hypertensive retinopathy?

A. Arteriolar narrowing
B. Gunn's sign
C. Copper and silver wire sign
D. Cotton-wool spots
E. Hemorrhages
F. Hard exudates

3. What are the main ophthalmoscopic signs of renal retinopathy?

A. Arteriolar narrowing
B. Retinal edema
C. Edema of the optic disc
D. Cotton-wool spots
E. Hemorrhages
F. Hard star-shaped exudates on the macula

4. What ocular complications of diabetes mellitus can cause significant visual impairment?

A. Lid styes and cellulitis
B. Cataract
C. Glaucoma
D. Diabetic retinopathy
E. Extraocular muscle palsy
F. Iridocyclitis

5. What are the complaints of patients with diabetic retinopathy?

A. No complaints
B. Blurred vision
C. Floaters
D. Distortion
E. Flashing lights
F. Sudden loss of vision in one or both eyes

6. What are the main ophthalmoscopic signs of nonproliferative diabetic retinopathy?

A. Microaneurysms
B. Hemorrhages
C. Retinal edema
D. Hard exudates
E. Cotton-wool spots
F. Neovascularization

7. What are the main ophthalmoscopic signs of proliferative diabetic retinopathy?

A. Microaneurysms and hemorrhages
B. Hard exudates
C. Cotton-wool spots
D. Neovascularization
E. Vitreous hemorrhages
F. Tractional retinal detachment

8. What are the treatment options of diabetic retinopathy?

A. Normalization of blood glucose and blood pressure
B. Laser photocoagulation
C. Laser panretinal photocoagulation
D. Intravitreal steroid injections
E. Intravitreal anti-VEGF drugs injection
F. Vitrectomy

9. What are the main ophthalmoscopic signs of AIDS-associated retinopathy?

A. Microaneurysms
B. Hemorrhages
C. Cotton-wool spots
D. Hard exudates
E. Roth spots
F. Neovascularization

10. What are the ocular complications of tuberculosis?

A. TB nodules over the eyelid skin
B. Dacryoadenitis or dacryocystitis
C. Pedunculated polyps and tuberculoma of the conjunctiva
D. Diffuse or nodular episcleritis or scleritis with nodules
E. Interstitial keratitis
F. Uveitis

C. Clinical Cases

Case 1

A 52-year-old man presents with headaches and blurred vision. Physical examination is unremarkable except for blood pressure, which is 190/118 mm Hg. Retinal examination reveals retinal arteriolar narrowing and increased retinal arteriolar light reflexes, retinal "hard" exudates, cotton-wool spots, and flame retinal hemorrhages. What is the diagnosis?

Case 2

A 46-year-old woman presented with gradual blurring of vision in both eyes over a period of two weeks. This was preceded by six weeks of intermittent headaches, which were aggravated by eye straining to focus when reading. Fundus examination showed narrowing of the arterioles with arteriovenous nipping, multiple cotton-wool spots at the posterior pole, a macular star with mild macular edema and bilateral papilledema with splinter hemorrhages. Her blood pressure was recorded as 230/130 mm Hg with a pulse rate of 84 beats per minute. Define the diagnosis and your following actions.

Case 3

A 73-year-old man presented to the ophthalmologist with blurring of vision, visual field scotomas, and sudden visual loss in one eye lasting just for seconds or minutes. Fundus examination revealed thickening of the arteriolar walls, copper and silver wire sign, retinal hemorrhages, cotton-wool spots, the optic discs are pale-pink with distinct margins. What is the diagnosis?

Case 4

A 28-year-old woman presents with visual blurring in her left eye. She has a 2-month history of easy bruising and increasing fatigue accompanied by unexplained weight loss. Retinal examination reveales a pale fundus with a yellowish tint, flame and blot hemorrhages scattered throughout the retina, neovascularization and edema of the optic disc with blurring of its margin. Make a diagnosis.

Case 5

A 64-year-old overweight man with type 2 diabetes mellitus is seen for routine examination. Both retinas show multiple scattered intraretinal hemorrhages of dot and blot shape, multiple microaneurysms, hard exudates of various sizes, cotton-wool spots and retinal edema. What is the diagnosis?

Case 6

A 29-year-old man who has suffered from type 1 diabetes since the age of 13 years came for an annual ophthalmoscopic exam. Fundus examination revealed microaneurysms, hemorrhages, cotton-wool spots, irregularly dilated vessels, venous beading and loops, and diffuse leakage in the macular region. Make a diagnosis.

Case 7

A 48-year-old woman who has suffered from type 1 diabetes since the age of 11 years came for a regular eye exam. Fundus examination revealed neovascularizations on the optic disc and temporal to the macula as well as a few microaneurysms, preretinal and vitreous hemorrhages, and tractional retinal detachment. What is the diagnosis?

Case 8

A 33-year-old HIV-infected woman presented with blurred vision and floaters in her left eye. On examination, she had obvious decreased visual acuity in her left eye. Fundus examination revealed cotton-wool spots, microaneurisms and hemorrhages, Roth spots and hyperemic discs with telangiectasia, and blurring of the disc margins, more prominent in the left eye.

Case 9

A 30-year-old man was diagnosed with HIV infection three years ago and started antiretroviral therapy. He presented to the ophthalmologist with blurring of vision, floaters and flashes. Ophthalmoscopic examination revealed peripheral scattered yellow-white areas of retinal necrosis and hemorrhages ("pizza pie appearance"). What is the diagnosis?

Case 10

A 24-year-old woman came for a routine ocular exam. Ophthalmoscopic picture revealed that the optic disk is pale pink, round, situated on the same level with the retina, the disc margins are distinct, the ratio of the arteries to the veins is 2:3. The area of the macula presented as a horizontal oval of red color. How do you interpret this fundus picture?

CHAPTER 19

Disorders of Ocular Motility And Strabismus

OBJECTIVES

Upon completion of the chapter the students should:

- know the main principles of binocular vision and requirements for its formation;
- know the types of ocular motility disorders;
- know the basic diagnostic methods of ocular motility disorder evaluation;
- describe the clinical sings and complications of strabismus;
- evaluate and manage patients with strabismus;
- know the main principles of strabismus treatment.

Plan:

1. **BINOCULAR VISION AND REQUIREMENTS FOR ITS FORMATION**
2. **CLASSIFICATION OF OCULAR MOTILITY DISORDERS**
3. **EXAMINATION METHODS**
4. **DISORDERS OF OCULAR MOTILITY AND STRABISMUS**
   - **4.1. Concomitant Strabismus**
   - **4.2. Incomitant Strabismus**
   - **4.3. Nystagmus**

1. Binocular Vision and Requirements for Its Formation

Binocular vision (or stereoscopic vision) may be defined as simultaneous vision with both eyes. Normal binocular vision is attained when both eyes are directed towards an object so that the images are projected on the fovea. The eyes receive their own images of the object, and these images are slightly dissimilar — that is based on the horizontal separation of the two eyes in the skull. However, the images are merged by the brain into a single visual image due to the process of fusion. So, as the result a person sees only one object.

Binocular vision is a very important visual function. It enables to determine the distance from one object to another, depth, volume and relative position of objects. It also allows for stereopsis, or the ability to see in three dimensions. Binocular vision expands the visual field and increases VA by 0.1—0.2. Humans have about 120° of the binocular field of view. The lack of binocular vision limits the professional suitability of man.

There are three distinct levels of binocular vision quality:

— simultaneous vision — the retina of both eyes perceives two images simultaneously;
— fusion — two retinal images blend into a single one;
— stereoscopic vision — perception of depth (single three-dimensional visual perception).

Binocular vision develops later than other visual functions such as light sensitivity, central vision, color vision, peripheral vision. Newborn babies have no coordinated eye movements; that occurs only at 2—3 weeks, so binocular vision is absent. At the age of 6—8 weeks a baby can fix an object with both eyes and keeps watching at it, and a 3—4-month-old baby has a rather steady binocular fixation. By 5—6 months of age the fusion reflex is formed. Binocular vision is considered formed by 3—4 years of age and is finally established in six- to seven-year-old children. Therefore, preschool age is the most dangerous for the development of binocular vision disorder (formation of strabismus). Full development of stereoscopic vision ends by 12 years old.

EYE FACTS

Binocular vision allows predators to judge the distance between themselves and their prey.

Formation of normal (steady) binocular vision requires the following conditions:
— the same visual acuity in both eyes (not less than 0.3—0.4);
— the same degree of refraction in both eyes that produce images of an equal size on the retina;
— transparency of the optical media;
— precise coordination of movement between the two eyes (the eye muscular system);
— location of both eyes in one frontal and horizontal plane;
— normal functional ability of the retina, pathways, and higher visual centers.

2. Classification of Ocular Motility Disorders

Ocular motility disorders can be classified according to:
— *the age of onset:*
  • congenital (infantile);
  • acquired;
— *the underlying etiology:*
  • primary;
  • secondary;
  • recurrent;
  • residual;
  • consecutive;
— *fixation:*
  • unilateral (monocular);
  • intermittent (alternating);
— *the duration of deviation:*
  • constant;
  • intermittent;
  • periodic;
— *the manifestation of deviation:*
  • false strabismus (apparent or pseudostrabismus);
  • true strabismus:
    – latent (heterophoria);
    – manifest (heterotropia);
— *direction of deviation:*
  • horizontal:

  - eso- (inward deviation or convergent);
  - exo- (outward deviation or divergent);
- vertical:
  - hyper- (upward deviation);
  - hypo- (downward deviation);
- torsional (cyclo-);

— *variation with the position of gaze:*

- concomitant:
  - accommodative;
  - partial accommodative;
  - non-accommodative;
- incomitant:
  - paralytic;
  - pseudo-paralytic (restrictive).

3. Examination Methods

— Ocular motility evaluation.
— Unilateral and alternating cover test.
— Hirschberg corneal light reflex test.
— Kalfa method or two pencils test.
— Sokolov test (test with "a hole in the palm").
— Test to read with a pencil (or pen).
— Belostotskiy-Friedman 4-dot color test.
— Bagolini striated glass test.

4. Disorders of Ocular Motility and Strabismus

The correct position of the eyes and their aligned motility required for the normal functioning of the visual analyzer are provided by the complex anatomical structure and functional organization of the oculomotor system. Disorders of the system are polyetiological. Some are present at birth, other may develop over time and be associ-

ated with ocular diseases or trauma, orbital floor fracture, weakness or paralysis of the eye muscles, neurological and degenerative neurological impairment, stroke, endocrine and systemic conditions, facial and brain tumors. The most common oculomotor system disorders are strabismus and nystagmus, which are observed in 15—35 % of people.

Strabismus, also known as squint, is misalignment of the visual axis of the eyes. In other words, it is a condition in which both eyes do not look at the same object at the same time. That is, one eye looks in one direction while the other eye turns in another direction.

Strabismus falls into one of four groups depending on the direction of the eye's deviation: inwards, outwards, up or down. In some cases, the same eye may turn each time, while in other cases, the eyes may alternate turning. Although strabismus can develop at any age, it usually starts in childhood. Up to 5 % of children are estimated to have strabismus.

Strabismus can be false or true. ***False strabismus***, also known as apparent strabismus or pseudostrabismus, is the false appearance of misaligned eye. This is common in infants and young children due to peculiarities of their facial structures. The wide bridge of the nose and small folds of the eyelid skin on the nasal side of the eyes (epicanthus) contribute to this appearance by covering the sclera of the eye on that side. This is especially noticed when the infant looks to the right or left. False strabismus requires no treatment. As the child grows, this appearance will improve and disappear.

To tell the difference between true and false strabismus the corneal light reflex test is used. If both eyes are aligned, the light reflection will appear in the center of each pupil. If a child has true strabismus and the eyes are not properly aligned, the reflection will appear in a different location in each eye. Because the light is not affected by the width of the nose or the folds of the eyelids skin a child with pseudostrabismus will have a normal reflection.

True strabismus is a state when one or both eyes deviate from the central visual axis. This deviation can be latent or manifest.

Latent strabismus (heterophoria) is a condition when deviation of the eye is noticeable only when one eye is covered and excluded from the binocular vision act. Without covering, deviation of the covered eye disappears due to muscular effort. This causes visual fatigue and headache. It is mostly due to refractive errors, and correction may relieve the condition. Latent strabismus may also appear under certain conditions such as fatigue, stress, illness, or when a person becomes lost in thoughts.

Orthophoria is a condition of perfect alignment of the two eyes, which is maintained even after the elimination of the influence of fusion.

Manifest strabismus (heterotropia) is a condition when deviation of the eye is noticeable with both eyes open. It can be constant or intermittent and may involve one eye or both eyes. Manifest strabismus is divided into 2 broad types: concomitant and incomitant (non-concomitant) strabismus.

4.1. Concomitant Strabismus

Definition. Concomitant strabismus, or comitant strabismus (from the Latin *comitare* — to accompany) is a condition when the deviating eye accompanies the leading eye in every direction of movement. The angle of deviation of the visual axis of the deviating eye remains the same in relation to the other eye in all directions of gaze and there is no associated limitation of ocular movement.

Etiology. Concomitant strabismus is commonly associated with children (6 months to 3—4-years-old). It may be idiopathic, developing in a child with normal eyes, or may arise secondary to another eye problem. It can result from refractive errors, anisometropia, corneal opacities, cataract, macular diseases, optic atrophy, congenital anomalies of the shape and size of the orbit or extraocular muscles and retinoblastoma.

Risk factors for developing squint include family history, prematurity, neurological illness (meningitis, encephalitis, cerebral palsy). Medical conditions such as Down syndrome, stroke, brain tumors or head injury can also cause strabismus.

Clinical Picture. Concomitant strabismus may occur as monocular strabismus, in which only one eye deviates, or as alternating strabismus, in which both eyes deviate alternately. By the duration, with which it occurs, it can be either constant or intermittent.

Depending on the direction of the patient's eye deviation various types of concomitant strabismus are distinguished (table 19.1, fig. 19.1). They are:

— esotropia — convergent strabismus (inward turning of the eye);
— exotropia — divergent strabismus (outward turning of the eye);
— hypertropia — supravergent strabismus (upward turning of the eye);
— hypotropia — infravergent strabismus (downward turning of the eye);
— cyclotropia — torsional strabismus (wheel rotation of the eye).

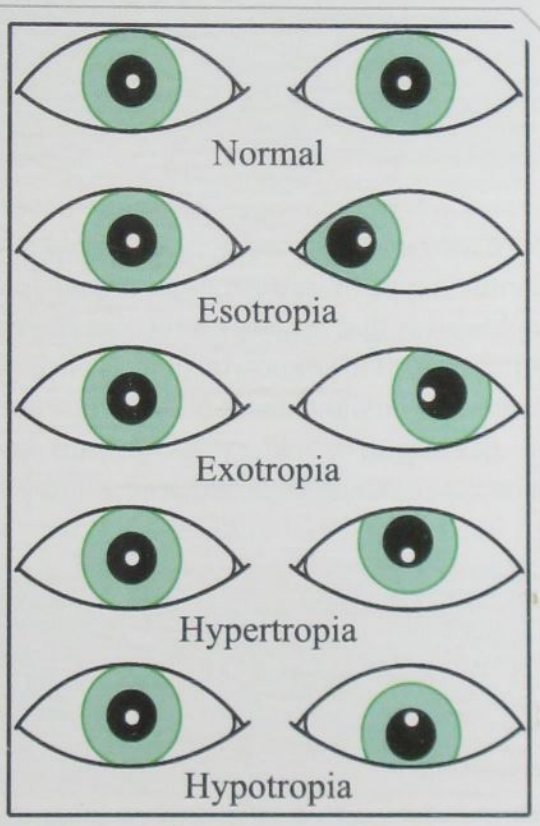


Fig. 19.1. Deviation of the eyes assessed by the Hirschberg test (corneal light reflex) (from http://medical-dictionary.thefreedictionary.com)

Table 19.1

Types of Strabismus Depending on the Direction of the Eye Deviation

Prefix-	Type of Strabismus		Description
	Latent (-phoria)	Manifest (-tropia)	
Eso-	Esophoria	Esotropia	Inward deviation of the eye
Exo-	Exophoria	Exotropia	Outward deviation of the eye
Hyper-	Hyperphoria	Hypertropia	Upward deviation of the eye
Hypo-	Hypophoria	Hypotropia	Downward deviation of the eye
*Cyclo-	Cyclophoria (incyclophoria or excyclophoria)	Cyclotropia (incyclotropia or excyclotropia)	Wheel rotation or torsional deviation of the eye

* Incyclo- — the eye rotates toward the nasal or medial side.
Excyclo- — the eye rotates toward the temporal or lateral side.

Esotropia, or crossed-eyes, is inward deviation of the eye (fig. 19.2). It is the most common type of strabismus in infants. In approximately half of cases it is caused by farsightedness that has not been corrected by glasses or contact lenses. The extra focusing effort needed to see nearby objects clearly may cause the eyes to turn inwards. Esotropia caused by farsightedness is called accommodative esotropia or refractive esotropia.

Exotropia, or wall-eye, is outward deviation of the eye (fig. 19.3). It is another common type of strabismus that occurs most often when a child is focusing on distant objects. It usually occurs intermittently rather than continuously and sometimes may be underestimated because of this.

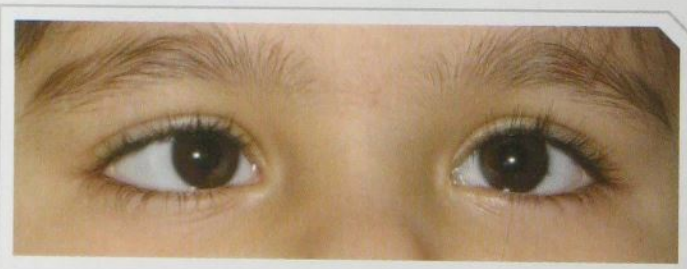


Fig. 19.2. Concomitant convergent strabismus (esotropia) (from https://bodlaeyecare.com)

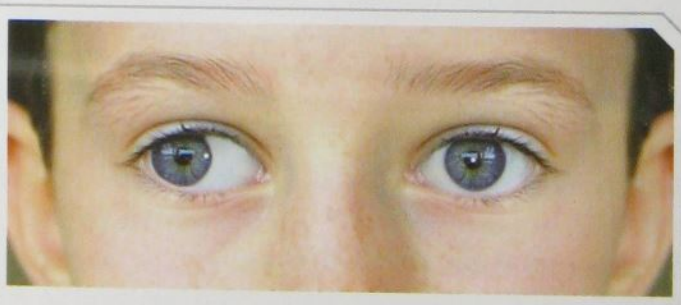


Fig. 19.3. Concomitant divergent strabismus (exotropia) (from http://leczeniezeza.pl)

Concomitant strabismus is divided into accommodative, partially accommodative or non-accommodative.

Accommodative esotropia is one of the most common forms of concomitant strabismus that occurs due to disorder of normal correspondence of accommodation and convergence (fig. 19.4).

Convergence of the eyes is controlled reflexively, along with focusing for near vision (accommodation). This normal integration is necessary to maintain eye alignment

and to focus on objects at various distances. With respect to the norm every accommodation diopter corresponds to one convergence meter angle. Patients with refractive errors requiring a greater than normal amount of accommodation (hypermetropia), and patients with inherently excessive reflexive convergence will converge too much and cross their eyes when accommodating. This means that the eyes must work harder to bring a close object into sharp focus. The closer the object is to the eye, the greater the amount of accommodation that is required. A side effect of the accommodative effort can be excess convergence or crossing of the eyes. Frequently, esotropia is present only when looking at near objects.

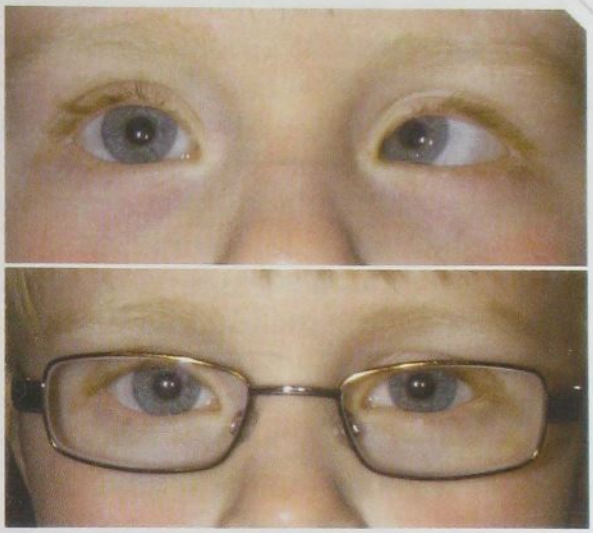


Fig. 19.4. Refractive accommodative esotropia, comparison of alignment without and with correction (from http://webeye.ophth.uiowa.edu)

Accommodative esotropia appears in children from six months to seven years of age, most commonly between two and three years of age. Rare cases have been reported from the age of 3 months to 11 years old. At the onset, the deviation is usually intermittent, but usually becomes constant in the following weeks to months.

If the angle of esotropia is fully corrected with glasses, the squint is said to be ***a fully accommodative esotropia***.

If the angle of the squint is not fully corrected with glasses, it is called ***a partially accommodative esotropia***. It is caused both by accommodation disorders and a hidden paretic component or may appear after surgical treatment of a non-accommodative strabismus.

If there is no change in the angle of the squint with long-sighted glasses, the squint is said to be ***a non-accommodative esotropia***. It may be caused by central nervous system (CNS) diseases and acquired visual impairment of one eye below 0.3 as result of corneal opacity, cataract, optic nerve atrophy, or trauma.

Some children have a large angle convergent squint when they look at a near object, but their eyes are straight or almost straight when they look at a distant target. This is known as ***a convergence excess esotropia***.

NOTE!

Not everyone with strabismus will have double vision.

The brain may quickly adapt to strabismus by ignoring visual information from the deviated eye. With time this can lead to permanent poor vision in the turned eye, a condition known as amblyopia or "lazy eye".

Complaints. Visible deviation of the eye, (cosmetic problems), the eyes that do not align in the same direction, uncoordinated eye movements, reduced vision, blurred vision, double vision, closing or covering of one eye when looking at something located at a close distance, loss of depth perception. Sometimes eye strain and fatigue, or headaches may be mentioned.

Signs of concomitant strabismus are:

— squinting;
— unsymmetrical points of reflection in each eye;
— the angle of deviation is constant regardless of gaze direction;
— primary (basic) angle of deviation (the angle of deviation of the squinting or affected eye) and secondary angle of deviation (the angle of deviation of the non-squinting or normal eye in case of squinting eye fixation) are equal;
— full range of eye motion, equal bilaterally;
— refraction anomalies — hyperopia, anisometropia, astigmatism;
— other ocular disorders — corneal opacity, cataract, retinoblastoma, etc.

Methods of Examination. Medical and family history; general inspection — facial symmetry, nose bridge, epicanthic folds and head position; VA; refraction; ocular movements evaluation; pupillary reactions — for detection of neurologic diseases; standard ophthalmic exam — slit-lamp exam, ophthalmoscopy to detect signs of corneal opacity, cataract, retinoblastoma; cover-uncover test; alternate cover test; estimation of angle of deviation — Hirschberg corneal light reflex; prism cover test; synoptophore test.

The prism cover test is an objective measurement of the angle of deviation with the help of prisms or prism bars, which contain a series of prisms of progressively increasing strength arranged one above the other. It combines alternate cover test with prisms. The apex of the prism must always point in the direction of deviation during the examination.

A patient is asked to fix on a near (33 cm) or distant (5 m) target with the leading eye. The examiner places prisms of different refractive power before the deviating eye and performs the cover/uncover test until the eye no longer makes any adjustment. This means that the angle of deviation corresponds to the strength of the respective prism and is fully compensated by that prism. To confirm that this is the end point indeed, the power of the prism is increased to check if the eye movement is reversed. The angle of deviation will be equal to the prism diopters (pd). One prism diopter is a deviation of the visual axes of 1 cm at a distance of 1 m.

Synoptophore (amblyoscope) is a basic orthoptic instrument for measurement of the objective and subjective angles of deviation, examination and training of binocular vision. It consists of chin and forehead rests with two cylindrical tubes with a mirrored right angle bend and contains a +6.50 D lens in each eyepiece, which optically sets the testing distance to about 5—6 meters. The synoptophore can be converted to a near test by adding —3.00 D lenses to induce accommodation. At the outer end of each tube there is a slide carrier, into which pictures are inserted, so that each eye is stimulated by a separate image. These tubes are placed horizontally and are movable in the horizontal and vertical planes. Therefore, any horizontal, vertical or torsional deviation can be measured in nine positions of gaze.

The objective and subjective angles of strabismus are measured with the aid of pictures for simultaneous perception. The objective angle of strabismus is measured by the disappearance of fixation movements of the eyes fixing two supplementing pictures. These pictures are called simultaneous perception pictures (e.g. a lion in a cage). The subjective angle is the angle that forms when the child places the lion into the cage. If both angles are identical, the correspondence of both retinas is normal. If the value of the objective angle differs from that of the subjective angle, anomalous retinal correspondence is present. The difference between the subjective and objective angles of strabismus is called the angle of anomaly.

Differential Diagnosis. Pseudoesotropia, Duane syndrome, incomitant strabismus.

Treatment should be started immediately after concomitant strabismus is diagnosed irrespective of the child's age. It is usually a long-lasting process, which requires a lot of patience and close cooperation of the physician with the treated child and his/her parents.

Treatment of strabismus aims to correct the misalignment of the eyes and to restore normal vision, i.e., visual acuity and binocular vision. It also improves the cosmetic appearance of the eyes.

Treatment for strabismus varies according to the type, severity, and cause of the disease. If it develops as a result of another ocular disease (for example, cataract), this needs to be treated.

Treatment modalities of concomitant strabismus include the following:

1. Optical correction with eyeglasses or contact lenses. This is necessary for children who have strabismus due to uncorrected refractive errors (farsightedness or astigmatism). Contact correction of ametropia is possible from the very first months after birth. Children can use spectacles from seven—nine months of age. Prism spectacles can be used for strabismus correction in 3—4-year-old children for some conditions such as constant strabismus angle. Prisms move the image of the fixing object onto the central retina fossae of both eyes, correct strabismus, recover binocular vision. The prism base is directed to the side opposite to the strabismus direction — temporal in case of convergent and nasal in case of divergent strabismus. Fresnel plastic prisms and their modifications are also used.

2. Patching or occlusion of the "good" eye. In children, the eye that is not properly aligned may not mature properly; and if misalignment is not corrected, permanent vision loss (amblyopia) can occur. In these cases, a patch is applied over the unaffected eye (fig. 19.5). This forces the child to

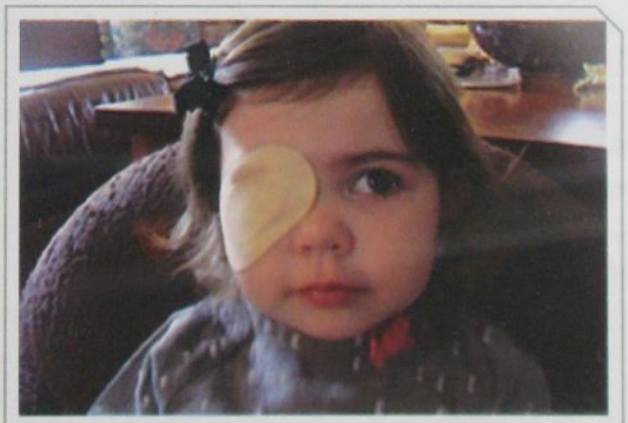


Fig. 19.5. Patching of the "good eye" for the treatment of amblyopia (from https://www.reviewofophthalmology.com)

fixate and use the affected eye that will help its visual development. The length of time the patch is worn depends on the severity of the condition and the age of the child.

Occasionally, cycloplegic drops may also be used in the good eye to temporarily blur the vision. This also forces the affected eye to fixate properly. These drops may be used as a substitute for patching.

3. Orthoptic therapy (eye exercises) is used to strengthen the eye muscles for improving eye coordination and eye focusing abilities. It is more useful for intermittent exotropia treatment. Vision therapy uses synoptophore, special computerized optical devices, including lenses and filters for monocular exercises of the amblyopic eye visual acuity as well as for binocular vision development (fig. 19.6).

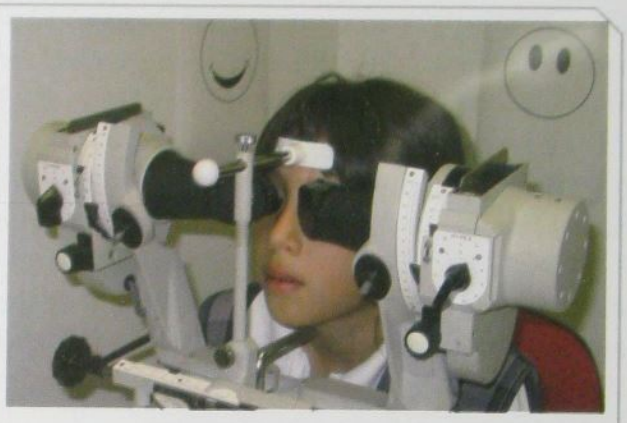


Fig. 19.6. Orthoptic therapy by means of a synoptophore (from http://medicalwiki.org)

This treatment is carried out in courses of everyday procedures. The treatment course lasts 10—15 days. For patients' ultimate recovery it is necessary to repeat such courses every 2 months during 1.5—2 years. It is the most effective for the treatment of 5—6-year-old patients. However, even under proper and forehanded care only in 20—30 % of the children ultimate recovery is observed.

4. Surgery is required when there is no identifiable cause, or in large squints that have not responded to conservative treatment. Immediate surgery (at the age of 8—12 months) is provided in case of congenital strabismus.

Surgery consists in weakening of the action of the enforced muscle in the direction of eye deviation or strengthening the action of the weakened antagonist muscle on the side opposite eye deviation. The most widely used operation aimed at weakening of the enforced muscle is recession surgery — muscle transplantation retrad to its usual location of junction with the sclera. To strengthen the weakened muscle the operation of its partial shortening, called resection, is carried out (fig. 19.7, 19.8). Very often both operations are carried out simultaneously. In case of alternating strabismus the surgery is usually performed simultaneously on both eyes; in monolateral strabismus only the squinting eye is operated. The recession and resection values are calculated in accordance with the magnitude of the deviation angle. Approximately 1 mm of recession or resection corrects 5—8 degrees of strabismus. The rates for successful realignment can exceed 80 %.

Often, children who had eye muscle surgery will also need vision therapy to improve eye coordination and to keep the eyes from becoming misaligned again.

Prognosis. With early detection, accurate diagnosis, and proper treatment, the prognosis for children with strabismus is excellent. The most satisfactory results are achieved if the condition is corrected before the age of seven years old.

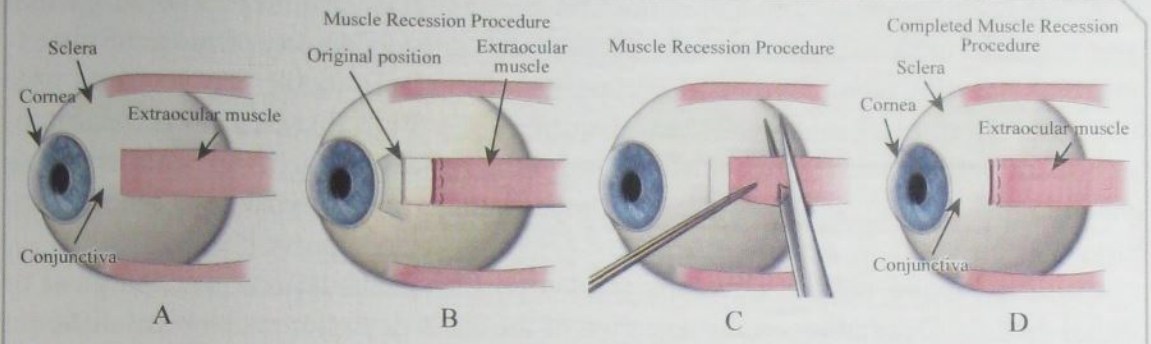


Fig. 19.7. Muscle surgery for strabismus treatment (from https://www.aapos.org): A — extraocular muscles; B — recession procedure, during which the muscle is cut from the surface of the eye and reattached a few millimeters behind depending upon the amount of weakening required; C, D — resection procedure, during which a small segment is cut from the extraocular muscle and the muscle is reattached at the original site of attachment on the globe. In resection, the extraocular muscle has not changed its position as in a recession, the only difference is its shorter length

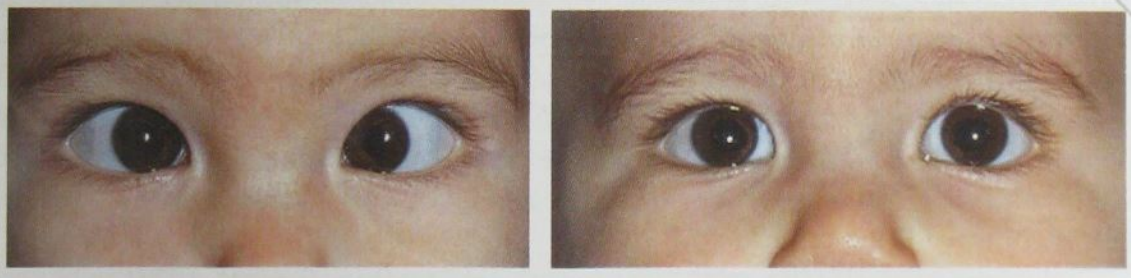


Fig. 19.8. Comparison of pre- and post-operative appearances of the child underwent bilateral medial rectus recessions (from http://webeye.ophth.uiowa.edu)

Complications. Blurry vision, double vision, poor three-dimensional (3-D) vision, amblyopia.

Prophylaxis. Strabismus cannot be prevented. However, complications of strabismus can be prevented if the problem is detected early and treated properly. Children should be monitored closely during infancy and preschool years to detect potential eye problems.

4.2. Incomitant Strabismus

Definition. Incomitant strabismus, or non-concomitant strabismus, is the condition in which the angle of deviation of the squinting eye varies with the direction of gaze and with the eye used for fixation.

Etiology. Incomitant strabismus may be congenital or acquired. The congenital type is due to birth injuries, some developmental anomaly of one or more of the extraocular muscles or of the neural component that serves them. The acquired type may result from head injury, blowout fracture of the orbit, diseases of the oculomotor system, inflammations, neoplasia, raised intracranial pressure, vascular lesions or systemic disease (e.g. multiple sclerosis, myasthenia gravis, thyroid eye disease, aneurysms).

Clinical Picture. Squint develops suddenly after recent trauma, neurological or orbital disease. Depending on the direction of the eye's deviation it can be of different type, but more common are eso- or exo-types, which are classified as "alphabet patterns": they are denoted as A- or V- or, more rarely, λ-, Y- or X-pattern depending on the extent of convergence or divergence when the gaze moves upward or downward.

Incomitant strabismus is usually seen in adults, although it may be also seen in infants and children, but much less commonly than the concomitant one.

According to the causative factor, incomitant strabismus is divided into paralytic or restrictive.

Paralytic strabismus is a squint caused by paralysis of the extraocular muscles due to defect of the III (oculomotor), IV (trochlear) or VI (abducens) cranial nerve. Each nerve may be affected at any point along its course from the brainstem to the orbit. In most cases there is no complete loss of function of the muscle, just partial loss and the condition is referred to as paretic strabismus.

In paralytic strabismus the affected eye will be deviated from the field of action of the involved muscle. For example, in palsy of the right VI cranial nerve the right eye will be adducted. Nerve palsy may be isolated or involve all the nerves, which results in immobility of the eye.

Diplopia is noticed if the paralysis is recent and usually accompanied by an abnormal head posture. It is important to make sure that this double vision disappears when either eye is closed — double vision that is maintained with one eye shut is due to another uniocular disease (i.e. refractive error or cataract).

Another symptom of the condition is confusion due to formation of an image of two different objects on the corresponding points of two retinas — the patient is not able to grasp or point at the object correctly on the side of action of the paralyzed muscle.

Restrictive strabismus is a condition caused by mechanical restriction of ocular movements that may be caused by fracture of the orbit, inflammatory diseases of the orbit tissue, tumors, or be of endocrine or aneurysmal origin. Apart from diplopia, discomfort or pain on eye movement is a common occurrence.

Complaints. Sudden onset of ocular deviation, restriction or total immobility of the eyes, diplopia, false orientation of the objects.

Signs of incomitant strabismus are:

— deviation of the eye;
— the angle of deviation differs in different directions of gaze;
— the secondary deviation angle is larger than the primary one;

— limitations or restriction of ocular movements;
— compensatory head posture (the head tilted in the direction of the affected muscle);
— VA is normal;
— diplopia;
— false projection;
— nausea or dizziness;
— may be accompanied by ptosis.

Methods of Examination. Medical history, general inspection of the eyes and head position, ocular motility test, VA, refraction, pupillary reaction, slit-lamp exam, ophthalmoscopy, cover tests, Hirschberg corneal light reflex, fixation test, Belostotskiy-Friedman 4-dot color test, synoptophore, forced duction test, orbital ultrasonography, skull X-ray, CT or MRI scan, neurological investigations.

The forced duction test is performed to differentiate between incomitant squint caused by paralysis of the extraocular muscles and that due to mechanical restriction of ocular movements. After topical anesthesia the affected eye is grasped near the limbus with two fixation forceps. Then the eye is gently rotated in the direction of a certain extraocular muscle to test for mechanical restriction. It may be possible to achieve the same result less traumatically by using a cotton wool bud (soaked in topical anesthetic) to push on the limbus in the desired direction. If ocular movements are free, then the test is negative and incomitant squint is caused by muscle palsy. If restricted, then the test is positive and the condition is a result of mechanical restriction.

Differential Diagnosis. Concomitant strabismus (table 19.2).

Table 19.2

Differences Between Concomitant and Incomitant Strabismus

	Concomitant	Incomitant
Age of onset	Typically, in childhood	In childhood and in adults
Type of onset	Usually slow	Usually sudden
Ocular movements	Full and equal bilaterally	Limited or restricted
Angle of deviation	Constant regardless of gaze direction	Varies with different gazes
Secondary to primary deviation	Secondary deviation equal to primary	Secondary deviation exceeds primary
Visual acuity	Reduced unilaterally	No change
Diplopia	Not always present	Present
Amblyopia	Usually present	Absent
Tilting head position	No	Yes

Treatment for incomitant strabismus is aimed at aligning the eyes and restoration of binocular vision.

1. ***Treatment of the underlying cause of the condition.*** If it is a neurogenic disorder, the patient must be referred to a neurologist. If it results from another ocular disease, it must be treated first. There can be improvement of the condition after the initial cause is cured.

2. ***Occlusion therapy.*** If diplopia troublesome, occlusion or patching of the affected eye is done.

3. ***Optical correction*** with the use of prisms (Fresnel) in glasses. With time, the prism is reduced in strength and eventually discarded.

4. ***Orthoptic therapy*** — although this type of strabismus does not usually respond well to it, especially if the deviation was caused by injury or a recent disease.

5. ***Medication.*** Injections of botulinum toxin type A may also be used to treat strabismus. The injection relaxes the contracted muscles in the eyes, making it easier for the eyes to focus where they need to.

6. ***Surgery*** is indicated only when recovery does not occur in 6—12 months to strengthen the paralyzed muscle by resection and weakening of the overacting muscle by recession or transposition/reposition procedures.

Prognosis depends on the underlying cause and is usually good after appropriate treatment of the causative disease.

Complications. Surgically over- or undercorrection, limitation of eye movements, diplopia.

Prophylaxis. Incomitant strabismus cannot be prevented.

4.3. Nystagmus

Definition. Nystagmus is fast, uncontrollable, and repetitive movements of the eyes, sometimes called "dancing eyes".

Etiology. There are two types of nystagmus: congenital and acquired. Congenital nystagmus develops in infancy, usually between six weeks and three months of age and then may disappear around 3 to 5 years old. It can be caused by a variety of eye conditions, including cataract, strabismus, and optic nerve hypoplasia or developmental neurological problems.

Acquired nystagmus may develop at any point in life depending on the underlying cause. This may include head injuries, stroke, brain diseases (i.e. multiple sclerosis, encephalitis, brain tumors), inner ear problems, certain medicines (i.e. lithium, anti-seizure medications), nutritional deficiency, alcohol or drug use. Many cases of nystagmus are idiopathic.

According to etiology nystagmus is divided into:

— optical — caused by eye diseases;

NOTE!

Nystagmus is a sign, not a disease. It may be the first sign of serious disorder of the brain. It is vital that the patient is referred to a neurologist when the condition develops for the first time.

— central or neurogenic — caused by CNS diseases;
— vestibular — associated with vestibular system diseases.

Clinical Picture. The movement of the eyes can vary in directions, between slow and fast motions, and it usually involves both eyes.

Nystagmus can be classified depending upon *the direction of motions* as:
— horizontal (side-to-side movements);
— vertical (up and down movements);
— rotary (circular movements).

According to the *type of eye motions* nystagmus can be:
— pendular (the speed of motions is the same in both directions);
— jerk (there is a slow and fast phase — the eyes move slowly in one direction and then seem to jerk back in the other direction);
— mixed (a combination of two types of nystagmus — pendular movements in the primary position of gaze (i.e. looking straight), but jerk movements on lateral gaze).

Nystagmus can be manifest — symptoms are present all the time; latent — symptoms are worsening when one eye is covered; fixation — occurs only when the eyes are focusing on an object; postrotatory — when the body is rotated and then stopped; miner's — after working in darkness for long periods. There are many other types of nystagmus.

Central, neurogenic, and vestibular types of nystagmus are mostly acquired and accompanied by symptoms of CNS and vestibular system disorders, such as sickliness, vomiting, faintness, oscillopsia, and overbalancing. These patients must be treated by proper specialists (neuropathologist, neurosurgeon, otolaryngologist).

Optic nystagmus is congenital and not accompanied by the mentioned above subjective symptoms. This kind of nystagmus occurs as a result of congenital or acquired during the first months of life sudden reduction of central vision of both eyes because of different retina or optic nerve diseases, opacity of the refractive medium as well as congenital pathology of the oculomotor system.

Complaints. Some people do not have complaints as their vision is not affected, only other people around them notice eye movements.

Most often complaints concern involuntary eye movements, troubles focusing and reading, sensitivity to light, sometimes blurry vision, loss of depth perception, difficulty seeing in darkness, holding head in the tilted position, dizziness, difficulties in balance and coordination.

Signs. Uncontrollable movements of the eyes, compensatory head posture, decreased central vision.

Methods of Examination. Medical history, general inspection of the eyes and head position, ocular motility test, VA, refraction, pupillary reaction, slit-lamp exam, ophthalmoscopy, ear exam, vestibular testing, neurologic exam, CT or MRI of the brain.

Differential Diagnosis. Strabismus.

Treatment. In most cases, there is no cure for nystagmus. Treatment helps to reduce symptoms and depends on the cause of the condition.

1. Treatment of the underlying cause. Sometimes there is a clear cause that can be addressed. For instance, a cataract may be treated or a tumor may be removed. If nystagmus is caused by a medicine, it may be stopped or replaced. Eliminating alcohol or drug abuse can also end the problem. When these underlying causes are treated, nystagmus may be cured.

2. Treatment of related vision problems. Contact lenses or eyeglasses can help correct problems such as nearsightedness or farsightedness. Special prisms may be used to adjust eye gaze. This helps to get the best possible vision and reduce symptoms.

3. Medicines. Botulinum injections may weaken some eye muscles, which helps reduce symptoms.

4. Surgery may be considered in some cases, especially for congenital nystagmus. The main goal of surgery is to alleviate a significantly abnormal head position or to ease nystagmus severity but it does not totally eliminate the condition.

In case of horizontal jerky nystagmus Anderson—Kestenbaum operation is effective if visual acuity significantly improves at forced head rotation. It consists of bilateral recession — resection surgeries on all the four muscles. When strabismus is not diagnosed, Anderson—Kestenbaum operation is carried out symmetrically on both eyes. When jerky nystagmus is combined with strabismus, Anderson—Kestenbaum operation is performed on the sighting non-squinting eye. As the result of Anderson—Kestenbaum operation forced head rotation is eliminated, visual acuity increases at straight head position, and in the primary position of the eyes nystagmus intensity is reduced.

Prognosis. Congenital nystagmus is usually a benign condition. It is not curable, but its symptoms can be diminished. Some types of congenital nystagmus resolve spontaneously before the child reaches school age. The prognosis for an acquired nystagmus depends on its cause. If the condition is due to a side effect of a drug, its dose can be reduced or the drug can be replaced to eventually resolve the nystagmus.

EYE FACTS

Of all the muscles in your body, the muscles that control your eyes are the most active. Your eye is constantly making tiny jerking movements called "microsaccades" to stop objects from fading from your vision.

Complications. Loss of focus, trouble reading, poor balance, blurry vision, and loss of vision in case of congenital pathology.

Prophylaxis. In general, nystagmus cannot be prevented. Acquired nystagmus may be reduced by prompt treatment of an underlying condition.

Review:

1. Key Points

Binocular vision is simultaneous vision with both eyes. It enables one to determine the distance from one object to another, the depth, volume, and relative position of objects. It expands the visual field and increases VA to 0.1—0.2. the requirements

for binocular vision formation are the same visual acuity in both eyes (not less than 0.3—0.4), the same degree of refraction in both eyes, transparency of the optical media, precise coordination of 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.

Disorders of ocular motility can be classified according to the age of onset — *congenital* or *acquired*; according to the underlying etiology — *primary, secondary, recurrent, residual, consecutive*; according to fixation — *unilateral* or *intermittent*; according to the duration of deviation — *constant, intermittent* or *periodic*; according to the manifestation of deviation — *false* and *true*, which in its turn is divided into *latent* and *manifest*; according to the direction of deviation — *horizontal, vertical* or *torsional*; according to the variation with the position of gaze — *concomitant* or *incomitant*.

The main *examination methods* of binocular vision and ocular motility disorders are ocular motility evaluation, cover/uncover test, prism cover test, Hirschberg corneal light reflex test, Kalfa method or two pencils test, Sokolov test (test with "a hole in a palm"), test to read with a pencil (or pen), Belostotskiy-Friedman 4-dot color test, Bagolini striated glass test, synoptophore.

Concomitant strabismus is commonly associated with children. Depending on the direction of eye deviation there can be esotropia, exotropia, hypertropia, hypotropia, cyclotropia. Esotropia according to the association with accommodation is divided into accommodative, partially accommodative or non-accommodative. Clinical presentation of the condition includes deviation of the eye, the angle of deviation is constant regardless of gaze direction, the primary and secondary angles of deviation are equal, full range of eye motion and equal bilaterally, refraction anomalies, amblyopia, or other ocular disorders. Treatment involves optical correction, occlusion of the healthy eye, orthoptic therapy and surgery.

Incomitant strabismus is more often seen in adults and according to the causative factor is divided into paralytic or restrictive. Clinical presentation includes sudden onset of the eye deviation, angle of deviation differs in different directions of gaze, secondary deviation angle is larger than primary one, restriction of ocular movements, compensatory head posture, diplopia, false projection, may be accompanied by ptosis, nausea or dizziness. Treatment depends on the causative factor and includes treatment of the underlying cause, occlusion of the affected eye, optical correction with prisms, injections of botulinum toxin, and surgical correction.

Nystagmus may develop in infancy or adults. According to the etiology it can be optical, neurogenic, or vestibular. Nystagmus is a sign, not a disease. It may be the first sign of serious disorders. Symptoms are involuntary eye movements, trouble focusing and reading, blurry vision, loss of depth perception, holding head in tilted position, dizziness, difficulties in balance and coordination. Treatment helps to reduce symptoms and depends on the cause of the condition. It includes treatment of the underlying cause and related vision problems, botulinum injections, and surgery.

2. Diagrams

STRABISMUS
According to the age of onset
Infantile
Acquired
According to fixation
Monocular
Alternating
According to the manifestation
False
Latent (heterophoria)
True
Manifest (heterotropia)
According to the variation with position of gaze
Incomitant
Concomitant
Paralytic
Restrictive
Accommodative
Partially accommodative
Non-accommodative
According to the direction of deviation
Horizontal
Convergent (eso-)
Divergent (exo-)
Vertical
Supravergent (hyper-)
Infravergent (hypo-)
Torsional (cyclo-)

3. The Review Questions

A. Control Questions

1. What is binocular vision? What are its advantages and requirements for its formation?
2. What examination methods are used to test binocular vision?
3. What is strabismus? Describe its types — false, latent, manifest.
4. What is concomitant strabismus and its types according to eye deviation?
5. What examination methods are used to evaluate the angle of deviation?
6. Dwell on accommodative esotropia and the main methods of its treatment.
7. What methods are used for amblyopia prevention?
8. What is incomitant strabismus and its types?
9. What are differential symptoms of concomitant and incomitant strabismus?
10. What is nystagmus and its types?

B. Tests

1. **Advantages of binocular vision are:**
   A. Increase of VA and expanding of the visual field
   B. Decrease of VA and narrowing of the visual field
   C. Improvement of color vision
   D. Three-dimensional visual perception
   E. Determinate relative position of objects
   F. Determinate distance from one object to another

2. **Diagnostic methods for binocular vision examination are:**
   A. Kalfa method or Two-pencil test
   B. Sokolov test (test with "a hole in a palm")
   C. Cover tests
   D. Hirschberg corneal light reflex test
   E. Belostotskiy-Friedman 4-dot color test
   F. Test to read with a pencil (or pen)

3. **What conditions are related to strabismus?**
   A. Orthophoria
   B. Heterophoria
   C. Heterotropia
   D. Heterochromia
   E. Esotropia
   F. Exotropia

4. **Diagnostic methods for strabismus evaluation are:**
   A. Ocular motility evaluation
   B. Cover/uncover tests
   C. Hirschberg corneal light reflex test
   D. Kalfa method or Two-pencil test
   E. Prism cover test
   F. Synoptophore

5. **Clinical characteristic of concomitant strabismus are:**
   A. The angle of deviation is constant
   B. The angle of deviation varies
   C. Full range of eye motion
   D. Restriction of ocular movements

E. Refraction anomalies
F. Amblyopia

6. Clinical characteristic of incomitant strabismus are:
A. The angle of deviation is constant
B. The angle of deviation varies
C. Full range of eye motion
D. Restriction of ocular movements
E. Diplopia
F. Amblyopia

7. Treatment methods for concomitant strabismus are:
A. Optical correction
B. Occlusion of the healthy eye
C. Occlusion of the deviating eye
D. Orthoptic surgery
E. Surgical treatment
F. Botulinum toxin injections

8. Treatment methods for incomitant strabismus are:
A. Treatment of the underlying cause
B. Optical correction
C. Occlusion of the healthy eye
D. Orthoptic surgery
E. Surgical treatment
F. Botulinum toxin injections

9. Nystagmus is:
A. Inward deviation of the eye
B. Outward deviation of the eye
C. Upward deviation of the eye
D. Downward deviation of the eye
E. Wheel rotation or torsional deviation of the eye
F. Uncontrollable and repetitive movements of the eyes

10. The main causes of nystagmus are:
A. Congenital cataract
B. Head injuries
C. Brain diseases
D. Diseases of the vestibular system
E. Certain medicines
F. Alcohol or drug use

C. Clinical Cases

Case 1

A 4-year-old girl was referred to the ophthalmologist with a history of right eye deviation over the past year. Examination showed that VA of both eyes is equal, the right eye deviates nasally, the angle of deviation according to the Hirschberg corneal light reflex test is 30° and equal in all the directions of gaze. Diplopia is absent, the movement of the eye is full in all directions. The secondary angle of deviation is equal to the primary one. Make a diagnosis.

Case 2

A 3-year-old boy was referred to the doctor with a complaint of crossed eyes. His left eye deviates temporally. The condition developed slowly over the last 2 years. Examination showed that VA is equal in both eyes, the angle of deviation according to the Hirschberg corneal light reflex test is 30—35° and equal in all the directions of gaze. Diplopia is absent, the movement of the eye is full in all directions. The secondary angle of deviation is equal to the primary one. Make a diagnosis.

Case 3

A child was referred at the age of 6 months with a history of crossed eyes noticed over the last 2 months. During examination she is alert and playful, the facial structure without peculiarities, the left eye is crossed inwards. The girl tracks toys well using both eyes. With the left eye covered, she fixes and follows easily. When the right eye is covered, she is fussing (she can't see as well with her left eye). Motility is full, the pupils are round and reactive to light. Hirschberg test shows that reflection is located temporally to the center of the left pupil. Upon covering the right eye, there is an outward shift of the left eye. When the eye is uncovered, the left eye shifts back inwards. Make a diagnosis. What is the first step of treatment?

Case 4

A 3.5-year-old girl was referred to the ophthalmologist with a history of crossed eyes and excessive blinking. The condition has worsened over six months. On examination: visual acuity OD — 0.7, OS — 0.1; prism cover test — constant slight right convergent deviation for near and distance fixation (near 12 prism diopters; distance 8 prism diopters). Ophthalmoscopy was normal and cycloplegic refraction revealed bilateral moderate hypermetropia with significant anisometropia (+5.00 diopter spheres (DS) in the right eye and +3.50 (DS) in the left eye). What is the diagnosis and first step of treatment?

Case 5

A 23-year-old woman referred to the hospital with complaints of alternately inward deviation of one and then another eye. Diplopia is absent. Examination showed that the patient can fix on an object with the right or with the left eye. The angle of deviation according to the Hirschberg corneal light reflex test is 30°. What is the diagnosis?

Case 6

A 21-year-old man presented with a cosmetically unacceptable outward turning of the left eye. There was a history of a penetrating injury to the left eye at the age of 7 years. A traumatic cataract was removed 6 months later and an intraocular lens was implanted. He also had left corneal scarring related to the penetrating injury. VA: OD — 1.0, OS — perception of light. Cover test — moderate angle outward deviation of the left eye with poor left fixation, angle of deviation — 35 prism diopters. Diplopia is absent. What is the diagnosis and recommendations?

Case 7

A 36-year-old woman presented with complaints of the absence of vision and outward deviation of the left eye. 4 years ago she was treated in hospital due to neuritis of the left eye and was discharged with very weak vision. On examination: VA of the left eye — hand movement near the face, ocular motions are not limited, the eye is deviated outward, the angle of deviation according to the Hirschberg corneal light reflex test is 45°. What is the diagnosis?

Case 8

A 30-year-old man presented with an outward deviation of the right eye and diplopia. Two weeks ago during a car accident he injured his head. During the examination: the right eye deviates outward and downward, almost full ptosis, mydriasis, the inward movement of the eye is absent. Diplopia disappears when the right eye is covered. The left eye is healthy. Make a diagnosis.

Case 9

A 60-year-old woman referred to the doctor with complaints of double vision. She has been suffering from hypertension for 10 years. Two days ago she noticed deterioration of general condition (headache, dizziness). After the use of antihypertensives her general condition improved, but diplopia appeared. Examination showed upward and inward deviation of the left eye, limited ocular movement downward and outward. Make a diagnosis, give recommendations.

Case 10

A 72-year-old man presented complains of blurred vision and moving images. There is a history of relapsing and remitting multiple sclerosis. On examination: horizontal jerk movements of the eyeballs, no manifest strabismus. Horizontal gaze-evoked nystagmus was present to right and left gaze with rebound nystagmus on refixation. A rotary element was also seen in the nystagmus.

CHAPTER 20

Blindness

OBJECTIVES

Upon completion of the chapter the students should:

— know the definition of blindness and its types;
— know international classification of visual impairment and blindness;
— know the causes of blindness and visual loss;
— know the rehabilitation methods of the blind;
— be acquainted with global initiatives for visual impairment prevention.

Plan:

1. **DEFINITION OF BLINDNESS AND ITS TYPES**
2. **INTERNATIONAL CLASSIFICATION OF VISUAL IMPAIRMENT**
3. **COMMON CAUSES OF BLINDNESS**
4. **METHODS OF REHABILITATION OF THE BLIND**
5. **GLOBAL INITIATIVES FOR PREVENTION OF BLINDNESS AND VISUAL IMPAIRMENT**

1. Definition of Blindness and Its Types

There are different definitions and terms for blindness but in ophthalmology the term **blindness** strictly implies complete absence of vision or inability to perceive light. However, blindness involves varying levels of vision disability that cannot be fully corrected with glasses or contact lenses and can range from mild visual impairment to total loss of vision.

According to the severity of VA decrease and functionality of vision blindness can be characterized as:

— *absolute or medical blindness* is a complete loss of vision, inability to distinguish between light and darkness. In this state VA is 0;

— *object blindness* is a level of vision when a patient does not distinguish the shape of objects located around him or her but feels light and change of its intensity, and correctly determines the direction of a light source. Visual acuity in these cases is designated as $1/\infty$;

— *civil or social blindness*, in which a patient loses the ability to navigate in the environment, to move out of the house without assistance, thus requiring dependence upon another person, agency, or device, but retains the perception of light. This type of blindness can also be called household blindness;

— *professional blindness* is a condition of visual functions, which makes it impossible to carry out professional work.

Along with complete blindness, which is also called also amaurosis, there is *partial* or *practical blindness* — a partial ability to see, when people distinguish light and dark or have a low visual ability that has no practical significance.

The category of the *almost blind* includes persons with visual acuity from light perception to 0.04 in the better eye with correction tolerated by conventional means. This is called residual vision.

According to the etiology blindness can be *congenital* (inherited, genetic, or result from action of infectious diseases, certain medications or toxins during pregnancy) or *acquired* due to traumas, ocular or systemic diseases.

Blindness can be *preventable* by effective preventive or prophylactic measures (e.g. xerophthalmia, trachoma) and *curable*, where visual impairment is reversible by prompt management (e.g. cataract).

Visual damage can affect not only central vision but other visual functions too or arise after certain conditions. *Color blindness* is inability to distinguish certain colors. *Night blindness* is a difficulty in seeing under decreased illumination. *Snow blindness* is temporary loss of vision after exposure to a large amount of UV light.

2. International Classification of Visual Impairment

In order to determine, which people may need special assistance because of their visual disabilities, in different countries various scales have been developed to describe the extent of vision loss and define blindness. These scales of visual impairment are based on central vision and visual field loss. In Ukraine a patient with a visual acuity of 0.04 or less, or a field of vision of 10 degrees from the point of fixation and less is recognized as blind. In the US and many European countries, any person with a vision that cannot be corrected to better than 20/200 in the best eye, or who has constriction of the visual field in the better eye to less than 20 degrees of normal, is considered legally blind. This means that a legally blind individual would have to stand 20 feet (6 m) from an object to see it with the same degree of clarity as a normally sighted person could from 200 feet (60 m).

By the Tenth Revision of the International Statistical Classification of Diseases, Injuries and Causes of Death, published by the World Health Organization (WHO), ***low vision*** is defined as VA of about 0.05—0.3 or corresponding visual field loss to less than 20 degrees in the better eye with the best possible correction. ***Blindness*** is defined as visual acuity of less than 0.05 or corresponding visual field loss to less than 10 degrees in the better eye with the best possible correction. This 10th Revision divided visual impairment into 6 categories, which are presented in table 20.1. Their use to classify different levels of visual impairment is shown in table 20.2.

Table 20.1

Categories of Visual Impairment Severity According to the 10th Revision of the International Statistical Classification

Category of Visual Impairment	Classified as	Presenting VA and Visual Field	Level of Disability
1	Mild vision loss, or near-normal vision	20/30 to 20/70 (0.5—0.3) or a visual field of less than 30 degrees	No special aids needed; may require reading glasses
2	Moderate visual impairment	20/80 to 20/160 (0.25—0.125), visual field not considered	Special aids needed for some tasks
3	Severe visual impairment (legally blind)*	20/200 to 20/400 (0.1—0.05) or a visual field of less than 20 degrees	Can read only with special aids at reduced speed and endurance

Table 20.1, continued

Category of Visual Impairment	Classified as	Presenting VA and Visual Field	Level of Disability
4	Profound low vision*	20/500 to 20/1,000 (0.04—0.02) or a visual field less than 10 degrees	Reading and mobility impaired; relies on other senses for some tasks
5	Near-total blindness*	Hand motion, light perception, or a visual field of less than 5 degrees	Relies on other senses; cannot benefit from magnification
6	Total blindness*	No light perception	No vision; relies completely on other senses

* Note: Categories 1 and 2 constitute "low vision", and categories 3—6 constitute "blindness".

Table 20.2

Classification of Visual Impairment in a Person According to the International Classification of Diseases (ICD)

H54 ICD Code	Level of Visual Impairment in a Person	Category of Visual Impairment
H54.0	Blindness in a person	Visual impairment category 3, 4, 5 or 6 in the better eye
H54.1	Moderate visual impairment in a person	Visual impairment category 2 in the better eye
H54.2	Mild visual impairment in a person	Visual impairment category 1 in the better eye
H54.3	Unspecified visual impairment in a person	Visual impairment category 9 in both eyes
H54.4	Blindness in one eye of a person	Visual impairment category 3, 4, 5 or 6 in one eye and no visual impairment in the other eye
H54.5	Moderate visual impairment in one eye of a person	Visual impairment category 2 in one eye and no visual impairment in the other eye
H54.6	Mild visual impairment in one eye of a person	Visual impairment category 1 in one eye and no visual impairment in the other eye
H54.7	Unspecified visual impairment in one eye of a person	Visual impairment category 9 in one eye and no visual impairment in the other eye

Visually impaired are persons, who have visual acuity of the better eye with conventional optical correction of about 0.05—0.2. This allows such persons using sight in particularly favorable conditions for learning and performing other activities that do not require high visual acuity. Residual vision promotes the development of some spatial representations, but it is not enough to use it for work that requires constant visual control. Therefore, almost blind persons as well as persons with absolute blindness are educated and trained without the participation of their visual system.

3. Common Causes of Blindness

Among the reasons that most commonly cause blindness the WHO indicates cataract (about 47.9 %), glaucoma (about 12.3 %), diseases associated with aging (about 8.7 %), clouding of the cornea (about 5.1 %), diabetic retinopathy (about 4.8 %), blindness in children (about 3.9 %), and trachoma (about 3.6 %).

.The causes of blindness differ according to the socioeconomic condition of the country. In Ukraine, in 2015 glaucoma ranked first among the pathologies leading to blindness. The second widespread cause is degenerative diseases of the retina, the third — the effects of trauma.

In developed countries, particularly in Western Europe and North America, most cases of blindness are associated with macular degeneration, glaucoma, diabetic retinopathy and traumatic injuries.

In developing countries the principal causes are infections, cataracts, glaucoma, injury, vitamin A deficiency, and uncorrected refractive errors. Infectious causes include trachoma, onchocerciasis (river blindness), and leprosy.

Other common causes of blindness include retinopathy of prematurity, blood vessel diseases involving the retina or optic nerve including stroke, congenital abnormalities, amblyopia, tumors of the eye such as retinoblastoma and optic glioma, complications of eye surgery and chemical poisoning from toxic agents such as methanol.

The statistics of blindness prevalence in the world is shown in fig. 20.1.

Fig. 20.1. The number of the blind in different countries

According to the WHO's data, 90 % of blind people live in developing countries, so 65 % (22 million) cases of blindness in these countries are caused by cataract. It is important that in 70—80 % cases the vision of blind people could be restored after adequate treatment. Blindness from glaucoma in developing countries occurs in about 6 million cases per year, while onchocerciasis causes approximately 1 million cases of blindness worldwide.

Blindness that grows with age, as well as blindness caused by uncontrolled diabetes is more common in the world. On the other hand, health actions reduced the number of cases of blindness that develops due to an infection. So the number of people suffering from trachoma leading to blindness decreased from 360 million as of 1985 to 40 million at the beginning of the 2000s, and over the past 10 years from 6.0 million to 1.3 million cases per year, making it the seventh in the list of causes of blindness worldwide.

4. Methods of Rehabilitation of the Blind

For the patients with low vision rehabilitation is designed to support the physical, psychological and social activities, to help them achieve social independence as well as to develop and improve their quality of life. Adaptation to the disability and psychological help are priority issues and must be confronted from the beginning. Not least important and almost as urgent is education of the patient and their family to confront the new situation. Social integration aids facilitate adapted leisure and cultural activities, and private and public initiatives tending to improve mobility and better access to information for everybody, including the visually impaired. Developed countries, taking care of the blind, create societies of the blind. In Ukraine it is the Ukrainian Society of the Blind. The blind are educated in specialized kindergartens, schools, and orphanages.

Many people with serious visual impairment can travel independently and orient themselves in space with the help of hearing and sense of touch, special devices or guides. For walking these people use a long cane to extend their range of touch sensation, swung in a low sweeping motion across the intended path of travel to detect obstacles. For a few hundred years specially trained dogs have been used as guides to help blind and visually impaired people to move around and avoid obstacles.

People who are not totally blind can read with the help of telescopic glasses, magnifiers and other special means of correction that greatly facilitate seeing of the and enhance their performance. Others read with the help of Braille — a special writing system. Frenchman Louis Braille injured his eyes at the age of 3 years, but thanks to his father, who was strongly engaged in the development of the child, he became a thoroughly educated man. At age 12 he began to work independently on the creation of the alphabet

for the blind. The method consists in using different combinations of six raised dots; each letter of the French alphabet corresponds to its point's location. The alphabet also includes punctuation and numbers. Continuing his studies, Louis adapted the method for musical notation. In 1878 at the World Congress in Rome the Braille method was adopted as the most suitable method of reading and writing for blind people.

There are audio books, computers with special hardware such as scanners and refreshable Braille displays and software written specifically for the blind, like optical character recognition applications and screen reading software as well as special computer games.

To facilitate the life of blind people, there are also a lot of tools, such as relief paving slabs at bus stops, traffic lights with dubbing audio signal, special barriers, etc. In some tourist areas, there are three-dimensional models of the environment on a small scale, which enable the blind to read the surrounding architecture with the help of touch.

People may use talking thermometers, enlarged or marked oven dials, talking watches, talking clocks, talking scales, talking calculators, talking compasses, and other talking equipment. Blind and partially sighted people participate in sports such as swimming, snow skiing, and athletics. Some sports have been invented or adapted for the blind such as cricket and golf. People with vision impairment have participated in the Paraolympic Games since 1976.

Scientists are working on stem cell treatment, gene therapy, the creation of artificial visual systems with implantable chips with video processors that can create opportunities of visibility of the outlines of objects.

The organ of vision is one of the most important sensory organs, providing about 80 % of perception of the surrounding world. Devoid of view people are deprived not only of the opportunity to see the sunrise, but also to live a full life and serve themselves. Therefore, it is necessary to learn ophthalmology to help people and save their vision.

5. Global Initiatives for Prevention of Blindness and Visual Impairment

According to the WHO data, globally about 80 % of all visual impairment can be prevented or cured. And in order to eliminate the main causes of avoidable blindness and to help create sustainable access to eye care for patients around the world the WHO initiated the global program *VISION 2020: The Right to Sight* along with the International Agency for the Prevention of Blindness (IAPB). The project is targeted to attain

the best possible vision for all people thereby improving their quality of life and its main goal is to eliminate avoidable and treatable blindness by 2020. This can be achieved through integration of eye care services into primary and secondary health care systems; campaigns to raise awareness, including school-based education; and stronger international partnerships, with engagement of the private sector and civil society.

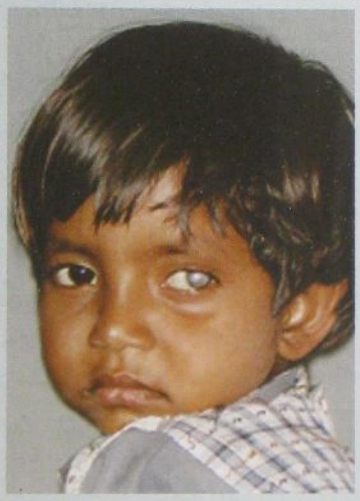
The core strategies of the program are:

— *disease control* — to facilitate the implementation of specific programs to control and treat the major causes of blindness;

— *human resource development* — to support training of ophthalmologists and other eye care personnel to provide eye care;

— *infrastructure and appropriate technology development* — to assist in improving the infrastructure and technology to make eye care more available and accessible.

FACTS about blindness and visual impairment according to estimates from the WHO Prevention of Blindness and Deafness program:



- About 314 million people are estimated to be visually impaired: 45 million are blind and 269 million have low vision.
- About 90 % of the world's visually impaired people live in developing countries.
- About 80 % of all visual impairment can be prevented or cured.
- 153 million people are visually impaired because of uncorrected refractive errors (near-sightedness, far-sightedness or astigmatism). Almost all of them could have normal vision restored with eyeglasses, contact lenses or refractive surgery.
- 39 % of all blindness is due to age-related cataract, the leading cause of blindness in developing countries.
- 65 % of visually impaired, and 82 % of blind people are over 50 years of age.
- Around 1.4 million children under the age of 15 are blind. Yet approximately half of all childhood blindness can be avoided by treating diseases early and by correcting abnormalities such as cataract and glaucoma at birth.
- The number of people visually impaired from infectious diseases has greatly reduced in the last 20 years.

For decades, the project has been working with global partners to eliminate the main causes of avoidable blindness, strengthening country-level efforts by providing technical assistance, monitoring and coordination. As the result, the data over the last 20 years show that there has been a significant progress in the prevention and cure of visual impairment in many countries. Furthermore, there has been a massive reduction in infectious-related blindness as part of a significant reduction in the disease. This has been achieved through a number of successful international partnerships.

Many international non-governmental organizations, professional associations, eye care institutions and medical corporations around the world are involved in eye care and prevention and management of blindness. Among them are Orbis International, SEE International, Mercy Ships, Optometry Giving Sight, SightSavers International, VisionSpring, Volunteer Optometric Services to Humanity and many other humanitarian eye care organizations, which aim is to bring eye care to underserved regions and/or segments of the population with vision challenges.

Review:

1. Key Points

In ophthalmology, the term *blindness* strictly implies complete absence of vision or inability to perceive light. However, blindness involves varying levels of vision disability that cannot be fully corrected with glasses or contact lenses and can range from mild visual impairment to total loss of vision.

According to the functionality of vision blindness can be characterized as *absolute*, *object*, *civil* or *professional*. Blindness can be *congenital* or *acquired*, *preventable* and *curable*. By affection of other visual functions and causes blindness can be *color*, *night*, *snow*.

By the 10th Revision of the International Statistical Classification visual impairment is divided into 6 categories, which are classified as *mild vision loss*, *moderate visual impairment*, *severe visual impairment*, *profound low vision*, *near-total blindness* and *total blindness*.

The most common causes of blindness in developed countries are macular degeneration, glaucoma, diabetic retinopathy and traumatic injuries, in developing countries — infections, cataracts, glaucoma, injury, vitamin A deficiency, and uncorrected refractive errors.

Rehabilitation for the patients with low vision is designed to support the physical, psychological, and social activities, to help them achieve social independence as well as to develop and improve their quality of life. These methods include

low vision aids such as magnifiers, sticks or dogs for moving around, the Braille system, audio books, computers with special hardware, talking equipment, etc.

The WHO initiated the global program VISION 2020: The Right to Sight, that is targeted to attain the best possible vision for all people and its main goal is to eliminate avoidable and treatable blindness by 2020. Many international non-governmental organizations, professional associations, eye care institutions, and medical corporations around the world are involved in eye care, prevention and management of blindness.

2. The Review Questions

A. Control Questions

1. What is blindness?
2. What types of blindness do you know?
3. What are the definitions of low vision and blindness according to the 10th Revision of the International Statistical Classification?
4. What are the categories of visual impairment severity according to the 10th Revision of the International Statistical Classification?
5. How is visual impairment classified according to the International Classification of Diseases (ICD)?
6. What diseases can cause blindness?
7. What methods of rehabilitation of the blind do you know?
8. What is the program VISION 2020 and its main goal?

Appendix 1

OPHTHALMIC PHARMACEUTICALS

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
		ANTIBIOTICS	
Ofloxacin 0.3 %	Floxal®, Ocuflox®, Uniflox®, Ofloxacin®	Ophthalmic solution, 5/10 ml	Days 1 and 2: 1 to 2 drops into the affected eye(s) every 2 to 4 hours; Days 3 through 7—1 to 2 drops into the affected eye(s) 4 times a day. *Corneal ulcers:* Day 1: 1 to 2 drops into the affected eye every 30 minutes while awake; Day 2: 1 to 2 drops into the affected eye hourly while awake; Day 3 through 14: 2 drops 4 hours while awake
Ofloxacin 0.3 %	Floxal®, Ofloxacin®	Ophthalmic ointment, 3 g	1 cm strip applied into the conjunctival sac of the affected eye(s) 2—3 times a day up to 14 days; *Chlamydial eye infections:* 1 cm strip applied into the conjunctival sac of the affected eye(s) 5 times a day up to 14 days
Ciprofloxacin 0.3 %	Ciloxan®, Ciprofarm®, Ciprolet®, Floximed, Cipromed	Ophthalmic/ear solution, 5/10 ml	Days 1 and 2: 1 to 2 drops into the affected eye(s) every 2 hours while awake; Days 3 through 7: 1 to 2 drops into the affected eye(s) 4 times a day. *Corneal ulcers:* Day 1: 2 drops into the affected eye every 15 minutes for the first 6 hours and then 2 drops into the affected eye every 30 minutes for the remainder of the day; Day 2: 2 drops into the affected eye hourly; Day 3 through 14: 2 drops 4 hours while awake

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Ciprofloxacin 0.3 %	Ciloxan®, Floximed	Ophthalmic ointment, 3 g	Days 1 and 2: 1.25 cm strip applied into the conjunctival sac of the affected eye(s) 3 times a day; Days 3 through 7: 1.25 cm strip applied into the conjunctival sac of the affected eye(s) 2 times a day
Levofloxacin 0.5 %	Oftaquix®, Signicef®, Quixin®, Levoximed	Ophthalmic solution, 5 ml	Days 1 and 2: 1 to 2 drops into the affected eye(s) every 2 hours while awake (8 times a day); Days 3 through 5: 1 to 2 drops into the affected eye(s) 4 times a day
Lomefloxacin 0.3 %	Okacin®	Ophthalmic solution, 5 ml	Day 1: 5 drops into the conjunctival sac of the affected eye(s) within 20 minutes (1 drop in 5 min) or 1 drop hourly for 6—10 hours. Thereafter, until Day 7—9: 1 drop 3 times daily into the conjunctival sac of the affected eye(s)
Moxifloxacin 0.5 %	Vigamox®	Ophthalmic solution, 3 ml	1 drop in the affected eye(s) 3 times a day for 7 days
Norfloxacin 0.3 %	Norfloxacin®	Ophthalmic/ear solution, 5/10 ml	1 or 2 drops to the affected eye(s) 4—6 times daily for up to 7 days. *For severe infections:* 1 or 2 drops to the affected eye(s) every 2 hours while awake on the first day. *For trachoma:* 2 drops to each eye 2—4 times daily for up to 1—2 month(s)
Norfloxacin 0.3 %	Norfloxacin®	Ophthalmic ointment, 5 g	1 cm strip applied into the conjunctival sac of the affected eye(s) 3 times a day on the first two days, and then 2 times a day for the next five days
Gatifloxacin 0.5 %	Zymaxid™	Ophthalmic solution, 2.5 ml	Day 1: 1 drop every two hours in the affected eye(s) while awake, up to 8 times. Day 2 through Day 7: 1 drop 2—4 times daily in the affected eye(s)

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Tobramycin 0.3 %	Tobrex®, Tobramycin, Tobrin®	Ophthalmic solution, 5 ml	1 or 2 drops into the affected eye(s) every four hours for 7—10 days. *For severe infections:* 2 drops to the affected eye(s) hourly until improvement and then by the above scheme
Tobramycin 0.3 %	Tobrex®	Ophthalmic ointment, 3.5 g	1.25 cm strip applied into the conjunctival sac of the affected eye(s) two to four times a day for 7—10 days.
Chloramphenicol 0.25 %	Levomycetin	Ophthalmic solution, 5/10 ml	1 drop into the affected eye(s) every 2 hours for the first 48 hours, then three to four times a day for 5—14 days
Gentamicin 0.3 %	Gentamicin	Ophthalmic solution, 5 ml	1 or 2 drops into the affected eye(s) 3—4 times daily for 7—14 days
Gentamicin 0.3 %	Gentamicin	Ophthalmic ointment, 3.5 g	1.25 cm strip applied into the conjunctival sac of the affected eye(s) two or three times a day for 7—10 days
Tetracycline 1 %	Tetracycline	Ophthalmic ointment, 3 g	1.25 cm strip applied into the conjunctival sac of the affected eye(s) three or five times a day for 7—10 days. *For trachoma:* 1.25 cm strip applied into the conjunctival sac of the affected eye(s) three or five times a day for up to 1—2 month(s)
Erythromycin 0.5 %	Erythromycin	Ophthalmic ointment, 3.5 g	1cm strip applied into the conjunctival sac of the affected eye(s) up to 6 times daily for 5—10 days. For *prophylaxis of neonatal gonococcal or chlamydial conjunctivitis*: 1cm strip of ointment into each lower conjunctival sac
Fusidic acid 1 %	Futaron	Ophthalmic suspension, 5 ml	1 drop into the conjunctival sac of both eyes every 12 hours (i.e. twice daily) for 7 days
TOPICAL STEROIDS			
Hydrocortisone 0.5 %; 1 %; 2.5 %	Hydrocortisone, Hydrocortison-POS®	Ophthalmic ointment, 2.5/3 g	1cm strip applied into the conjunctival sac of the affected eye(s) up to 2—3 times daily for not longer than 14 days

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Dexamethasone 0.1 %	Dexamethasone, Pharmadex, Maxidex®, Dexapos COMOD, Medexol, Oftan® Dexamethason	Ophthalmic solution/ suspension, 5/10 ml	1 or 2 drops into the affected eye(s) every hour during the day and every two hours during the night as initial therapy until improvement is observed Then the dosage is reduced to 1 drop every four hours. Later, further reduction in dosage to one drop three or four times daily
Dexamethasone	Ozurdex®	Intravitreal implant, 0.7 mg	For intravitreal injection in cases of macular edema following retinal vein occlusion, posterior uveitis, diabetic macular edema
Prednisolone acetate 1 %	Pred Forte®	Ophthalmic suspension, 5/10 ml	1 to 2 drops topically into the conjunctival sac of the affected eye(s) two to four times daily. During the initial 24 to 48 hours, the dosing frequency may be increased if necessary. (!) If signs and symptoms fail to improve after 2 days, the patient should be re-evaluated
Cefuroxime	Aprokam®	Powder for solution for injection, 50 mg	Intracameral use For *prophylaxis of postoperative endophthalmitis after cataract surgery*
		TOPICAL NSAIDS	
Diclofenac 0.1 %	Uniclophen®, Diphthal®, Diclo-F®, Naclof, Clodifen	Ophthalmic solution, 5/10 ml	For *prevention of per-operative miosis during cataract surgery:* 1 drop every 30 minutes within 2 hours preceding the operation. For *prevention of complications:* *at cataract surgery* — 1 drop into the affected eye 4 times daily beginning 24 hours after cataract surgery and continuing for 2 weeks; *at corneal refractive surgery* — 1—2 drops into affected eye within the hour prior to surgery, within 15 minutes after surgery, and then continue for 4 times daily up to 3 days.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
			For *treatment of pain, discomfort, and post-traumatic inflammation:* 1 drop every 4—6 hours for 2 days. When pain is due to a surgical procedure (e.g. refractive surgery), 1 to 2 drops in the hour preceding surgery, 1 to 2 drops within the first 15 minutes after intervention and 1 drop every 4—6 hours for 3 days thereafter. For *reducing of signs and symptoms of seasonal allergic conjunctivitis:* 1 drop into the conjunctival sac of the eyes 4 times daily
Indometacine 0.1 %	Indocollyre®	Ophthalmic solution, 5 ml	For *inhibition of miosis during surgery:* 4 drops the day before surgery and 4 drops in the 3 hours preceding surgery. For *prevention of inflammations related to cataract and to anterior segment of the eye surgery:* 1 drop 4 to 6 times daily 24 hours before operation, 1 drop every 30 minutes within 2 hours preceding the operation, and 1 drop 4 times per day for one month. For *treatment of ocular pain following photorefractive keratectomy:* 1 drop 4 times daily for the first few days after surgery
Nepafenac 0.1 %	Nevanac®	Ophthalmic suspension, 3/4 ml	For *prevention of inflammations and pain related to cataract surgery:* 1 drop into the affected eye 3 times daily beginning 1 day prior to surgery, continued on the day of surgery and through the first 2 weeks of the postoperative period. For *reduction of the risk of postoperative macular edema associated with cataract surgery in diabetic patients:* 1 drop into the affected eye 3 times daily beginning 1 day prior to cataract surgery, continued on the day of surgery and up to 60 days of the postoperative period

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Ketorolac 0.45 %	Acuvail®	Ophthalmic solution, 0.4 ml	For *prevention of inflammations and pain related to cataract surgery:* 1 drop into the affected eye 2 times daily beginning 1 day prior to surgery, continued on the day of surgery and through the first 2 weeks of the postoperative period
Ketorolac 0.5 %	Medrolgin	Ophthalmic solution, 5 ml	For *prevention of inflammations and pain related to cataract surgery:* 1 drop into the affected eye 2 times daily beginning 1 day prior to surgery, continued on the day of surgery and through the first 2 weeks of the postoperative period
ANTIBACTERIAL COMBINED DRUGS			
Tobramycin 0.3 % + Dexamethasone 0.1 %	TobraDex® Dex-Tobrin®, Medetrom, Tobrosodex	Ophthalmic solution/ suspension, 2,5/5/10 ml	1 or 2 drops into the conjunctival sac(s) every four to six hours. *In severe cases:* Initial dosage is 1 or 2 drops every two hours for 24 to 48 hours. Then frequency should be decreased gradually as warranted by improvement in clinical signs.
Tobramycin 0.3 % + Dexamethasone 0.1 %	TobraDex®	Ophthalmic ointment, 3.5 g	1 cm strip into the conjunctival sac(s) up to three or four times daily.
Dexamethasone 0.1 % + Gentamicin 0.5 %	Dexa-Gentamicin®	Ophthalmic solution, 5 ml	1 to 2 drops into the conjunctival sac(s) up to 4—6 times daily at regular intervals. Treatment should not extend a period of 2 weeks, or maximally 3 weeks.
Dexamethasone 0.1 % + Neomycin Sulfate 3.5 mg + Polymyxin B Sulfate 10.000 units	Maxitrol®, Neladex	Ophthalmic/ear solution/ suspension, 5 ml	1 to 2 drops into the conjunctival sac of the affected eye(s) up to 4—6 times daily. *In severe cases:* Initial dosage is 1 or 2 drops every hour for 24 hours. Then frequency should be decreased gradually as warranted by improvement in clinical signs.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Dexamethasone 0.1 % + Neomycin Sulfate 3.5 mg + Polymyxin B Sulfate 10.000 units	Maxitrol®, Neladex	Ophthalmic ointment, 3.5/5 g	1 cm strip into the conjunctival sac(s) up to 3 or 4 times daily or, may be used adjunctively with drops at bedtime.
Hydrocortisone 1 % + Neomycin Sulfate 3.5 mg + Polymyxin B Sulfate 10.000 units	Cortisporin®	Ophthalmic ointment, 15 g	1 cm strip into the conjunctival sac(s) up to 3 or 4 times daily, depending on the severity of the condition.
Framycetin 0.5 % + Gramicidin C 0.005 % + Dexamethasone 0.05 %	Sofradex®	Ophthalmic solution, 5 ml	1—2 drops into the conjunctival sac of the affected eye(s) up to 6 times daily. *In severe cases:* 1—2 drops every one or two hours for 2—3 days, then reducing to 1—2 drops three or four times daily.
ANTIVIRAL AGENTS			
Interferon alfa-2b 200 000 IU	Ocoferon®	Lyophilized powder for ophthalmic solution preparation and solvent, 5 ml	Before administration the powder is dissolved in 5 ml of solvent (0.1 % nipagin solution). 2 drops into the conjunctival sac of the affected eye(s) every 2 hours, but not less than 6 times per day. With reduction of symptoms the volume of instillation can be reduced up to 1 drop. The course of treatment is 7—10 days.
Aciclovir 3 %	Aciclovir® Zovirax®, Virolex	Ophthalmic ointment, 4.5 g	1 cm of strip applied into the lower conjunctival sac 5 times a day at approximately four hourly intervals, omitting the night time application. Treatment should continue for at least 3 days after healing is complete.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Ganciclovir 0.15 %	Virgan®	Ophthalmic gel, 5 g	1 drop into the inferior conjunctival sac of the affected eye(s) 5 times a day until complete corneal re-epithelialisation. Then 3 instillations a day for 7 days after healing. The treatment should not exceed 21 days.
Trifluridine 1 %	Viroptic®	Ophthalmic solution, 7.5 ml	1 drop into the inferior conjunctival sac of the affected eye(s) every 2 hours while awake for a maximum daily dosage of nine drops until complete corneal re-epithelialisation. Then 1 drop every 4 hours while awake for a minimum daily dosage of five drops for 7 days. The treatment should not exceed 21 days.
Idoxuridine 0.1 %	Herplex®	Ophthalmic solution, 10 ml	1 drop into the inferior conjunctival sac of the affected eye(s) every hour during the day and 2 hours at night until complete corneal re-epithelialisation. Then 1 drop every 4 hours while awake for an additional 5—7 days. Maximal treatment period is 21 days.
Idoxuridine 0.5 %	Herplex®	Ophthalmic ointment, 4.5 g	1 cm strip applied into the lower conjunctival sac every four hours (five times a day) during the day and once before bedtime until complete corneal re-epithelialisation.
		ANTIFUNGAL AGENTS	
Natamycin 5 %	Natacyn®	Ophthalmic suspension, 15 ml	1 drop into the conjunctival sac of the affected eye(s) every 1 or 2 hours for three or four days, then one drop six to eight times a day for 14 to 21 days or until there is resolution.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
		DISINFECTANTS	
Sulfacetamide 20 %, 30 %	Sulfacyl, Sulfacyl-Sodium	Ophthalmic solution, 5/10 ml	1—2 drops into the conjunctival sac of the affected eye(s) up to 5—6 times a day for 7—10 days. For *prophylaxis of neonatal gonococcal and chlamydial conjunctivitis*: 2 drops in each eye immediately after birth and after 2 hours (20 %).
Myramistin 0.01 %	Okomistin® Oftamirin®	Ophthalmic/ear solution, 5/10 ml	1 to 2 drops into the conjunctival sac of the affected eye(s) up to 4—6 times daily for 7—14 days. For *prophylaxis of neonatal gonococcal and chlamydial conjunctivitis*: 1 drop in each eye right immediately birth 3 times with an interval of 2—3 min. For *prevention of surgical infections:* 2 drops 3 times daily for 2—3 days prior to surgery and for 10—15 days after the intervention. At *treatment of ocular thermal burns*: after irrigation of the eye with water — 2 drops every 5—10 min for 1—2 hours, then 2 drops 4—6 times a day.
Decamethoxin 0.02 %	Oftadek®, Oftalmodek, OkoDek	Ophthalmic solution, 5 ml	1 to 2 drops into the conjunctival sac of the affected eye(s) up to 4—6 times daily for 7—14 days. For *prophylaxis of neonatal gonococcal and chlamydial conjunctivitis*: 2 drops in both eyes immediately after birth and 2 drops in 2 hours. For *prevention of surgical infections:* 2 drops up to 6 times daily on the day before surgery and for 3—5 days after the intervention.
Zinc Sulfate 0.25 % + Decamethoxin 0.02 %	Cidelon	Ophthalmic solution, 5/10 ml	1 to 2 drops into the conjunctival sac of the affected eye(s) 3 times daily.
Zinc Sulfate 0.25 % + Boric Acid 2 %	Ziborat-Ophtan	Ophthalmic solution, 10 ml	2 drops into both eye(s) 2—3 times daily up to 1—3 weeks.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
		ANESTHETICS	
Proxymetacain 0.5 %	Alcaine®	Ophthalmic solution, 15 ml	For rapid and short-acting local anesthesia of the eye surface as: *for cataract extraction*: 1 drop every 5 to 10 minutes for 3 to 5 doses. *for removal of sutures:* 1 or 2 drops 2 or 3 minutes before removal of stitches. *for removal of corneal foreign bodies:* 1 or 2 drops prior to operating; *for tonometry and gonioscopy performing:* 1 or 2 drops immediately before measurement.
Oxybuprocaine 0.4 %	Benox®, Inocain®	Ophthalmic solution, 5 ml	For rapid and short-acting local anesthesia of the eye surface as: *for tonometry and gonioscopy performing:* 1 or 2 drops immediately before measurement; *for removal of conjunctival and corneal foreign bodies:* 3-multiple instillations with the time interval of 4—5 minutes; *for incision of a meibomian cyst through the conjunctiva*: 3-multiple instillations at 90 second intervals before the surgery.
Tetracaine 0.5 % — 1.0 %	Tetracaine Hydrochloride	Ophthalmic solution, 5 ml	For rapid and short-acting local anesthesia of the eye surface as: *for tonometry and other diagnostic purposes:* 1 or 2 drops just prior to evaluation; *for superficial foreign bodies or suture removal:* 1 or 2 drops in every five minutes for one to three doses; *for cataract extraction*: 1 drop every 5 to 10 minutes for up to 5—7 doses; *for laser microsurgery:* 1 drop 3 times every 2 minutes prior to operating.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
MYDRIATICS AND CYCLOPLEGICS			
Atropine 1 %	Atropine sulfate	Ophthalmic solution, 5 ml	For *diagnostic purposes:* 1 or 2 drops into the conjunctival sac 40—60 minutes prior to evaluation; for *pre-operative mydriasis:* 1—2 drops into the conjunctival sac 30—60 minutes prior to operating; for *iridocyclitis, uveitis, keratitis, ocular trauma, reducing ocular pain, retinal artery spasm and the tendency to thrombosis:* 1 drop into the affected eye three times daily (with intervals of 5—6 hours).
Cyclopentolat 1 %	Cyclomed®	Ophthalmic solution, 5 ml	For *ophthalmoscopy:* 3-multiple instillations by 1 drop at 10 minute intervals 30 minutes prior to evaluation; for *refraction examination:* 1 or 2 drops into the conjunctival sac 2—3 times with the interval of 15—20 minutes prior to examination; for *anterior and posterior uveitis and for the breakdown of posterior synechiae:* 1 to 2 drops 3 times per day, in the heavy cases 1 drop each 3—4 hours is permitted.
Tropicamide 1 %	Mydriacyl®, Tropicamide, Unitropic®	Ophthalmic solution, 5/10 ml	For *ophthalmoscopy and refraction examination:* 1 or 2 drops into the eyes repeated in 5 minutes 20 minutes prior to examination.
Scopolamine 0.25 %	Scopolamine	Ophthalmic solution, 5 ml	For *refraction examination:* 1 or 2 drops into the conjunctival one hour prior to evaluation; for *iridocyclitis treatment:* 1 or 2 drops into the conjunctival sac up to 4 times daily; for *ocular pain control:* 1 drop into the affected eye three times daily (with intervals of 5—6 hours).

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Phenylephrine hydrochloride 2.5 % or 10 %	Phenylephrine®, Irifrin®, Mezaton, Phenephrin	Ophthalmic solution, 5 ml	For *ophthalmoscopy (2.5 %):* 1 or 2 drops into the conjunctival sac 40 minutes prior to evaluation; for *performing a provocative test for suspected angle-closure glaucoma and differential diagnosis of conjunctival injection (2.5 %):* 1 drop into the affected eye as a single dose; for *iridocyclitis treatment (2.5 % or 10%):* 1 drop into the conjunctival sac 2—3 times daily (8 hourly) up to 10 days; for *pre-operative mydriasis (2.5 %):* 1—2 drops into the conjunctival sac 30—60 minutes prior to operating; for *combined therapy of spasm of accommodation and asthenopia in schoolchildren (2.5 %):* 1 drop into the conjunctival sac at bedtime every day for 2—4 weeks.
RE-EPITHELIZATION DRUGS AND ARTIFICIAL TEARS			
Dexpanthenol 5 %	Cornere-gel®	Ophthalmic gel, 5 g	1 one drop into the conjunctival sac 3—5 times per day and 15 minutes before going to bed until complete disappearance of symptoms.
Sodium Hyaluronate 0.1 %	Hylo-Comod®	Ophthalmic solution, 10 ml	1 one drop into the conjunctival sac 3 times daily.
Dexpanthenol 2 % + Sodium Hyaluronate 0.1 %	Hylo-Care®	Ophthalmic solution, 10 ml	1 one drop into the conjunctival sac 3—4 times daily, but it can be used more often.
Dexpanthenol 3 % + Polyvinyl Alcohol 0.14 %	Siccaprotect®	Ophthalmic solution, 10 ml	1 one drop into the conjunctival sac up to 6 times daily, but it can be used every hour in severe cases.
Solcoseryl 20 %	Solcoseryl®	Ophthalmic gel, 5 g	1 one drop into the conjunctival sac 3—4 times per day until complete disappearance of symptoms.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Carbomer 0.2 %	Vidisic®, Sikapos	Ophthalmic gel, 10 g	1 to 2 drops into the conjunctival sac 3—5 times per day and 30 min before going to bed.
Carbomer 0.25 %	Oftagel®, Oftagel® Uno	Ophthalmic gel, 10 g	1 one drop into the conjunctival sac 4 times per day.
Hypromellose 0.32 %	Artelac®	Ophthalmic solution, 10 ml	1 one drop into the conjunctival sac 3—5 times per day.
Hypromellose 0.3 % + Dextran 70 0.1 %	Artificial tears, Tears Naturale	Ophthalmic solution, 5/10/15 ml	1 or 2 drops into the affected eye(s) as needed, up to once every 1—2 hours.
Povidon 2 %	Wet-Comod	Ophthalmic solution, 10 ml	1 drop into the conjunctival sac of both eyes up to 4—5 times daily.
Povidon 0.6 % + Polyvinyl alcohol 1.4 %	Ophtolique®	Ophthalmic solution, 10 ml	1 or 2 drops into the conjunctival sac of both eyes 3—4 times daily.
Carmellose 0.5 % + Glycerol 0.9 %	Optive ®	Ophthalmic solution, 10/15 ml	1 or 2 drops into the affected eye(s) 1—2 times daily or as needed.
Polyethylene Glycol 0.4 %, Propylene Glycol 0.3 %	Systane®, Systane® Ultra	Ophthalmic solution, 10/15 ml	1 or 2 drops into the affected eye(s) 1—3 times daily.
Cyclosporine 0.05 %	Cyclosporine, Restasis®	Ophthalmic emulsion, 0.4 ml	1 drop into each eye twice daily (approximately 12 hours apart) around the same times every day.
Trehalose (3 %) + Hyaluronic acid (0.15 %)	Thealoz Duo	Ophthalmic solution, 10 ml	1 drop into the conjunctival sac 4—6 times daily. Drops are also suitable for contacts lens wearers to apply either before or during wear.
ANTIHISTAMINES			
Cromoglicate 2 %	Lecrolyn®, Cromohexal®, Cromopharm®, Allergocrom, Ifiral®	Ophthalmic solution, 10 ml	1 or 2 drops into each eye up to four times daily at regular intervals. If possible, start treatment before the usual allergy season.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Olopatadine 0.1 %	Opatanol®, Patanol®, Opatadin, Pallada	Ophthalmic solution, 5 ml	1 to 2 drops into the conjunctival sac of the affected eye(s) twice daily (8 hourly). Treatment may be maintained for up to four months, if considered necessary.
Lodoxamide 0.1 %	Alomide®	Ophthalmic solution, 5 ml	1 or 2 drops in each affected eye 4 times a day at regular intervals for up to 3 months.
Emedastine 0.05 %	Emadine®	Ophthalmic solution, 5 ml	1 or 2 drops in the affected eye two times daily.
Azelastine 0.05 %	Allergodil®	Ophthalmic solution, 6 ml	1 or 2 drops in the affected eye two times daily, if necessary up to 4 times a day for 6 weeks.
Ketotifen 0.025 %	Ketotifen	Ophthalmic solution, 5 ml	1 drop in the affected eye(s) 2 times a day (every 8 to 12 hours).
MEDICATIONS USED IN CATARACT			
Pirenoxine 0.005 %	Catalin®	Ophthalmic solution, 15 ml	1 to 2 drops into the conjunctival sac 3 to 5 times every day.
Cytochrom C 0,675 mg + Adenosin 2 mg + Nicotinamid 20 mg	Oftan® Catachrom	Ophthalmic solution, 10 ml	1 to 2 drops into the conjunctival sac 3 times daily for 10—15 days then one week off and then repeat the course.
ANTIGLAUCOMA AGENTS			
Drugs that improve aqueous humor outflow			
Cholinomimetics			
Pilocarpine 1 %	Pilocarpine	Ophthalmic solution, 15 ml	For *reduction of IOP in open angle glaucoma or ocular hypertension:* 1 or 2 drops into the affected eye(s) up to three or four times daily; for *management of acute angle-closure glaucoma:* 1 drop into the affected eye(s) every 15 minutes for 1 hour; then 1 drop every 30 minutes for 2—3 hours; after this 1 drop in an hour during 4th—6th hour, thereafter 1 drop 3—6 times daily until arresting the attack; for *prevention of postoperative elevated IOP associated with laser surgery:* 1 drop into the affected eye(s) 15 to 60 minutes prior to surgery; for *induction of miosis:* 1 drop or 2 drops five minutes apart.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
		Postaglandin analog	
Latanoprost 0.005 %	Xalatan®, Lanotan™, Vizipres®, Glaumax®, Xaloptic, Latanox®, Latanoprost, Latasopt, Prolatan®, Unilat®	Ophthalmic solution, 2.5 ml	1 drop into the affected eye(s) once daily in the evening.
Travoprost 0.004 %	Travatan®, Glautan	Ophthalmic solution, 2.5 ml	1 drop into the affected eye(s) once daily in the evening.
Tafluprost 0.0015 %	Taflotan®, Zioptan®	Ophthalmic solution, 0.3/2.5 ml	1 drop into the conjunctival sac of the affected eye(s) once daily in the evening.
Bimatoprost 0.03 %	Lumigan®, Bimican®	Ophthalmic solution, 5 ml	1 drop into the affected eye(s) once daily in the evening.
		Drugs that suppress aqueous humor production	
		β-blockers (selective and non-selective)	
Betaxolol 0.25—0.5 % (selective)	Betoptic® S, Betalmic	Ophthalmic suspenson, 2.5/5/10/15 ml	1 drop inti the affected eye(s) twice daily.
Timolol 0.25—0.5 % (non-selective)	Oftimol®, Timolol Maleate, Aritumol®, Glaumol, Kuzimolol®, Oftan® Timolol, Normatin	Ophthalmic solution, 5 ml	1 drop inti the affected eye(s) twice a day.
		Carbonic anhydrase inhibitors	
Dorzolamide 2 %	Trusopt®, Rezlod, Dorzamed, Dorzol®, Dorzoptic	Ophthalmic solution, 5/10 ml	1 drop into the affected eye(s) 3 times daily.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Brinzolamide 1 %	Azopt®, Brizal®	Ophthalmic solution, 10 ml	1 drop into the affected eye(s) 2—3 times daily.
		α-adrenergic agonists	
Brimonidine 0.2 %	Alphagan®, Luxfen®, Brimonal	Ophthalmic solution, 2.5/5/10 ml	1 drop into the affected eye(s) twice daily, approximately 12 hours apart.
Apraclonidine 1 %	Iopidine®	Ophthalmic solution, 5/10 ml	1 to 2 drops into the affected eye(s) three times daily.
		Combined drugs	
Latanoprost + Timolol	Lanotan®, Xalacom®, Xaloptic®, Latamed	Ophthalmic solution, 2.5 ml	1 drop into the affected eye(s) once daily in the evening.
Pilocarpine + Timolol	Fotil®, Fotil® forte	Ophthalmic solution, 5 ml	1 drop into the affected eye(s) two times daily.
Travoprost + Timolol	DuoTrav®	Ophthalmic solution, 2.5 ml	1 drop into the affected eye(s) once daily, in the morning or evening. It should be administered at the same time each day.
Brinzolamide + Timolol	Azarga®	Ophthalmic suspension, 5 ml	1 drop into the affected eye(s) two times daily.
Dorzolamide + Timolol	Cosopt®, Mardozia, Dorzotimol®, Dorzasopt	Ophthalmic solution, 5 ml	1 drop into the affected eye(s) two times daily.
Bimatoprost + Timolol	Ganfort®	Ophthalmic solution, 3 ml	1 drop into the affected eye(s) once daily, administered either in the morning or in the evening. It should be administered at the same time each day.
Brimonidine + Timolol	Combigan®	Ophthalmic solution, 5 ml	1 drop into the affected eye(s) twice daily approximately 12 hours apart.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
ANTI-VASCULAR ENDOTHELIAL GROWTH FACTOR (VEGF) AGENTS			
Ranibizumab 6/10 mg/ml	Lucentis®	Solution for injections, 0.05 ml in a single-use pre-filled syringe	For *wet age-related macular degeneration, macular edema following retinal vein occlusion, and diabetic macular edema, diabetic retinopathy:* intravitreal injections once a month (approximately 28 days) for up to three months.
Aflibercept 40 mg\ml	Eylea®	Solution for injections, 3 ml, single-use glass vial	For *wet age-related macular degeneration, macular edema following retinal vein occlusion, and diabetic macular edema, diabetic retinopathy, myopic choroidal neovascularization:* intravitreal injections of 0.05 ml (2 mg) once every 4 weeks (monthly) for the first 12 weeks (3 months), followed by intravitreal injection once every 8 weeks (2 months).
Pegaptanib	Macugen®	Solution for injections, 0.3 mg in a single-use pre-filled glass syringe	For *wet age-related macular degeneration:* intravitreal injections once every six weeks.
OTHER DRUGS			
Potassium iodide 2/3 %	Potassium iodide	Ophthalmic solution, 5/10 ml	For *cataract, corneal and vitreous opacities, subconjunctival hemorrhages, degenerative diseases of the retina, fungal conjunctivitis and keratitis:* 1 or 2 drops into the affected eye(s) up to 4 times daily for 14 days.
Taurin 4 %	Taufon®	Ophthalmic solution, 5 ml	For *cataract, corneal degenerative pathologies and injuries, open-angle glaucoma, degenerative diseases of the retina:* 1 or 2 drops into the affected eye(s) 2—4 times daily for a month or up to three months.

Active Ingredient(s)	Trade Name	Pharmaceutical Form(s)	Administration and Dosage
Bibrocathol 2%	Posiform-in®	Ophthalmic ointment, 5 g	For *irritations of the exterior of the eye and chronic blepharitis not caused by pathogens, uninfected fresh injuries of the cornea:* 0.5 cm strip applied into the conjunctival sac of the affected eye(s) 3—5 times a day up to 14 days.
Emoxipin 1 %	Emoxil-N, M-Oxi-plex®	Solution for injections, 1/3 ml	For *intraocular hemorrhages, thrombosis of the central retinal vein and its branches, diabetic retinopathy, central chorioretinal dystrophies, uveitis, complicated myopia and glaucoma:* subconjunctival, parabulbar or, if necessary, retrobulbar injection.
Prourokinase 5000 IU	Gemasa	Lyophilized powder for ophthalmic solution preparation	For *hyphema, hemophthalm, subretinal hemorrhages, occlusion of the central retinal artery and its branches, thrombosis of central retinal vein and its branches, prevention of adhesions after glaucoma surgery:* subconjunctival or parabulbar injections.
Poloxamer 188 + PEG 90 + Carbomer etc.	Blephagel	Ophthalmic gel, 30 g	For *daily hygiene of eyelids and eyelashes with blepharitis or dry eye, and is suitable for contact lens wearers.* 1 drop to the skin around the eye, the eyelid and eyelid margins twice a day, in the morning and before going to sleep.
Poloxamer 184 Propylene glycol + PEG-6,8 Zinc sulfate Sodium + Hyaluronate etc.	Blephaclean	Sterile pads	For *daily hygiene of eyelids and eyelashes with blepharitis, dry eye or sensitive skin, it also is suitable for contact lens wearers.* Twice a day, in the morning and evening.

Appendix 2

KEYS TO TESTS AND CLINICAL CASES

Chapter 1 — Anatomy of the Eye and Visual Functions

Tests

1. A, B, C, E, F; **2.** B, C, E, F; **3.** B—F; **4.** D, C, A, E, B; **5.** A, D, F; **6.** A, B, D; **7.** A, C, D; **8.** A, F; **9.** B, D, F; **10.** B; **11.** A, E, F; **12.** C; **13.** A, B, D; **14.** A, F; **15.** A, C, E, F; **16.** D, B, F, G, C, E, A; **17.** A, B, D, F.

Chapter 2 — The Ophthalmic Examination

Tests

1. C; **2.** C; **3.** B; **4.** D; **5.** C; **6.** F; **7.** B; **8.** B, D, E; **9.** A, C, D, F; **10.** B; **11.** A—E; **12.** D; **13.** B, D, E; **14.** B, E; **15.** A, D, F; **16.** D; **17.** C; **18.** B, D, F; **19.** B-E; **20.** B; **21.** B, C, E, F; **22.** E; **23.** A, D; **24.** A, F.

Chapter 3 — Optical System of the Eye. Refractive Errors, Accommodation and Amblyopia

Tests

1. C; **2.** B; **3.** D; **4.** A; **5.** D; **6.** A; **7.** B; **8.** D; **9.** C; **10.** C.

Clinical Cases

Case 1 — The combination of decreased distance vision with preserved near vision is typical of myopia, which often becomes symptomatic during adolescence. Presumptive evidence of refractive error is provided by the marked improvement in visual acuity that occurs with the use of pinhole. Note that visual acuity with pinhole frequently does not reach 20/20. The patient should be referred to an ophthalmologist as a regular rather than an urgent consultation;

Case 2 — Cataract is a common cause of painless progressive loss of vision in older individuals. Her complaints about her visual ability are an indication for referral to an ophthalmologist for evaluation for possible cataract surgery;

Case 3 — Referral is not indicated since the cause of decreased vision is established and progressive loss is not occurring. Note that this healthy individual has better than 20/20 acuity in his right eye;

Case 4 — The patient has an unexplained loss of vision of unknown duration in one eye. An unexplained decrease in vision in one or both eyes requires referral to an ophthalmologist, because it may indicate occult disease of the eyes or central nervous system that is not detectable by examination methods available to the primary care physician. In this case, the patient's decreased vision was due to a macular disturbance detectable only by more precise methods of examination (e.g., special lenses and fluorescein angiography);

Case 5 — Visual acuity testing should be attempted using the tumbling E chart or a picture card, with each eye alternately covered by an adhesive patch. A difference in visual acuity

between the eyes or decreased vision in both eyes is significant. Test the alignment of the eyes by evaluating the corneal light reflex. Then proceed to the cover test. Unequal positioning of the light reflex or movement of the uncovered eye to pick up the fixation would suggest a misalignment of the eyes. Perform an ophthalmoscopic examination, preferably through dilated pupils, to determine if there is any intraocular basis for visual loss, such as cataract, retinoblastoma, or a retinal abnormality. If visual acuity is asymmetric or if there is a suspicion of intraocular disease, the patient should be referred for an urgent ophthalmologic evaluation. If visual acuity and fundus examination are normal but strabismus is suspected because of other examination findings or patient history, a nonurgent referral should be made;

Case 6 — Inquire about any family history of strabismus or amblyopia and evaluate for the presence of epicanthus. Place your hand in front of one eye and then the other to see if the child exhibits displeasure. Observe the alignment of the child's eyes as well as extraocular movements if possible. Use a penlight to assess the position of the corneal light reflex. Examination reveals that significant lid folds are present. The corneal light reflex is the same in each eye, and full extraocular movements are seen in all cardinal fields of gaze. Although in this case the appearance of a crossed eye is probably the result of epicanthus, continued observation on the next visit is indicated. Remember that strabismus and amblyopia can occur in a patient with epicanthus, and strabismus may be intermittent. Any suspected abnormality should be referred to an ophthalmologist;

Case 7 — Show the child a toy and cover his left eye with your hand; evaluate his response. Cover his right eye to compare his response. Evaluate the corneal light reflex and perform the cover test. In particular, note whether an abnormal response is elicited on covering one eye. Test the pupillary light reflexes. Perform an ophthalmoscopic examination, preferably through a dilated pupil, to assess the red reflex and observe for organic pathology. Examination reveals equal pupillary light reflexes. A white reflex is noted on ophthalmoscopic examination of the right eye as compared with the left one. No detail can be seen in the right fundus. Your findings indicate the need for an urgent referral. Following ophthalmologic sultation, esotropia in this child was diagnosed as secondary to a retinoblastoma;

Case 8 — The poor vision in the right eye may be due to a long-standing amblyopia. If this is the case, an ophthalmologist will conclude that removal of the cataract would result in vision only as good as that during the adolescent years and surgery would probably not be recommended;

Case 9 — Often children become uncooperative with visual acuity testing due to poor vision in the eye being tested. This is sometimes misinterpreted as a behavioral rather than an ocular issue. This patient can return on another day to have the left eye tested first, with the right eye covered by an adhesive patch. If visual acuity measurement is still unsuccessful, the patient should be referred to an ophthalmologist for evaluation;

Case 10 — c: The child can have glaucoma, which causes buphthalmos, or enlargement of one or both eyes. Although glaucoma may be associated with increased tearing, sensitivity to light, and a hazy or white cornea, the only sign of glaucoma in some children may be enlargement of the eye or eyes. This child should be referred immediately to an ophthalmologist for diagnosis and management.

Chapter 4 — Diseases of the Eyelids

Tests

1. A—D; **2.** F; **3.** C; **4.** E; **5.** E; **6.** F; **7.** E; **8.** B; **9.** A, B; **10.** C.

Clinical Cases

Case 1 — chalazion; **Case 2** — lagophthalmos; **Case 3** — hordeolum; **Case 4** — blepharitis; **Case 5** — ptosis; **Case 6** — chalazion; **Case 7** — eyelid allergic edema; **Case 8** — eyelid abscess; **Case 9** — simple blepharitis; **Case 10** — lagophthalmos.

Chapter 5 — Disorders of the Lacrimal System

Tests

1. A, B, D, E, F; **2.** B, C, D, F; **3.** A, D, E; **4.** A, E, F; **5.** A, B, D, E, F; **6.** A, C, D, E, F; **7.** D; **8.** C; **9.** B, C, D, F; **10.** A—F;

Clinical Cases

Case 1 — acute dacryoadenitis; **Case 2** — chronic dacryoadenitis; **Case 3** — benign tumor of the lacrimal gland; treatment: complete excision without disruption including the excision of the periorbital tissues or the bone to avoid seeding of the tumor into the adjacent tissues; **Case 4** — dry eye syndrome of level 2 (moderate); treatment: dietary modification (drinking enough water, omega-3 fatty acid), environmental management, artificial tear drops and ointments, topical anti-inflammatory agents, control of ocular allergy, stop wearing contact lenses; **Case 5** — neonatal dacryocystitis, massage of the lacrimal passages followed by cleaning of any discharge and instillation of disinfectants and antibiotic drops; **Case 6** — acute dacryocystitis; treatment: local hot compresses several times a day and systemic NSAIDs to relieve the pain, topical application of antibiotic drops and ointment and systemic antibiotics for 7 days; **Case 7** — chronic dacryocystitis; topical antibiotics, forceful lacrimal irrigation, surgical probing, dacryocystorhinostomy, dacryocystectomy.

Chapter 6 — Disorders of the Orbit

Tests

1. A—E; **2.** A; **3.** B; **4.** E; **5.** B; **6.** A; **7.** E; **8.** B; **9.** C; **10.** E; **11.** A; **12.** E; **13.** D; **14.** E; **15.** D.

Clinical Cases

Case 1 — E: MRI, CT; **Case 2** — E: orbital cellulitis; **Case 3** — A: carotid-cavernous fistula; **Case 4** — B: cavernous sinus thrombosis; **Case 5** — A: thyrotoxic exophthalmos; **Case 6** — B: edematous exophthalmos; **Case 7** — D: endocrine myopathy; **Case 8** — C: pseudotumor; **Case 9** — E: dermoid cyst; **Case 10** — A: optic nerve glioma.

Chapter 7 — Diseases of the Conjunctiva

Tests

1. A—F; **2.** A, B, D, F; **3.** A—F; **4.** A, B; **5.** A, D—F; **6.** A, B, C, F; **7.** B, C, E, F; **8.** B—F; **9.** A—D; **10.** A, C, D, E.

Clinical Cases

Case 1 — acute conjunctivitis of both eyes; **Case 2** — chronic bacterial conjunctivitis; **Case 3** — neonatal gonococcal conjunctivitis; **Case 4** — diphtheritic conjunctivitis; **Case 5** — adenoviral conjunctivitis; **Case 6** — adult inclusion conjunctivitis (paratrachoma); **Case 7** — trachoma; **Case 8** — giant papillary conjunctivitis; **Case 9** — seasonal allergic conjunctivitis; **Case 10** — atopic keratoconjunctivitis.

Chapter 8 — Diseases of the Cornea

Tests

1. A, B, C, D, F; **2.** A—F; **3.** B, C, E; **4.** F; **5.** A—C, E, F; **6.** A—E; **7.** A, B; **8.** A, D; **9.** A—E; **10.** A—D; **11.** A—C; **12.** A—E.

Clinical Cases

Case 1 — corneal erosion, removal of the pieces of mascara, topical antibiotics for infection prevention, lubricants for epithelization; **Case 2** — Thygeson's superficial punctate keratitis, topical steroids or cyclosporine drops; **Case 3** — corneal ulcer, systemic and topical broad-spectrum antibiotics, cycloplegics, oral pain medication; **Case 4** — dendritic herpetic keratitis, systemic and topical anti-viral therapy; **Case 5** — herpetic disciform keratitis, combined corticosteroid and antiviral therapy, timolol; **Case 6** — acanthamoeba keratitis, the patient should stop wearing contact lenses, topical cationic antiseptic biguanides with or without a diamidine, aminoglycosides, cycloplegics, oral NSAIDs; **Case 7** — keratomycosis, frequent topical anti-fungal drugs, oral antimycotic agents, topical cycloplegics; **Case 8** — neuroparalytic keratitis, MRI for identifying an underlying cause, topical application of lubricants; **Case 9** — syphilitic interstitial keratitis, systemic treatment of syphilis, systemic corticosteroids, topical steroids, cycloplegics; **Case 10** — arcus senilis, does not require any topical treatment; **Case 11** — keratoconus, soft contact lenses, RGP contact lenses; **Case 12** — Fuchs' corneal dystrophy, instillation of hypertonic solutions and/or ointments, lubricant eye drops, therapeutic soft contact lenses.

Chapter 9 — Diseases of the Sclera

Tests

1. A—C, E, F; **2.** B—D, F; **3.** C, D; **4.** A—F; **5.** E; **6.** A—F.

Clinical Cases

Case 1 — diffuse anterior scleritis, polyarteritis, topical and systemic steroids, NSAIDs; **Case 2** — episcleritis; **Case 3** — necrotizing scleritis, complicated cataract, secondary glaucoma on both eyes; **Case 4** — the patient has posterior scleral staphyloma due to a high myopia.

Chapter 10 — Diseases of the Uvea

Tests

1. A—F; **2.** A, C—F; **3.** B—D; **4.** A, C—E; **5.** A, C, D, F; **6.** A, B, D, F; **7.** B, E, F; **8.** A—D, F; **9.** B—F; **10.** B—F.

Clinical Cases

Case 1 — acute anterior uveitis of the right eye; **Case 2** — acute anterior uveitis of both eyes as a complication of influenza infection; **Case 3** — acute rheumatic iridocyclitis of the left eye; **Case 4** — mydriatic eye drops, such as atropine 1 %, tropicamide 1 %; topical steroids — prednisolone acetate 1 %, dexamethasone sodium phosphate 0.1 %; systemic analgesics such as paracetamol, ibuprofen; sunglasses; warm compresses. Refer the patient to an ophthalmologist for possible initiation of the underlying condition treatment; **Case 5** — choroiditis of the left eye; **Case 6** — acute post-traumatic endophthalmitis; **Case 7** — syphilitic choroiditis; **Case 8** — tuberculous choroiditis; **Case 9** — toxoplasmosis choroiditis; **Case 10** — iris nevus, observation every six months; **Case 11** — choroidal melanoma.

Chapter 11 — Lens Diseases

Tests

1. D; **2.** C; **3.** E; **4.** B; **5.** E; **6.** C; **7.** A; **8.** C; **9.** B; **10.** E.

Clinical Cases

Case 1 — C: Marfan's syndrome; **Case 2** — A: Weill—Marchesani syndrome, microspherophakia; **Case 3** — B: congenital cataract; **Case 4** — D: blue dot cataract; **Case 5** — A: nuclear cataract; **Case 6** — B: senile nuclear cataract; **Case 7** — C: incipient senile cataract; **Case 8** — A: immature senile cataract; **Case 9** — E: Morgagnian cataract; **Case 10** — C: secondary cataract.

Chapter 12 — Glaucoma

Tests

1. D, F, A, E, C, B; **2.** A—F; **3.** A, C, E, F; **4.** A—E; **5.** A, B, C, F; **6.** E, B, D, C, A, F; **7.** A—F; **8.** A, B, C, F; **9.** A, B, C, F; **10.** A—D.

Clinical Cases

Case 1 — A: primary open-angle glaucoma; **Case 2** — A: Timolol 0.25—0.5 % 2 times a day; **Case 3** — D: acute angle-closure glaucoma; **Case 4** — F: Pilocarpine 1 % eye drops every 15 min for an hour, then every hour; A: Timolol 0.25—0.5 % 2 times a day; B: analgesics; D: osmotic therapy; **Case 5** — E: acute angle-closure glaucoma — peripheral laser iridotomy; **Case 6** — C: normal tension glaucoma; **Case 7** — E: congenital glaucoma — surgical treatment, goniotomy; **Case 8** — C: pseudoexfoliation glaucoma;

Case 9 — D: phacomorphic glaucoma; **Case 10** — E: neovascular glaucoma.

Chapter 13 — Diseases of the Retina

Tests

1. E; **2.** E; **3.** E; **4.** B, C, D; **5.** E; **6.** E; **7.** E; **8.** A, C, D; **9.** E; **10.** E.

Clinical Cases

Case 1 — D: central retinal artery occlusion; **Case 2** — A: antispasmodics; **Case 3** — B: central retinal vein thrombosis; **Case 4** — A: retinal degeneration, B: secondary glaucoma, D: optic nerve atrophy; **Case 5** — B: retinal detachment; **Case 6** — retinitis pigmentosa; treatment: vasodilators, vitamins, and antioxidants; **Case 7** — dry form of AMD; **Case 8** — C: central retinal vein outflow obstruction due to thrombosis; **Case 9** — C: chronic inflammatory focus; **Case 10** — A: fovea.

Chapter 14 — Diseases of the Optic Nerve

Tests

1. A, C, D, E; **2.** A—F; **3.** A, B, C, E, F; **4.** A—E; **5.** B, D, E; **6.** A—F; **7.** C, D; **8.** B, D, E, F; **9.** C, E, F; **10.** A—F.

Clinical Cases

Case 1 — B: retrobulbar neuritis; **Case 2** — D: ischemic optic neuropathy; **Case 3** — C: papilledema; **Case 4** — F: toxic optic neuropathy; **Case 5** — E: optic atrophy.

Chapter 15 — Diseases of the Vitreous Body

Tests

1. B, C, E, F; **2.** B—D, F; **3.** F; **4.** A, C, D, F; **5.** F; **6.** A—E; **7.** A—F.

Clinical Cases

Case 1 — persistent hyperplastic primary vitreous; **Case 2** — synchysis scintillans (or cholesterolosis bulbi); **Case 3** — vitreous hemorrhage.

Chapter 16 — Ocular Trauma

Tests

1. C; **2.** B; **3.** D; **4.** B—F; **5.** A, B, D—F; **6.** E; **7.** C; **8.** C; **9.** A—E; **10.** A, B; **11.** C, D; **12.** E; **13.** F; **14.** A—F; **15.** C.

Clinical Cases

Case 1 — traumatic corneal erosion; topical use of antibiotic ointment, epithelizing gels, monocular bandage; **Case 2** — right eye — subluxation of the lens, mild myopia; left eye — luxation of the lens into the anterior chamber, secondary phakotopic glaucoma; the patient probably has Marfan's syndrome; left eye — intracapsular lens extraction is indicated; **Case 3** — penetrating corneal injury of the right eye, prolaps of the iris; first aid: topical and systemic antibiotics, binocular bandage, tetanus prophylaxis if needed; treatment: surgical repair of the laceration after investigation for an intraocular foreign body; **Case 4** — posttraumatic hyphema of the left eye; strict bed regimen with the head raised at 30—45 degrees, hemostatic therapy, and binocular eye bandage to prevent recurrent bleeding; **Case 5** — chemical burn of the left eye; copious irrigation with sterile water or balanced saline solution until pH is neutral; topical antibiotics and antiseptics, tetanus prophylaxis if not current; urgent transportation of the patient to the nearest ophthalmologic department; **Case 6** — orbital floor fracture with entrapment, surgical repair to release the herniated tissue and repair the orbital floor; **Case 7** — a metallic intraocular foreign body, tetanus prophilaxis, intravenous antibiotics, surgical repair of corneal laceration, lensectomy, pars plana vitrectomy, IOFB removal and postoperation treatment; **Case 8** — III-grade alkali burn; tetanus prophylaxis, topical antibiotic ointments, midriatics, steroids, lubricants, oral ascorbic acid, analgetics, therapeutic soft contact lenses and massage of the conjunctival fornices with glass plates twice a day for symblepharon formation prevention; **Case 9** — thermal burn of both eyes; topical antibiotic ointments, topical mydriatics, instillation of lubricating gels, bandage contact lenses, oral analgesics, cool compresses to reduce pain; **Case 10** — blunt trauma (contusion) of the left eye, eyelid hematoma, subconjunctival hemorrhage, retinal Berlin's edema; the cause for visual decrease is retinal Berlin's edema; the recovery prognosis relatively good since all the observed conditions can be treated successfully; **Case 11** — the patient suffers from photokeratitis or electroophthalmia; atropine drops, ophthalmic antibiotic ointments, lubricating eye drops, oral pain medication, cold compresses and sunglasses for symptomatic relief; the disease could be prevented by wearing protective glasses or a welding helmet.

Chapter 17 — Ocular Emergencies

Tests

1. A—C, F; **2.** B; **3.** A, B, D, F; **4.** A, E; **5.** D, F; **6.** A, C, D; **7.** A—E; **8.** A—C, E; **9.** A; **10.** A—D, F; **11.** D; **12.** A—C, E, F; **13.** C.

Clinical Cases

Case 1 — acute angle-closure glaucoma; instillations of Pilocarpine 1 % eye drops every 15 min for an hour, followed by instillation every hour; Timolol 0.25—0.5 % 2 times a day; Brinzolamide 1 % 2 times per day and Dexamethasone; intravenously Acetazolamide 500 mg, Mannitol 20 %, or Glycerol orally, systemic analgesics and sedatives, counter-irritant procedures: hot foot baths, mustard plasters on the gastrocnemius muscles, salt laxatives, and hirudotherapy (leeches to the temple); referral to the ophthalmology department; **Case 2** — acute iridocyclitis; instillation of mydriatic drops (Atropine sulfate 1 %, Tropicamide 1 %), topical steroids (Prednisolone acetate 1 % or Dexamethasone sodium phosphate 0.1 %) every 2 hours, systemic analgesics; **Case 3** — adenoviral conjunctivitis; cool compresses, topical antiviral medications (Interferon) 6—8 times a day; ointment 2—3 times a day, lubricants (Polyethylene Glycol 0.4 %, Propylene Glycol 0.3 %, Hypromellose 0.3 %, Carbomer 0.2 %), systemic antihistamine medications (Mebhydrolin 0.05—0.1 g, Chloropyramine hydrochloride 25 mg, Loratadine 10 mg) and vitamins; **Case 4** — corneal ulcer serpens, topical broad-spectrum antibiotics (Ofloxacin 0.3 %, Levofloxacin 0.5 %, Lomefloxacin 0.3 % and Gentamicin 0.3 %), artificial tears (Carbomer 0.2 %, Dexpanthenol 5 %, Solcoseryl 20 %), systemic analgesics and antihistamine medications; referral to an ophthalmologist for analysis of the causative agent and possible treatment modification; **Case 5** — acute dacryoadenitis of the left eye; instillation of disinfectant medications (Furacilin 1:1500, Sulfacyl Sodium 30 %), antibiotic ointments (Tobramycin 0.3 %, Tetracycline 1 %), systemic analgesics (Paracetamol 500 mg, Ibuprofen 200—400 mg), antipyretics for fever, UHF-therapy, consultation with an ophthalmologist; **Case 6** — neonatal dacryocystitis; instillation of disinfectants and antibiotic drops, massage of the lacrimal passages; **Case 7** — orbital phlegmon of the right eye; intravenous broad-spectrum antibiotics in high doses up to 10 days (Ceftriaxone 1 g, Cefuroxime 750 mg, Gentamicin 40 mg/ml), analgesics (Paracetamol 500 mg, Ibuprofen 200—400 mg) and anti-inflammatory drugs (Diclofenac 0.1 %, Indometacin 1 ml), systemic antihistamine (Mebhydrolin 0.05—0.1 g, Chloropyramine hydrochloride 25 mg, Loratadine 10 mg), referral to an ophthalmologist and an ENT specialist for close observation and consultation; **Case 8** — central retinal artery occlusion; firm ocular massage, sublingual Nitroglycerin, Acetazolamide 500 mg intravenously, topical Timolol 0.5 %, Papaverine 40 mg intramuscularly, Mannitol 20 % or Glycerol 50 % intravenously, carbogen therapy — breathing into a paper bag or inhalation of 95 % oxygen and 5 % carbon dioxide mixture for 10 minutes; after first aid administration — referral to an ophthalmologist for a through medical evaluation and further management; **Case 9** — central retinal vein occlusion; intravenously Aminophylline 2.4 % 5—10 ml in Sol. Glucose 20% 20 ml, intramuscularly solution of Magnesium Sulfate 25 % 10 ml + Papaverine 40 mg + Dibasol 2 % 0.5 ml; Heparin 10 IU; parabulbar Fibrinolizin 1 IU + Heparin 0.5 IU + Dexamethasone 0.5 ml; orally vitamins B, C, E; the patient must be referred to the ophthalmology department for thorough examination and following treatment; **Case 10** — hyphema, bed rest with the head elevated by 30°—45°, limit any eye movement with binocular eye bandage, cool compresses, systemic analgesics (Paracetamol 500 mg, Ibuprofen 200—400 mg); **Case 11** — penetrating corneal injury with IOFB; washing the wound with disinfectant solutions, instillation of broad-spectrum antibiotic drops (Ofloxacin 0.3 %, Tobramycin 0.3 %, Levofloxacin 0.5 %), tetanus prophylaxis, binocular aseptic bandage and immediate referral to the ophthalmic trauma department for detailed examination and surgical removal of the IOFB; **Case 12** — chemical ocular burn, irrigation with sterile isotonic saline or with any watery solution of neutral pH for at least 30 minutes, until ocular surface pH is neutralized; application of antibiotic ointment and monocular bandage, tetanus prophylaxis, systemic analgesics and referral to an eye clinic

for examination and following treatment; **Case 13** — posttraumatic endophthalmitis; intravitreal and systemic broad-spectrum antibiotics (Vancomycin 1 mg/0.1 ml, Amikacin 0.4 mg/0.1 ml, Ceftazidime 2.25 mg/0.1 ml, Ceftazidime 2 mg/0.1 ml; Gentamicin 40 mg/ml, Ciprofloxacin 250—500 mg, Cefazolin 0.5—1.0 g, Tobramycin 40 mg/ml), topical steroids (Prednisolone acetate 1 %, Dexamethasone 0.1 %), mydriatics (Atropine sulfate 1 %, Tropicamide 1 %), systemic analgesics, referral to the ophthalmology department for the following treatment.

Chapter 18 — Ocular Manifestations of Systemic Diseases

Tests

1. A—C, E, F; **2.** A—F; **3.** A—F; **4.** D; **5.** B—F; **6.** A—E; **7.** A—F; **8.** A—F; **9.** A—E; **10.** A—F.

Clinical Cases

Case 1 — hypertensive retinopathy; **Case 2** — malignant hypertensive retinopathy, consultation with a nephrologist for suspected renal failure; **Case 3** — atherosclerotic retinopathy; **Case 4** — acute leukemia; **Case 5** — moderate nonproliferative diabetic retinopathy; **Case 6** — severe nonproliferative diabetic retinopathy; **Case 7** — proliferative diabetic retinopathy; **Case 8** — AIDS-associated retinopathy; **Case 9** — CMV retinitis; **Case 10** — the picture of a normal fundus.

Chapter 19 — Disorders of Ocular Motility and Strabismus

Tests

1. A, D, E, F; **2.** A, B, E, F; **3.** B, C, E, F; **4.** A, B, E, F; **5.** A, C, E, F; **6.** B, D, E; **7.** A, B, D, E; **8.** A, E, F; **9.** F; **10.** A—F.

Clinical Cases

Case 1 — concomitant convergent heterotropia (esotropia) of the right eye; **Case 2** — concomitant divergent heterotropia (exotropia) of the left eye; **Case 3** — infantile esotropia and amblyopia of the left eye; patching of the right eye for 6 hours a day for a month; **Case 4** — right constant esotropia with anisometropic hypermetropia, amblyopia; optical correction; **Case 5** — alternating convergent strabismus; **Case 6** — secondary exotropia of the left eye; cosmetic surgery; **Case 7** — concomitant divergent strabismus and amblyopia of the left eye; cosmetic surgical treatment; **Case 8** — incomitant exotropia of the right eye due to oculomotor nerve damage; **Case 9** — paralytic strabismus of the left eye caused by IV (trochlear) cranial nerve paresis due to hypertensive crisis; **Case 10** — nystagmus due to multiple sclerosis.

Appendix 3

LIST OF RECOMMENDED READING

Textbooks

Denniston A.K.O., Murray P.I. Oxford Handbook of Ophthalmology / Oxford University Press; 2014. — 1069 p.

Hollwich F. Ophthalmology. A Short Textbook/ Thieme; 1985. — 363 p.

Jogi Renu. Basic Ophthalmology / Jaypee Brothers Medical Publishers Ltd; 2009. — 502 p.

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Internet Sources

https://www.aao.org — American Academy of Ophthalmology; contains updates on every aspect of ophthalmology.

http://www.djo.harvard.edu — Digital Journal of Ophthalmology; contains clinical case presentations and quizzes from American ophthalmic hospitals.

http://dro.hs.columbia.edu — Digital Reference of Ophthalmology; it is an educational resource for medical students that contains color atlas and clinical cases reviews from Edward S Harkness Eye Institute, Columbia University.

http://www.eyerounds.org — a service of the University of Iowa Department of Ophthalmology and Visual Sciences; contains case reports, images, videos and tutorials for students, residents, physicians, and patients.

http://www.sarawakeyecare.com — a service of the Ophthalmology Unit of University of Malaysia, Sarawak or UNIMAS; contains educational materials, publications, clinical cases and atlas of ophthalmology.

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