OPHTHALMOLOGY

for international students for the speciality « Dentistry»

Минск БГМУ 2016

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для иностранных учащихся по специальности «Стоматология»

OPHTHALMOLOGY

for international students for the speciality «Dentistry»

Учебно-методическое пособие

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Содержит понятие о специальности «Офтальмология», вопросы к практическим занятиям, задачи, клинические случаи, тематические тесты, описание офтальмо-стоматологических синдромов и рекомендуемую литературу.

Предназначено для студентов-стоматологов 4-го курса медицинского факультета иностранных учащихся, изучающих учебную дисциплину «Офтальмология» на английском языке.

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INTRODUCTION TO OPHTHALMOLOGY

«Ophthalmology» is the medical discipline containing modern scientific data on etiology, pathogenesis, clinical picture, diagnostics, treatment and prevention of diseases of the organ of vision.

This guide manual is mainly devoted to ocular-dental symptoms.

There is a group of general diseases with characteristic ocular and dental manifestations, having, as a rule, hereditary nature.

TOPICS AND QUESTIONS FOR THE SEMINARS

GENERAL OPHTHALMOLOGY

Anatomy, Physiology, Examination Methods

Anatomy of the Eye

1. Brief anatomy of the eyeball. Coats, segments and chambers of the eyeball.
2. Nerve supply of the eyeball. Ciliary ganglion.
3. Blood supply of the eyeball. Venous drainage of the eyeball and orbit, clinical significance.

Physiology of Eye and Vision

4. The light sense. Light and dark adaptation.
5. Colour sense, tests for colour vision.

Examination Methods

6. Central vision (visual acuity).
7. Normal visual field, physiological blind spot. Visual field defects. Etiology. Diagnosis. Clinical value.
8. Computerised tomographic examination methods.

Practical Part

- External ocular examination.
- Testing of distant visual acuity.
- Slit-lamp biomicroscopy. Applications of biomicroscopy.
- Administration of eye drops and ointment to the inferior conjunctival sac.
- Ophthalmoscopy. Description of the normal fundus.
- Red reflex examination in diagnosing opacities in the refractive media.
- Anaesthesia in ophthalmology.

Optics and Refraction. Strabismus

Optics and Refraction

9. Refractive states of the eye. Correction of refractive errors with spectacles.
10. Accommodation: mechanism, examination methods, anomalies of accommodation.
11. Spasm and paralysis of accommodation. Causes, treatment, correction. Asthenopia.

12. Presbyopia: pathophysiology and causes, clinical features, optical treatment.
13. Hypermetropia: classification, optical treatment.
14. Myopia. Classification. Optical treatment of myopia.
15. Progressive myopia. Complications of myopia.
16. Aphakia. Treatment. Modern tendencies of intraocular lens implantation.
17. Astigmatism: types, correction.
18. Contact lenses for correction of refractive errors. Types of contact lenses.
19. Refractive surgery (advantages and disadvantages of various surgical techniques). Patient selection criteria. Contraindications.

Strabismus

20. Extraocular muscles: anatomy, blood supply, innervation, functions.
21. Prerequisites for development of binocular vision, examination methods. Heterophoria.
22. Amblyopia. Types. Treatment.
23. Concomitant strabismus. Clinical features. Examination. Treatment.
24. Paralytic strabismus: causes, diagnosis, treatment.

Practical Part

− Subjective method of finding out the clinical refraction of the eye. Identification of lens types and power.
− Objective method of finding out the error of refraction, retinoscopy.
− Prescription of appropriate glasses for patients with presbyopia according to refractive error for distance.
− Pupillary distance (PD) measurement.
− Differential diagnosis between concomitant and paralytic strabismus.

SPECIAL OPHTHALMOLOGY

Diseases of the Conjunctiva

1. Conjunctiva: three parts, anatomy, glands of conjunctiva. Blood supply, innervation, functions. Examination methods.
2. Infective conjunctivitis. Classification. Bacterial conjunctivitis (gonococcal, staphylococcal, pneumococcal, diphtheric). Investigations. Treatment. Prevention.
3. Degenerative conditions: pinguecula, pterygium. Dry eye. Diagnosis. Treatment.

Diseases of the Eyelids

4. Eyelids: gross anatomy, structure. Muscles of the eyelids. Blood supply, innervation, functions. Examination methods.
5. Diseases of the eyelids (blepharitis, chalazion, external hordeolum (stye), tumours). Etiology. Diagnosis. Treatment.
6. Anomalies in the position of the eye lashes and lid margin (trichiasis, entropion, ectropion, symblepharon, lagophthalmos, ptosis).

Diseases of the Lacrimal Apparatus

7. Lacrimal apparatus: anatomy, blood supply, innervation, functions. Examination methods. Tear film.
8. Lacrimal passages: anatomy. Elimination of tears. Examination methods.
9. Dacryoadenitis. Dacryocystitis. Etiology. Clinical features. Treatment.

DISEASES OF THE CORNEA, SCLERA, UVEAL TRACT

Diseases of the Cornea

10. Cornea: anatomy, source of nutrients, nerve supply, features, functions. Examination methods.
11. Keratitis. Classification. Clinical picture. Resolution of keratitis.

Diseases of the Sclera

12. Sclera, limbus: anatomy, functions.
13. Episcleritis, scleritis. Etiology. Diagnosis. Treatment.

Diseases of the Uveal Tract

14. Anatomy of the uveal tract.
15. Uveitis. Classification. Diagnosis. Treatment.
16. Malignant melanoma of the uveal tract. Clinical picture. Diagnosis. Treatment.

DISEASES OF THE LENS, GLAUCOMA

Diseases of the Lens

17. Lens: anatomy, metabolism, functions. Examination methods.
18. Congenital and acquired cataract. Classification. Diagnosis. Clinical features. Treatment.

Glaucoma

19. Chambers of the eyeball: anatomy, aqueous humour. Angle of anterior chamber: anatomy, functions.
20. Dynamics of aqueous humour: production, functions, aqueous outflow system.
21. Optic nerve: anatomy, blood supply. Examination methods.
22. Glaucomatous changes in the optic nerve. Pathogenesis. Diagnosis.
23. Glaucoma. Classification. Pathogenesis.
24. Congenital glaucoma. Classification, pathogenesis. Treatment.
25. Early diagnosis of glaucoma. Indications for surgical therapy. Surgical procedures for glaucoma.
26. Primary open angle glaucoma. Pathogenesis. Classification. Diagnosis. Management.
27. Acute primary angle-closure glaucoma. Etiology. Diagnosis. Treatment.
28. Secondary glaucomas. Classification, pathogenesis. Treatment.

Ocular Injuries, Systemic Ophthalmology

Ocular Injuries

29. Blow-out orbital fractures. Clinical features. Investigations. Management.

30. Injuries of the eyelids, conjunctiva; lacrimal apparatus lesions: clinical features. Investigations. Emergency intervention. Treatment.

31. Contusions of the eye. Classification. Clinical features. Investigations. Treatment.

32. Closed globe injuries. Clinical features. Investigations. Treatment. Prevention.

33. Subconjunctival scleral rupture. Diagnosis. Treatment.

34. Scleral and corneal penetrating/perforating injuries. Diagnosis. Emergency intervention. Treatment.

35. Penetrating injury with foreign body. Diagnosis. Emergency intervention. Complications. Treatment.

36. Sympathetic ophthalmitis. Clinical picture. Treatment. Prophylaxis.

37. Chemical injuries and thermal injuries. Classification. Clinical picture. Emergency intervention and further treatment.

Ophthalmo-dental syndromes

1. Ganglionitis (ciliary ganglionitis, naso-ciliary ophthalmia)
2. Neurofibromatosis.
3. Mikulicz syndrome.
4. Behcet's disease.
5. Stevens-Johnson syndrome.
6. Sjogren's disease.
7. Rare ophthalmo-dental syndromes and diseases with a common pathological process involving eyes and the oral cavities.

OPHTHALMO-DENTAL SYNDROMES IN DETAILS

Ganglionitis (ciliary ganglionitis, naso-ciliary ophthalmia)

The ciliary ganglion is a parasympathetic ganglion located in the posterior orbit. It measures 1–2 mm in the diameter and contains approximately 2,500 neurons. Preganglionic axons from the Edinger-Westphal nucleus travel along the oculomotor nerve and form synapses with these cells. The postganglionic axons run in the short ciliary nerves and innervate two eye muscles:

1) the sphincter pupillae constricts the pupil, a movement known as miosis. The opposite, mydriasis, is the dilation of the pupil.

2) the ciliaris contracts, releasing tension on the Zonular Fibers, making the lens more convex, also known as accommodation.

Both of these muscles are involuntary – they are controlled by the autonomic nervous system.

It is one of four parasympathetic ganglia of the head and neck. (The others are the submandibular ganglion, pterygopalatine ganglion, and otic ganglion).

Diseases of the ciliary ganglion produce a tonic pupil. This is a pupil that does not react to light (it is «fixed») and has an abnormally slow and prolonged response to attempted near vision (accommodation).

When a patient with an Adie pupil attempts to focus on a nearby object, the pupil (which would normally constrict rapidly) constricts slowly. On close inspection, the constricted pupil is not perfectly round. When the patient focuses on a more distant object (say, the far side of the room), the pupil (which would normally dilate immediately) remains constricted for several minutes, and then slowly dilates back to the expected size.

Tonic pupils are fairly common — they are seen in roughly 1 out of every 500 people. A patient with anisocoria (one pupil bigger than the other) whose pupil does not react to light (does not constrict when exposed to bright light) most likely has Adie syndrome.

But some other diseases can also denervate the ciliary ganglion. Peripheral neuropathies (such as diabetic neuropathy) occasionally produce tonic pupils. Herpes zoster virus can attack the ciliary ganglion. Trauma to the orbit can damage the short ciliary nerves.

Adie syndrome. Idiopathic degeneration of the ciliary ganglion.

It is a tonic pupil plus absent deep tendon reflexes. Adie syndrome is a fairly common, benign, idiopathic neuropathy that selectively affects the ciliary ganglion and the spinal cord neurons involved in deep tendon reflex arcs. It usually develops in the middle age, although it can occur in children. Early in

the course of Adie syndrome (when the cells of the ciliary ganglion have been destroyed, but before regeneration has occurred) the pupil will be fixed and dilated. The sphincter pupillae will be paralyzed. There will be no response to accommodation — the ciliary muscle is also paralyzed (fig. 1).

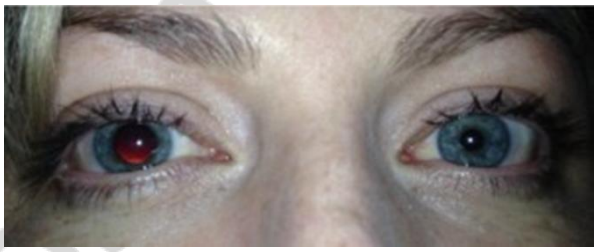


Fig. 1. Adie syndrome, right eye

With aberrant nerve regeneration, the pupil will remain fixed, but it will constrict with attempted near vision. The constriction will be abnormal («tonic»). Late in the course of Adie syndrome, the pupil becomes small (as all pupils do with old age). It will still be «fixed» (it will not constrict to bright light) and it will continue to show abnormal, tonic constriction with attempted near vision.

Argyll Robertson syndrome. It is a neurogenic dysfunction of the pupil in various diseases of the central nervous system. The pupil does not react to light, but its reaction to accommodation and convergence is saved; the reaction of the

pupil to sensory or mental irritation is weakened or absent; relative or fully-qualified (absolute) miosis; anisocoria or deformation of the pupils.

Parinaud syndrome. Cat-scratch-oculo-glandular syndrome.

It is caused by the tumor of the pineal gland which compresses the vertical gaze center at the rostral interstitial nucleus of medial longitudinal fasciculus.

Eye symptoms: paralysis of upgaze, downward gaze is usually preserved; the absence or weakening of the convergence; ptosis, violation of pupillary reactions, miosis, vertical nystagmus.

General manifestations: disturbance of sleep rhythm, dizziness.

Mikulicz syndrome. Morbus Mikulicz.

Benign lymphoepithelial lesion of the salivary and lacrimal glands (in cases of unilateral lesions are talking about a solid adenolymphoma).

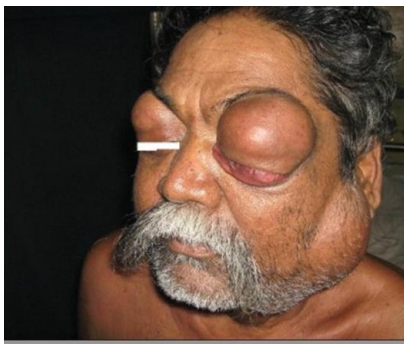


Fig. 2. Mikulicz syndrome

Progressive symmetrical and painless swelling of lacrimal and salivary glands accompanied by a decrease in their secretory function; often, also the mucous glands of the cheeks and tongue are involved to the process; the disease lasts for years. Late complications: atrophy of salivary glands with xerostomia and xerophthalmia. Expressed dental caries. Sometimes this syndrome is the partial manifestation of reticuloses or other leukemic blastosis. The prognosis is uncertain (fig. 2).

Neurofibromatosis. Recklinghausen's disease.

It is a systemic congenital disorder characterized by the formation of multiple benign tumors growing out of nervous sheath (neurilemma) and still undifferentiated (parent) nerve cells. These tumors can develop in the area of spinal nerves, their roots, inside the skull and in the spinal canal and cause spinal cord compression with corresponding neurological symptoms. Typically the nodes located on the trunk, neck and limbs, rarely on the soles and palms, the mucous membrane of the mouth, and rarely the bones and endocrine glands are affected. Men get sick about 2 times more often than women.

Eye symptoms occur in about 20 % of patients and sometimes may be the only manifestation of neurofibromatosis. Mainly neurofibromatosis of eyelid can occur. Nodes can be localized in the conjunctiva of the eyelids and eyeball, in the sclera, in the iris, very rarely-in the cornea. Changes in the fundus are in the form of flat, sharply restricted nodes and plaques of pinkish-yellow and white

color in the retina and at the optic disk. Choroid is a very rare localization of nodes. Sometimes develops hydrophthalmos and secondary glaucoma. The location of the tumor intraorbitally causes ptosis, exophthalmos, paresis or paralysis of extraocular muscles. Localization of nodes inside the skull is manifested by paresis of the abducens nerve, paralytic strabismus, congestive of the optic disk (fig. 3).

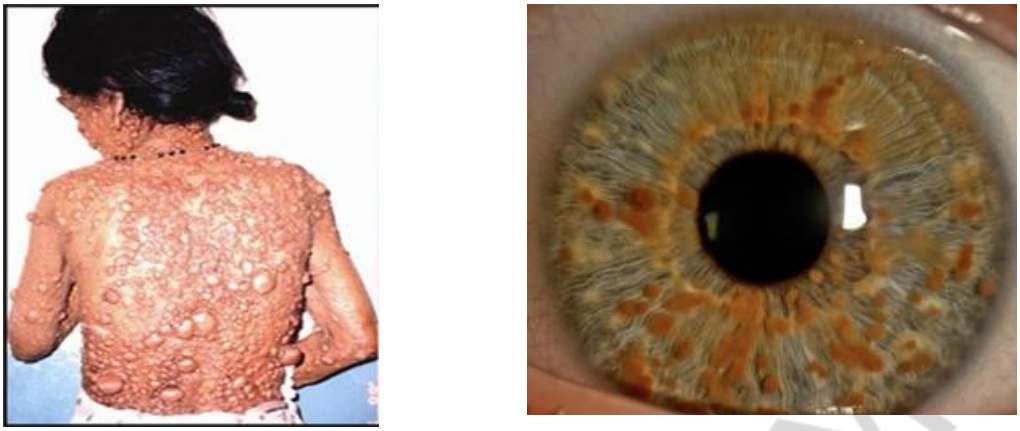


Fig. 3. Neurofibromatosis

Behcet's Disease. Presumably immune-mediated systemic vasculitis, often presented with mucous membrane ulceration and ocular involvements.

Triple-symptom complex of recurrent **oral aphthous and** genital ulcers and **uveitis.** The disease begins acutely, with fever, sometimes up to 40 □, chills and general malaise. As a systemic disease, it can also involve visceral organs such as the gastrointestinal tract, pulmonary, musculoskeletal, and neurological systems.

Ocular involvement can be in the form of anterior uveitis, posterior uveitis, or retinal vasculitis. Anterior uveitis presents with painful eyes, conjuctival redness, hypopyon, and decreased visual acuity, while posterior uveitis presents with painless decreased visual acuity and visual field floaters. A rare form of ocular (eye) involvement in this syndrome is retinal vasculitis, presented with painless decrease of vision and possibility of floaters or visual field defects.

Stevens-Johnson syndrome. Malignant exudative erythema.

It is a form of toxic epidermal necrolysis, is a life-threatening skin condition, in which cell death causes the epidermis to separate from the dermis. It is assumed that the disease has infectious or infectious-allergic etiology.

Eye symptoms include the following: red eye, tearing, dry eye, pain, blepharospasm, itching. In 98 % of cases catarrhal or purulent conjunctivitis is observed, in more severe cases, there is a membranous conjunctivitis with the formation of light yellow false membrane, often extending onto the cornea. It is likely the development of symblepharon and ankyloblepharon, inversion of the eyelids, trichiasis. In more severe cases ulcers of the cornea may develop.

The disease usually begins with fever, sore throat, and fatigue, which is commonly misdiagnosed and therefore treated with antibiotics. Ulcers and other lesions begin to appear in the mucous membranes, almost always in the mouth and lips, but also in the genital and anal regions. Those in the mouth are usually extremely painful and reduce the patient's ability to eat or drink. The disease also manifests by a characteristic macular-vesicular or bullous rash on the face, extensor surfaces of arms and legs, dorsum of the hands and feet, pubis and external genitals (fig. 4).

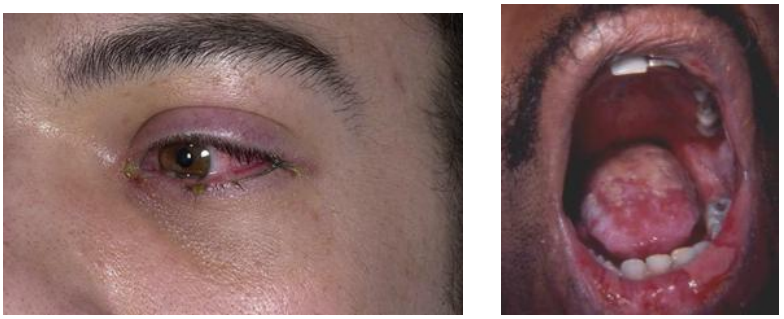


Fig. 4. Stevens-Johnson syndrome

Sjogren's disease. It is a chronic autoimmune disease in which people's white blood cells attack moisture-producing glands in the body. Although the hallmark symptoms are dry eyes and dry mouth, Sjögren's disease may also cause dysfunction of other organs such as the kidneys, gastrointestinal system, blood vessels, lungs, liver, pancreas, and the central nervous system. Patients may also experience extreme fatigue and joint pain with a high risk of developing lymphoma.

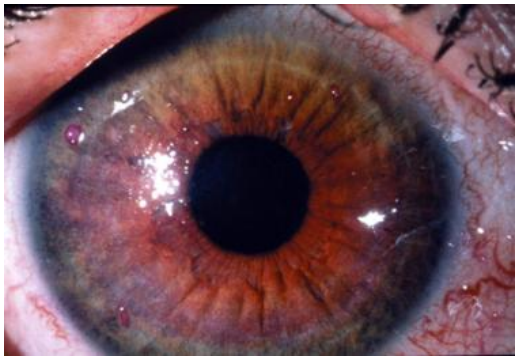


Fig. 5. Sjogren's disease

Dry eyes is a classic symptom of Sjogren's disease. Dryness of the eye can lead to more severe eye complications such as corneal ulcerations and eye infections. The **symptoms** also include eye itching, burning, photophobia. There are three stages of the disease: 1) hyposecretion of tear with the decrease of concentration of lysozyme; 2) dry conjunctivitis; 3) dry keratoconjunctivitis, filamentous keratitis (fig. 5).

It occurs predominantly in women older than 40 years and correlates with systemic lupus erythematosus, systemic sclerderma.

RARE OPHTHALMO-DENTAL SYNDROMES

Albers-Schönberg syndrome. Generalized diffuse osteosclerosis (Marble bone disease).

A syndrome of excessive calcification of bones causing marble like appearance with increased radiological density of the skeleton, is characterized mainly by multiple fractures.

Eye symptoms are caused by narrowing of the channels of the optic nerve and other bone holes of the eye sockets and the strangulation of the passing of vessels and nerves. Lagophthalmos, congestion of the optic nerve followed by secondary atrophy, tapetoretinal dystrophy, nystagmus, bilateral exophthalmos due to the deformation of the eye sockets, exotropia with impaired convergence, perhaps esotropia due to degeneration of abducens nerves, the weakening or absence of reaction of pupils to light.

Dental symptoms include caries, pathological changes of the mandible (fig. 6).

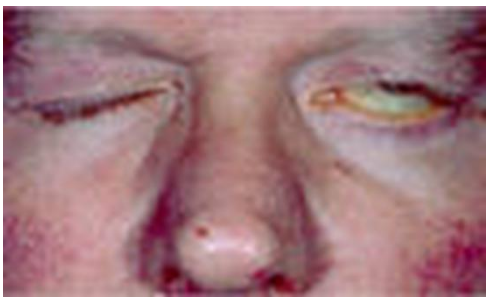


Fig. 6. Albers-Schönberg syndrome

Angelman syndrome. «Happy puppet» syndrome.

It is a neuro-genetic disorder characterized by severe intellectual and developmental disability, sleep disturbances, seizures, jerky movements (especially hand-flapping), frequent laughter or smiling, and usually a happy demeanor.

Eye symptoms are due to divergent strabismus. **Dental** changes: wide interdental spaces (fig. 7).

Fig. 7. Angelman syndrome

Appelt-Gerken-Lenz syndrome (syndrome Roberts).

This syndrome is usually observed in children, born as a result of post-term pregnancy, insufficient weight and body length. Tetraphokomelia (hands and feet are attached to shortened arms and legs), oxycephalia, cleft palate, cleft lip are clearly marked.

Eye symptoms: exophthalmia, hypertelorism, «antimongoloid» location of the palpebral fissure, corneal clouding.

Dental symptoms include hypoplastic nasal alae (narrowing of the nostrils that can decrease the width of the nasal base), beaked nose (a nose with a prominent bridge that gives it the appearance of being curved).

Ascher- Laffer syndrome.

The association of droopy eyelids, double lip and goiter.

Eye symptoms: blepharochalasis due to atrophy of skin and subcutaneous tissue. Possible spastic entropion, the offset under the skin of the lacrimal gland.

General manifestations: double upper lip due to edema of the mucosa of the lips and mucous membrane of the gums. Very rare double lower lip. Hypertrophy of the thyroid gland without endocrine disorders. Sometimes, these patients have long hooked nose and a long pale tongue (fig. 8).

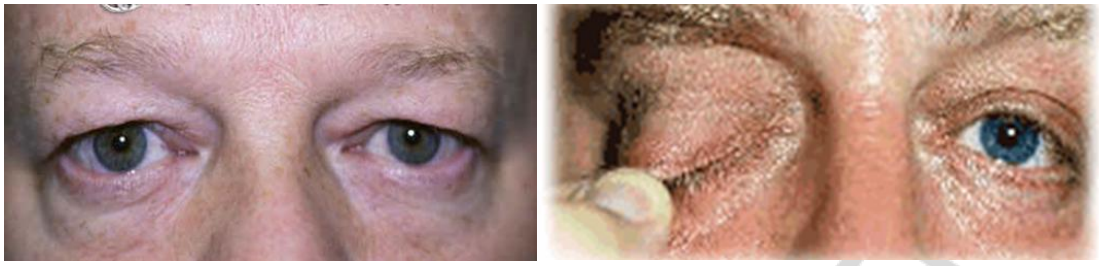


Fig. 8. Ascher-Laffer syndrome

Baader syndrome. Dermatostomatitis.

General catarrhal or pseudomembranous inflammation of mucous membranes with multiple skin rash of unknown etiology

Eye symptoms: hemorrhagic conjunctivitis, erosions of the conjunctiva of the eyelids and the eyeball.

General manifestations: fever, catarrhal or pseudomembranous inflammation of the mucous membranes of the nose, mouth, multiple rashes on the skin, especially the bright red maculopapular or papulovesicular rash, predominantly on the extremities. Often bronchitis, pneumonia (fig. 9).

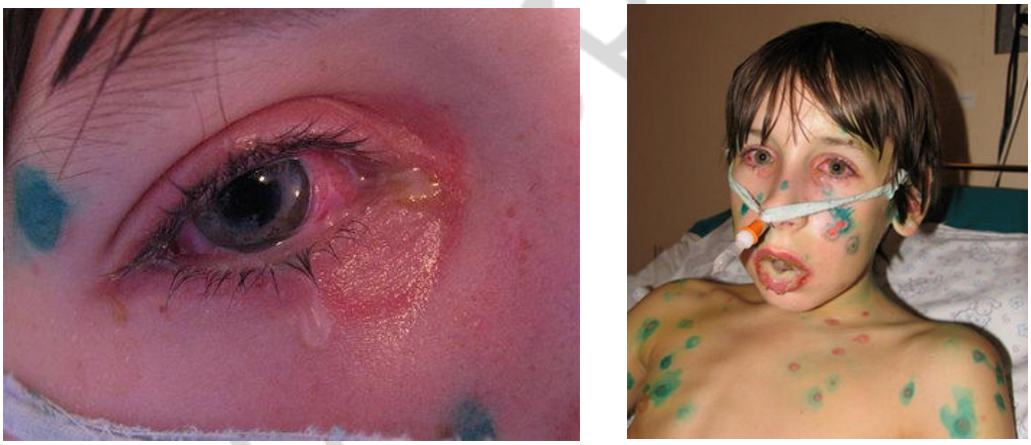


Fig. 9. Baader syndrome

Bamatter's syndrome. It is a rare developmental disturbance of connective tissue with too early aging processes of the skin and generalised osteopenia, also characterized by growth retardation, hyperlaxity, atrophy, and predisposition to fractures.

Eye symptoms — microcornea, microphthalmia, clouding of the corneas.

General manifestations: dwarf growth, premature aging (progeria), bone dysplasia, pigmented teeth. The appearance is reminiscent of the dwarfs.

Biemond syndromes.

1) **Eye symptoms:** strabismus, nystagmus. **General manifestations:** cerebellar ataxia, brachydactylia, mental retardation.

2) **Eye symptoms:** colobomas and atrophy of the stroma of the irises. **General manifestations:** even obesity, hypogenitalism, infantilism with primary amenorrhea, impotence, dysfunction of the hypothalamic-pituitary system.

3) **Congenital analgesia. Eye symptoms:** the absence of pain, decreased tactile and temperature sensitivity of corneas. **General manifestations:** the absence of pain and reduced tactile and thermal sensitivity of the teeth, bones, joints. Tendon areflexia.

Bloch-Sulzberger syndrome. Incontinentia pigmenti. Naevus pigmentosus systematicus.

The combination of pigmentary dermatosis with congenital anomalies dirty-brownish pigmented (usually symmetrical) skin irregular spots on the sides of the torso, shoulders and hips; often, the pigmentation begins with bullous or verrucose rash. The intensity of pigmentation decreases over time. Affects only women.

Eye symptoms: strabismus, congenital opacity of the cornea and the lens. **General manifestations:** alopecia, nail dystrophy, dental defects (the conical shape and the shortage of dentin), mental retardation, hirsutism, congenital dislocation of the hip joint (fig. 10).

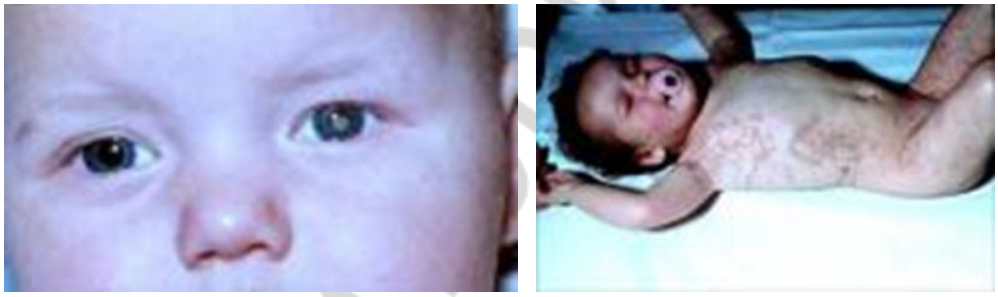


Fig. 10. Bloch-Sulzberger syndrome

Bremer syndrome. The combination of birth defects caused by disturbances of embryonic development.

Eye symptoms: in 80 % of patients — Horner syndrome and Fuchs heterochromia. Rarely — the decrease of sensitivity of the cornea, keratitis, paresis and paralysis of the trigeminal, abducent and facial nerve.

General manifestations: the asymmetry of the skull and chest, deformity of the spine (curvature, splitting arches) and foot, arachnodactyly, uneven growth of the teeth and improper position, changes in tendon reflexes, a breakdown of surface sensitivity, acrocyanosis. Often — cleft lip, cleft palate.

Waardenburg syndrome. Syndrome of outer germ layers dysplasia.

It is a rare genetic disorder most often characterized by varying degrees of deafness, minor defects in structures arising from the neural crest, and pigmentation anomalies.

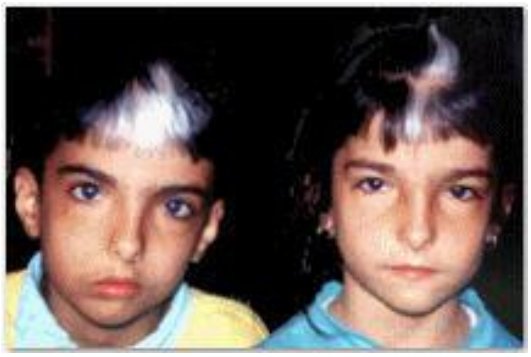


Fig. 11. Waardenburg syndrome

Eye symptoms : very pale or brilliantly blue eyes, eyes of two different colors (complete heterochromia), or eyes with one iris having two different colours (sectoral heterochromia). Appearance of wide-set eyes due to a prominent, broad nasal root (dystopia canthorum) also known as telecanthus. Rarely strabismus, refractive errors, areas of depigmentation on the periphery of the fundus, bilateral hydrophthalmos.

General manifestations: moderate to profound hearing loss, improper growth of teeth. A forelock of white hair (poliosis), or premature graying of the hair. A low hairline and eyebrows that touch in the middle. Patches of white pigmentation on the skin have been observed in some people. Sometimes, abnormalities of the arms (fig. 11).

Syndrome Weyers-Tyr. Syndrome of outer germ layer dysplasia.

Eye symptoms: unilateral abnormalities of the eyeball (hypo — or dysplasia of the iris, cataract, glaucoma, microphthalmos, blepharophimosis due to dysplasia of the bones of the orbit). Visual function is severely reduced.

General manifestations: facial asymmetry due to unilateral underdevelopment of the bones of the upper and lower jaws, the presence of teeth in the newborn, hypoplasia of tooth roots, premature eruption and loss of teeth. Deformation of the nose and the ear. Premature fusion of skull bones.

Weill-Marchesani Syndrome. Spherophakia-brachymorphia syndrome, congenital mesodermal dystrophy.

Eye symptoms include unusually small, round lenses of the eyes (spherophakia), which may be prone to dislocating (ectopia lentis), as well as other ocular defects (disorders of accommodation, secondary glaucoma). Due to such abnormalities, affected individuals may have varying degrees of visual impairment, ranging from myopia to blindness.

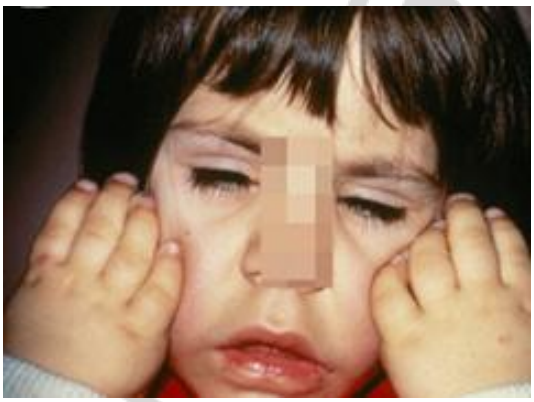


Fig. 12. Weill-Marchesani Syndrome

General manifestations: small (often dwarf) height, short, broad head (brachy-cephaly) and other facial abnormalities; hand defects, including unusually short fingers (brachydactyly). Frequently observed hypoplasia of the upper jaw, narrow palate, dental anomalies, joint stiffness, congenital heart defects (fig. 12) .

Syndrome Wildervanck. Cervico-oculo-acoustic syndrome, cervico-oculo-facial dystrophy.

Eye symptoms: uni — or bilateral paralysis of the abducens nerve retraction of

the eyeball, heterochromia of the iris, subconjunctivally lipoma, dermoid cyst of the eyelids.

General manifestations: congenital deaf**,** hypoplasia of the teeth. Mainly women are affected (fig. 13).

Fig. 13. Syndrome Wildervanck

Williams syndrome. The syndrome of «the face of an elf».

It is a rare neurodevelopmental disorder characterized by: a distinctive, «elfin» facial appearance, an unusually cheerful demeanor and ease with strangers.

Eye symptoms: epicanthus, strabismus.

General manifestations: a low nasal bridge; low height, small body weight at birth (2700), a wide upper jaw, small jaw, open mouth, protruding ears, developmental delay coupled with strong language skills; cardiovascular problems, such as supravalvular aortic stenosis and transient high blood calcium (fig. 14).

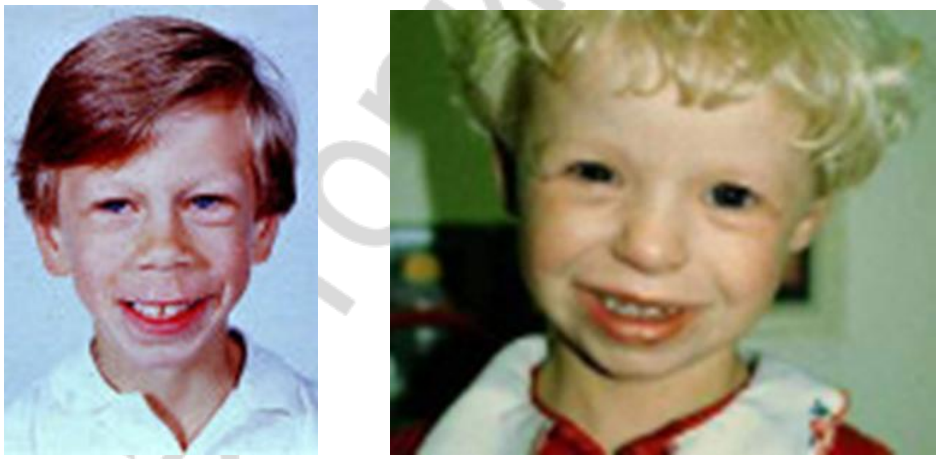


Fig. 14. Williams syndrome

Hutchinson-Gilford syndrome. Progeria, nanismus senilis.

Eye symptoms: exophthalmus, nystagmus, blurred lenses.

General manifestations: children have a senile appearance, thin gray hair, thin skin, resembling the skin in scleroderma, dis - or atrophic nails, acromicria, proportional dwarf growth. Growth stops in early childhood. Long retained baby teeth, delayed cutting of the teeth, which are arranged abnormally, sometimes absent. Often genital hypoplasia, osteoporosis, poor development of skeletal muscles (fig. 15).

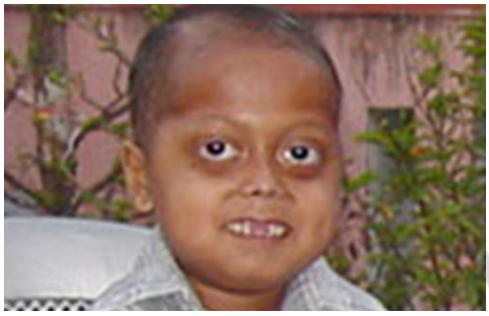



Fig. 15. Hutchinson-Gilford syndrome

Goldenhar syndrome. Oculo-Auriculo-Vertebral syndrome.

It is a rare congenital defect characterized by incomplete development of the ear, nose, soft palate, lip, and mandible. It is associated with anomalous development of the first and second branchial arch.

Eye symptoms: limbal dermoids, upper eyelid colobomas, (rarely lower), antimongoloid eyes, strabismus, ptosis.

General manifestations: anomalous auricles, hypoplasia of the lower and upper jaw, high arcuate palate, cleft palate, open bite, anomalies of the ribs and vertebrae, scoliosis (fig. 16).

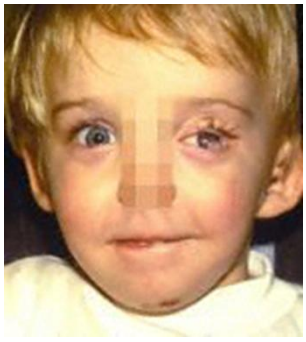
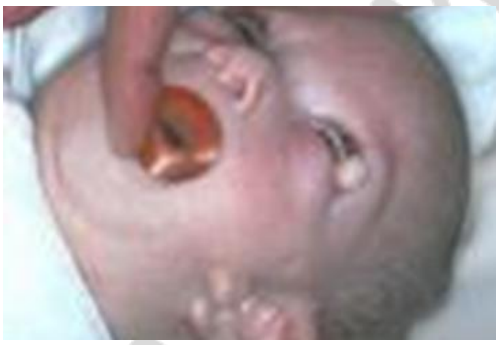
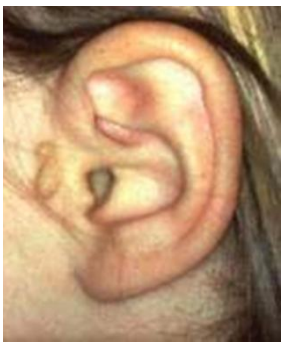




Fig. 16. Goldenhar syndrome

Gorlin-Cohen syndrome.

Eye symptoms: the hypertelorism (excessive width between two eyes), supraorbital hyperostosis.

General manifestations: microgenia (abnormal smallness of the chin), hypoplasia of teeth, aplasia of the permanent lateral incisors, asymmetry of the chest, chicken breast, stiffness of the elbow, carpal, hip, knee and ankle joints.

Grob syndrome. Lingual-facial dysplasia.

Complex of congenital anomalies.

Eye symptoms are non-constant: hypoplasia of the eyes, congenital cataract, refractive errors.

General manifestations: hypoplasia of the tongue or its complete absence, cleft lip and palate, multiple scars of the upper and lower lips, flat face.

Danbolt-Kloss syndrome. Acrodermatitis enteropathica.

Eye symptoms: conjunctivitis, blepharitis, photophobia, tearing, itching of the eyelids.

General manifestations: vesiculo-bullous rash around the mouth, in the elbow, popliteal fossae. The mucous membrane of the mouth is affected (severe stomatitis, cracks and erosion around the mouth).

De Lange syndrome.

Eye symptoms: hypertelorism, mongoloid location of the eyes, a bluish tinge to the skin around the eyes, ptosis, strabismus, nystagmus.

General manifestations: «clown face», high arcuate palate, late eruption of teeth, large gaps between teeth (fig. 17).

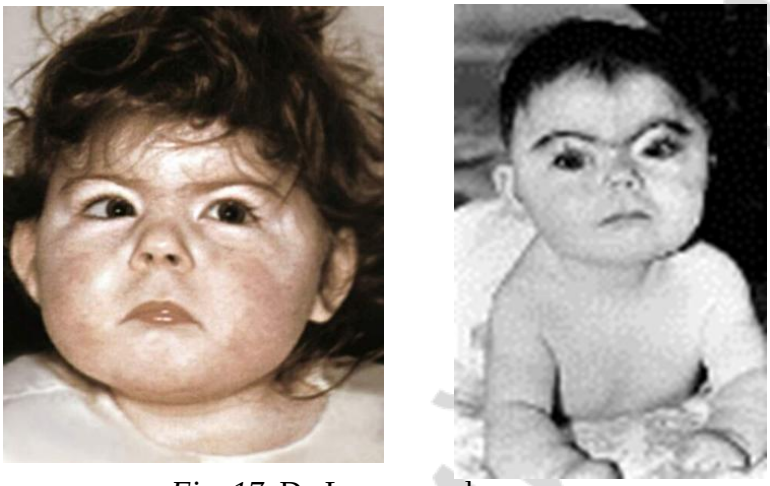


Fig. 17. De Lange syndrome

Seckel-Virchow syndrome. Dwarfism with the «bird's head».

Eye symptoms: hypertelorism, epicanthus, relatively large eyes, a light exophthalmos, strabismus.

General manifestations: dwarfism in combination with microcephaly, and a distinctive facial structure. High palate, partial adentia, enamel hypoplasia.

Oral-palpebrale synkinesia of Zoldan.

Eye symptoms: lifting upper eyelid with ptosis when opening the mouth and lowering of the eyelid when closing the mouth.

Cause: lesion of the brain stem with dysfunction of the oculomotor, trigeminal and facial nerves.

Klippel-Feil disease. Syndrome of short neck. Syndrome of cervical vertebrae synostosis.

Eye symptoms: paralytic strabismus, nystagmus, hypermetropia.

General manifestations: shortening of the neck, asymmetry of the face, splitting the soft palate, nonclosure of foramen ovale. Appearance — «the frog».

Kozlovoy syndrome. Kyrgyz dermatoosteolysis, syndrome of ulcerative lesions of the skin, acroosteolysis, keratitis and oligodentia.

Eye symptoms: recurrent keratitis, corneal clouding, impaired visual functions. **General manifestations:** multiple ulcerating and scaring papules on the oral mucosa and skin of the face; abnormal growth of the teeth; partial adentia.

Cockayne syndrome (2).

Eye symptoms caused by the development of dacryocystitis.

General manifestations: cleft lip and palate, cleft hands and feet, deafness.

Cohen Syndrome.

Eye symptoms: antimongoloid eyes, strabismus, myopia, microphthalmia, iris colobomas.

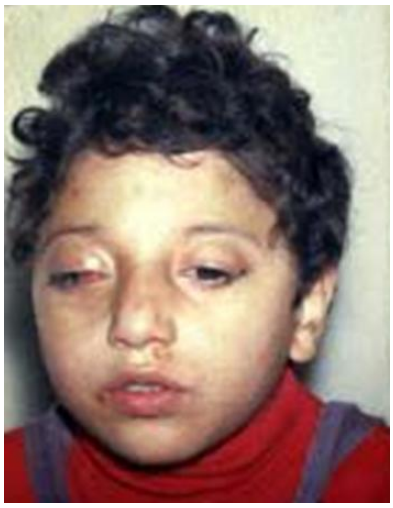


Fig. 18. Crouzon syndrome

General manifestations: open mouth with protruding incisors, hypoplasia of the upper jaw.

Crouzon syndrome. Craniofacial dysostosis.

Eye symptoms: hypertelorism, diverging strabismus, exophthalmos due to the reduction of the volume of the eye sockets; congestion of the optic nerve due to either increased intracranial pressure or compression of nerves in the restricted optical channel.

General manifestations: premature fusion of the sutures of the skull with its deformation as an extension in the fronto-temporal parts; hypoplasia of the upper jaw, the extension of the nose root; beaked nose, short upper lip; high palate, rare spinous teeth, large tongue, atresia of acoustic duct, deafness (fig. 18).

Curtius syndrome (1). Partial congenital gigantism with ectodermal dysplasia and endocrine disorders.

Eye symptoms: hemeralopia (night blindness), amblyopia.

General manifestations: the enormous growth of one half of the face or parts of the face (especially the upper jaw), or separate extremities; the tooth enamel dysplasia, hypodentia; dysplasia of the nails, hair; nevi on the face.

Curtius syndrome (3).

Eye symptoms: amblyopia with congenital nystagmus and marked convergent strabismus, hemeralopia (night blindness), projecting into the vitreous body optic nerve head.

General manifestations: microcephaly with severe debility, genital and breast hypoplasia, marked hypodentia.

Martin-Albright syndrome. Pseudoparathyreoid syndrome.

The disease is associated with genetically determined resistance of the kidney to the action of parathyroid hormone

Eye symptoms: progressive cataract, a disorder of ocular motility, diplopia.

General manifestation: short stature, short neck, hypoplasia of tooth enamel with early loss of teeth; pale brown skin pigmentation that resembles a geographic map.

Meyer-Schwickerth–Gruterich-Weyers syndrome. Oculodentodigital syndrome.

Eye symptoms: microphthalmos, microcornea, corneal opacity, congenital cataract; hypoplasia of the anterior mesodermal leaf of the iris — the iris has the form of porous tissue without crypts and lacunae; hypertension or glaucoma, pseudohypertelorism; often epicanthus, ptosis.

General manifestation: typical face — small nose with a pointed tip, hypoplasia of the alae and a wide nose bridge, generalized dysplasia of dental enamel with brown staining.

Mende syndrome.

Eye symptoms: epicanthus, mongoloid location of the eye, weak pigmentation of eyelashes and eyebrows, chronic blepharitis.

General manifestations: physical underdevelopment, short stature, partial albinism with pigmentary anomalies of the skin and hair, adherent ear lobes, cleft lip, delayed teething.

Mohr syndrome.

Eye symptoms: the hypertelorism, telecanthus (lateral displacement of the inner corner of eyes, with the normal interpupillary distance (unlike hypertelorism, wherein the interpupillary distance is increased)).

General manifestations: shortened, lobed, sometimes cleft tongue, multiple "bridle" between the alveolar process of the jaws and the mucous membrane of the lips, cleft palate, median pseudocleft of lip; the absence of central incisors.

Nager syndrome. Dysostosis acrofacialis.

Eye symptoms: antimongoloid eyes, hypoplasia or complete absence of eyelashes, coloboma of the lower eyelid in the area of the outer edge, microphthalmia.

General manifestations: the combination of symptoms of maxillofacial dysostosis (hypoplasia of the mandible, hypoplasia of the rudimentary molars) with hypoplasia of the 1st finger and radial bone.

Opitz syndrome. Syndrome C.

Eye symptoms: hypertelorism, mongoloid eyes, epicanthus, convergent strabismus.

General manifestations: anomalies of skull and face (among them macrostomia, high palate, extra bridle in the mouth); skeletal anomalies; other anomalies.

Pallister W syndrome.

Eye symptoms: telecanthus.

General manifestations: typical face with a high forehead, wide nose-tip, broad and flat upper jaw, cleft lip and palate, anomalies of the upper limbs.

Papiyon-Leage-Psoma syndrome. Oro-facio-digital syndrome type I.

Eye symptoms: epicanthus, telecanthus.

General manifestations: anomalies of the face and oral cavity (cleft palate, lobed tongue that is fixed by shortened frenulum; dystopia of teeth, aplasia of the lower incisors, increased tendency to caries of the teeth, extra teeth); skeletal anomalies.

Parry–Romberg syndrome. Progressive hemifacial atrophy.

This is a rare neurocutaneous syndrome characterized by progressive shrinkage and degeneration of the tissues beneath the skin, usually on only one

side of the face (hemifacial atrophy) but occasionally extending to other parts of the body.

Eye symptoms: lagophthalmus, enophthalmos, ptosis, miosis, redness of the conjunctiva, and decreased sweating (anhidrosis) of the affected side of the face; ophthalmoplegia, strabismus, uveitis, heterochromia.

General manifestations: trigeminal neuralgia, migraine; delayed eruption of teeth, dental root exposure or resorption of the dental roots on the affected side; difficulty or inability to normally open the mouth; temporomandibular joint disorder and spasm of the muscles of mastication on the affected side; atrophy of one side of the upper lip and tongue; total alopecia (fig. 19).

Patau syndrome. D1 trisomy syndrome. Trisomy of chromosome 13 syndrome.

This is one of the most common chromosomal abnormalities.

Eye symptoms: narrow palpebral fissures, epicanthus, micro - or anophthalmos, developmental disorders of the cornea, iris, anterior chamber angle; colobomas of the iris and choroid, nuclear cataract, persisting hyperplastic primary vitreous, dysplasia and congenital retinal detachment.

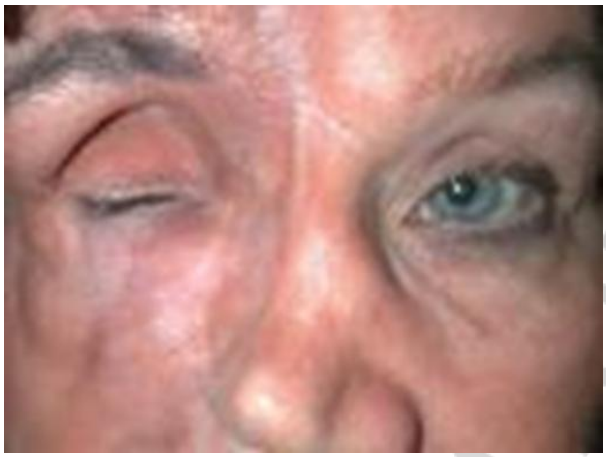


Fig. 19. Parry–Romberg syndrome

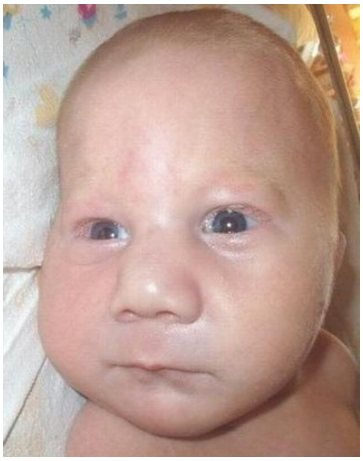


Fig. 20. Patau syndrome

General manifestations: small body weight at birth, microcephaly, deformation of the cerebral and facial skull, splitting his upper lip and / or palate, sunken nasal bridge, broad nasal root, low-set deformed auricles, and micrognathia, short neck, polydactyly, diffuse capillary hemangioma, malformations of the internal organs (fig. 20).

Peters syndrome.

Eye symptoms: central corneal leukoma, defect in descemet's membrane, iridocorneal adhesions, flattening of the anterior chamber with a possible secondary hydrophthalmus, persistent residues of pupillary membrane, sometimes buphthalmos.

General manifestations: ankylosis of limbs, cleft palate, intestine atresia, mental retardation (fig. 21).

Pillay syndrome. Ophthalmo-mandibular dysplasia.

Fig. 21. Peters syndrome

Eye symptoms: clouding of the corneas.

General manifestations:

1) anomalies of the lower jaw – temporomandibular fusion, the absence of the coronoid process, obtuse mandibular angle;

2) abnormalities of the upper extremities.

Rutherford syndrome.

Eye symptoms: dystrophy of the cornea with its opacity.

General manifestations: hypertrophy of the gums with violation of teething, hypodontia, sometimes mental retardation.

Rieger syndrome. Mesodermal dysgenesis of the cornea and iris.

Eye symptoms: megalocornea or microcornea, flat cornea; rear embryotoxon; remains of mesodermal tissue in the angle of the anterior chamber; hypoplasia of iris mesodermal leaf; deformation of the pupil in the form of a slit and its offset; ectopia of the lens, congenital cataract.

General manifestations: hypoplasia of the upper jaw, cleft palate, everted lower lip, oligodontia, the conical shape of the front teeth, the tooth enamel dysplasia (fig. 22).

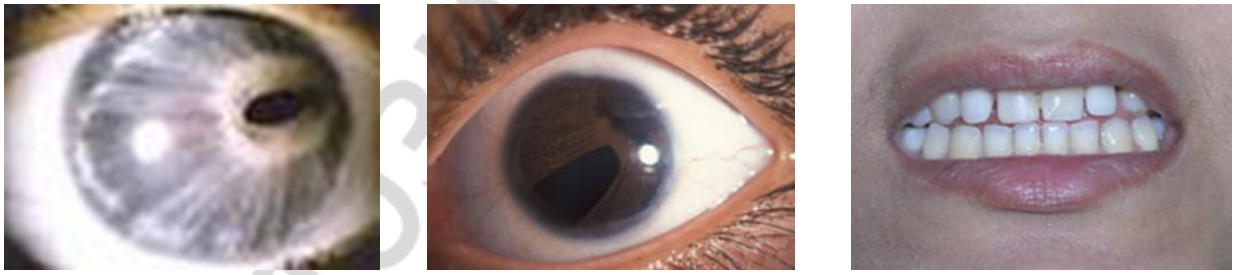


Fig. 22. Rieger syndrome

Robin syndrome.

Eye symptoms: bilateral microphthalmia or buphthalmos, strabismus, myopia, congenital cataract, glaucoma, retinal detachment.

General manifestations: sharp hypoplasia of the lower jaw, splitting the hard and soft palate, glossoptosis, microglossia. Anomalies of the jaw and the mouth are the cause of respiratory disorders and swallowing disorders.

Frequency in population 1 : 12 000.

Rossolimo-Melkersson-Rosenthal syndrome.

Eye symptoms: recurrent swelling and ptosis of the upper eyelid, the increase of the lacrimal gland, exophthalmos, keratoconjuctivitis, retrobulbar neuritis.

General manifestations: recurrent swelling of the lips and cheeks, facial paralysis; swelling of the oral mucosa; the decrease of salivation, folded tongue with atrophy of papillae.

Rapp-Hodgkin syndrome. Anhydrotic ectodermal dysplasia with cleft lip and palate.

Eye symptoms: frequently occurring purulent conjunctivitis.

General manifestations: dry thin skin, sunken nasal bridge, thin nose, hypoplasia of the mandible, small mouth, cleft upper lip, palate and uvula; hypoplasia of the teeth.

Stanescu syndrome. Craniofacial dysostosis with diaphyseal hyperplasia.

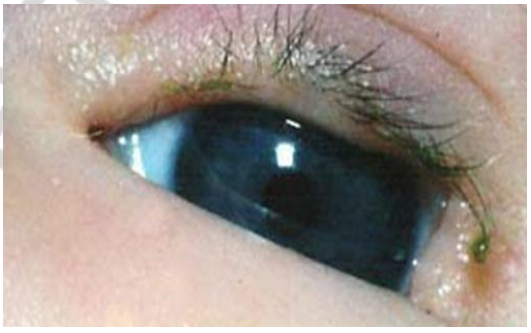
Eye symptoms: exophthalmos due to shallow orbits.

General manifestations: indentation of frontoparietals and occitoparietals seams; hypoplasia of the mandible, flattened palate; small curved teeth with enamel hypoplasia.

Thompson syndrome (2). Mandibular dysostosis. Berry-Frenceschetti syndrome, Frenceschetti- Zwahlen syndrome.

Eye symptoms: antimongoloid eyes, colobomas of lower eyelids in the outer third, rarely — microphthalmia, coloboma of upper eyelids, absence of eyelashes on the lower eyelids; exophthalmos, dermoid cysts of the limbus and conjunctiva, hypoplasia and a complete lack of separate eye muscles; cataracts; colobomas of the iris, choroid and optic nerve head.

General manifestations: gross violations of the formation of the facial skull, which are expressed in bilateral hypoplasia of the zygomatic bones, eye sockets, lower jaw, rarely — upper one; increasing the slit of mouth from one or both sides toward the ear (macrostomia); fistulas from the corner of mouth to ears; sharp hypoplasia of the auditory processes of the temporal bone; gross deformity of the ears with preauricular outgrowths which cover underdeveloped ear canals, resulting in deafness; body hair of hands in form of tongue; developmental disorders of teeth; high palate or its cleft; open bite; the absence of the parotid gland (fig. 23).



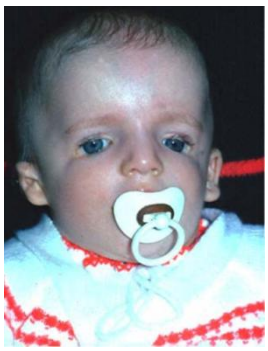


Fig. 23. Thompson syndrome (2)

Francois-Haustrate syndrome. Oto-mandibularis syndrome.

Eye symptoms: congenital iris coloboma, hypoplasia of the optic nerve. Usually both eyes are affected.

General manifestations: aplasia or atresia of the external auditory canal; hypoplasia of the upper jaw; beaked nose («bird face»), the splitting of the upper palate.

Hallermann-Streiff-Francois syndrome. Dysmorphia mandibulo-oculo-facialis.

Eye symptoms: congenital cataracts, microphthalmia; sparse eyelashes and eyebrows, colobomas of eyelids, microcornea, glaucoma.

General manifestations: dyscephalia with hypoplasia of the mandible, «bird face», teeth in newborns, improper growth of teeth, extra teeth, palate in the form of arch (fig. 24).

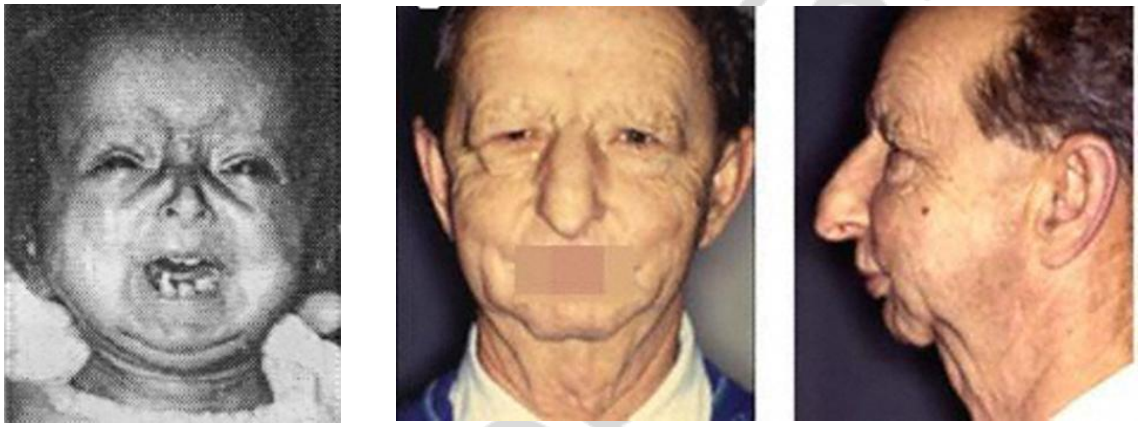


Fig. 24. Hallermann-Streiff- Francois syndrome

Steiner syndrome.

Eye symptoms: pigmentary degeneration of the retina.

General manifestations: congenital unilateral gigantism of separate parts of body, microcephaly, hypodontia. Dysplasia of the tooth enamel. Schizophrenia.

Elsahy-Waters syndrome. Brachio-skeleto-genital syndrome.

Eye symptoms: hypertelorism, ptosis, convergent strabismus, nystagmus.

General manifestations: hypoplasia of the midface, mandibular progna-thism, high palate in form of arch, cleft uvula, funnel chest, scrotal hypospadias, hypoplasia of dentin. X-ray: multiple cysts of the jaws.

Jacobsen-Brodwall syndrome.

Eye symptoms: iridocyclitis and posterior uveitis with progressive deterioration of vision.

General manifestations: dysplasia and decreased concentrating ability of the kidneys; skeletal anomalies; at the age of 9 months all teeth had already erupted, permanent teeth in 3 years.

CLINICAL CASES: THE EYE EXAMINATION

1. A 14-year-old boy is seen for a physical examination at school. He admits difficulty in reading from the blackboard but not in reading textbooks. He does not wear glasses. You record VA as OD 20/100, pinhole 20/25; and OS 20/100, pinhole 20/20. What is your diagnosis? Would you manage or refer this patient?

Answer: 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 the pinhole. Note that visual acuity with use of the pinhole frequently does not reach 20/20. The patient should be referred to an ophthalmologist as a regular patient rather than for an urgent consultation.

2. A 78-year-old woman is seen for an annual physical examination who complains of mild difficulty in reading and in seeing street signs. You record a VA of 20/70 (OD), no improvement with pinhole, and 20/50 (OS), no improvement with pinhole. Upon direct ophthalmoscopy, you note a dullness of the red reflex and you have difficulty seeing fundus details in both eyes. What is your diagnosis? Would you manage or refer this patient?

Answer: Cataract is a common cause of painless progressive loss of vision in older individuals. Her complaints about her visual ability is an indication for referral to an ophthalmologist for evaluation and possible **cataract** surgery.

3. A 40-year-old man is seen for an annual executive physical examination. He has no complaints and does not wear glasses. You record VA as OD 20/15 and OS 20/100, no improvement with pinhole. During examination, it was revealed that the patient had been aware since childhood that his left eye is a so-called lazy eye — in other words, that he suffered from **amblyopia**. Would you refer this patient?

Answer: 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.

4. A 50-year-old man visits your office because he noted decreased visual acuity in the right eye the preceding day while accidentally occluding his left eye. When his present glasses were prescribed 2 years ago, his vision was equal in both eyes. You record VA as OD 20/50, no improvement with pinhole; and OS 20/20. Upon ophthalmoscopy, no abnormalities were detected. What if any, is your diagnosis? Would you manage or refer this patient?

Answer: The patient has an unexplained loss of vision of unknown duration in his right 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).

5. A 55-year-old man, wearing goggles, was sawing wood in his garage shop. He removed the goggles to clean up and, while sweeping up some small wood chips, he had sudden onset of a foreign-body sensation in his right eye. The irritation was not relieved with artificial tears, and it intensified with every blink. His wife rushed him to their family doctor for emergency treatment. The physician was able to examine him after placing a topical anesthetic agent in the right eye. Visual acuity in the right eye was 20/80. Fluorescein staining revealed multiple vertical linear abrasions of the cornea. Explain the clinical findings. What further examination is required, and how is it performed?

Answer: By history, this man has been exposed to small particles that could have abraded his eye. The vertical linear abrasions in conjunction with the feeling of irritation with each blink imply the presence of **a foreign body** under the upper lid. Eversion of the upper lid will expose the foreign body, which can then be removed using a cotton-tipped applicator stick.

THE RED EYE

1. A 23-year-old teacher complains that her right eye is red and irritates. You note moderate injection of the larger conjunctival vessels, watery discharge, and a palpable preauricular lymph node. From this information alone, what tentative diagnosis would you make? Again based on the above information, which of the following symptoms or facts might be elicited by careful history-taking? (sore throat, exposure to children with colds, blurred vision, itching). Which of the following are also the likely findings in this case of viral conjunctivitis.

a. A small pupil in the right eye.

b. Lymphocytes in a smear of conjunctival scrapings.

c. Keratic precipitates.

Management by a primary care physician should consist of which of the following?

a. Corticosteroid.

b. Broad-spectrum antibiotic.

c. Referral to an ophthalmologist.

d. Instruction to the patient to use cool compresses and stay home from school until the redness resolves.

Answer: The conjunctival injection and discharge suggest conjunctivitis. The serous nature of the discharge, plus the preauricular adenopathy, indicate that she has **viral conjunctivitis**.

Sore throat often accompanies viral conjunctivitis; in such cases, a history of exposure to other individuals with upper respiratory tract infections can often be elicited. Blurred vision, a danger signal of serious ocular disease, is not

a feature of simple conjunctivitis. Itching is a symptom of allergic, not viral, conjunctivitis.

b. Lymphocytes are usually found in scrapings from eyes with viral conjunctivitis. A small pupil and keratic precipitates are signs of iritis.

Instruction to the patient should include the use of cool compresses and stay home from school until the redness resolves. Because the disease is contagious, the patient should be instructed to remain home from work. There is no specific medical treatment for viral conjunctivitis. Corticosteroids may be used only under the close supervision of an ophthalmologist and for a short period.

2. A young woman complains of a red eye and associated pain above the eye. The patient's mother has chronic open-angle glaucoma and the patient wants to know whether she has the same thing. The patient describes several bouts of having a red, painful, left eye that is relieved by sleeping. You notice an irregular pupil in addition to the injection of her left eye. This would be more consistent with which of the following?

a. Angle-closure glaucoma.

b. Iridocyclitis.

c. Conjunctivitis.

d. Keratitis sicca.

e. Chronic open-angle glaucoma.

On further questioning, the patient describes seeing colored halos around lights. What is the most likely diagnosis?

You measure the intraocular pressure to be 48 mm Hg. An ophthalmologist is not immediately available. Which of the following treatments would be appropriate?

a. Topical beta-adrenergic blocker.

b. Topical pilocarpine hydrochloride 1 % or 2 %.

c. Systemic carbonic anhydrase inhibitor.

d. Topical cycloplegic drop to relieve the pain.

Answer: a and b. Both **angle-closure glaucoma and iridocyclitis** can have an irregular pupil, pain and redness. Anterior surface problems like conjunctivitis and keratitis sicca (dry eyes) should not result in an irregular pupil. Chronic open-angle glaucoma does not present with a red, inflamed eye. It does not give an irregular pupil.

Angle-closure glaucoma.

a. b, and c. Cycloplegia of the eye helps relieve pain in patients with iridocyclitis by decreasing ciliary spasm, but can make symptoms worsen in angle-closure glaucoma. Cycloplegic drops would work against the pilocarpine trying to constrict the pupil and help lower pressure.

3. A 45-year old man reports a 2-day history of redness, severe pain, and photophobia of his left eye. He denies any trauma to the eye. Which of the following signs convince you the patient does not have conjunctivitis?

a. Visual acuity of 20/200.

b. Conjunctival injection.

c. Ciliary flush.

d. Serous discharge.

You note there is staining of the cornea in a branching pattern. What is the most likely diagnosis? If possible herpetic keratitis, what would your management be? a. A telephone request to an ophthalmologist for immediate examination b. Corticosteroid drops to decrease inflammation and follow up with an ophthalmologist in 3 to 5 days.

Answer: a and c. Reduced visual acuity, as well as ciliary flush, often signals ocular disease more serious than conjunctivitis.

Keratitis, possibly herpetic An oral antiviral like acyclovir and follow up in a week.

a. **Herpetic keratitis** is a serious infection and can be vision threatening. Immediate referral is indicated to decrease the potential scarring and permanent loss of vision. Steroid drops are never indicated in a patient with active disease and epithelial staining. Steroid drops can cause corneal melting and possible perforation of the eye.

4. A 38-year-old woman complains of a 3-day history of a red tender right eyelid. Physical examination reveals a tender nodule of the right lower eyelid with minimal injection of the inferior conjunctiva. Which of the following would constitute appropriate management by the primary care physician? (More than one course of action may be possible.)

a. Hot compresses.

b. Broad-spectrum systemic antibiotics.

c. Topical antibiotics.

d. Immediate surgical incision and drainage to prevent cellulitis.

If the patient reports she has had numerous nodules in this same area over the last 5 years, how should the primary care physician change the management plan?

Answer: a and c. The patient has a **stye**. Because she has only had symptoms for 3 days and the lesion is tender to touch, she would benefit from hot compresses. Topical antibiotic ointment might benefit a small percentage of patients. Incision and drainage is indicated only when lesions do not resolve spontaneously or with medical therapy. Usually surgical intervention occurs only after the lesion has been present for several weeks. Systemic antibiotics are not indicated.

A persistent or recurring lid mass should undergo biopsy to rule out an eyelid malignancy. Referral to an ophthalmologist is indicated.

5. An 88-year-old nursing home patient has had red, irritated eyes for months. She feels like she has «sand in her eyes» all the time. On examination, all four eyelid margins are inflamed and edematous with debris on the lashes. What is the most likely diagnosis? Treatment would consist of which of the following?

a. Immediate referral to an ophthalmologist.

b. Cleansing of the eyelids daily.

c. Antibiotic ointment to alleviate any staphylococcal infection.

Answer: blepharitis.

b and c. Blepharitis is a chronic, often relapsing inflammation of the eyelids that can irritate the eyes. A low-grade bacterial infection may be involved. It is not an ophthalmologic emergency and treatment is long-term daily lid hygiene. For difficult cases, an appointment with an ophthalmologist is indicated.

6. A man returned recently from an African journey. During his trip he had three episodes of blurring and pain in his left eye; each episode lasted about 2 hours and was relieved by sleep. A few hours before consulting you, his symptoms recurred. Which of the following signs convince you that the patient does not have conjunctivitis?

a. Visual acuity of 20/200 in the left eye.

b. Conjunctival injection.

c. Ciliary flush.

d. Absence of exudates.

You note a diffuse haziness of the patient's left cornea. What is the most likely diagnosis? You seek confirmatory data for your tentative diagnosis. What do you expect the following tests to show if your diagnosis is correct?

a. Estimation of anterior chamber depth: deep or shallow?

b. Determination of pupil diameter: large, middilated, or small?

c. Measurement of intraocular pressure: high or low?

Your management should be which of the following?

a. Corticosteroid eyedrops.

b. Advice to see an ophthalmologist the next day.

c. A telephone request to an ophthalmologist for immediate examination.

Answer: a and c. Reduced visual acuity, as well as ciliary flush, often signals ocular disease more serious than conjunctivitis.

Diffuse haziness of the cornea is usually due to edema. This and the history of recurrent attacks relieved by sleep suggest the diagnosis of **acute angle-closure glaucoma**.

In angle-closure glaucoma, the anterior chamber is shallow, the pupil is usually middilated, and the intraocular pressure is high.

c. Angle-closure glaucoma requires emergency treatment to lower the intraocular pressure. The patient should be referred immediately to an ophthalmologist. If an ophthalmologist is not immediately available, you may begin topical pilocarpine hydrochloride 1 % or 2 % a topical beta-adrenergic blocker, and a systemic carbonic anhydrase inhibitor.

7. After working in his garden, a 57-year-old man complains of moderate discomfort and redness in his right eye. You note a visual acuity of 20/25 in the right eye and 20/15 in the left eye. The right eye has mild hyperemia of

the conjunctival vessels; the right cornea appears clear to penlight examination. You diagnose a probable allergy to pollen and advise the patient to use topical dexamethasone sodium phosphate 0.1 % for 3 days. Give two reasons why your diagnosis of allergic conjunctivitis is unlikely to be correct. What other diagnostic techniques should you have performed to be certain that the cornea is normal? Is there any hazard in your prescribed course of treatment? Explain.

Answer: Unless the patient has always had weaker vision in right eye, this finding should alert you to the possibility of a more serious inflammation, such as **keratitis, or glaucoma**. Also, the patient did not complain of itching, which you might expect in an **allergic reaction**.

If the patient has an early herpes simplex keratitis or if his cornea has been scratched by a twig, the epithelial disruption might not be easily seen during a penlight examination. However, it most likely would be rendered visible by fluorescein staining of the cornea.

Yes. The virulence of both herpes simplex and fungal infections, which can result from trauma involving organic material, is markedly potentiated by the application of topical corticosteroids to the eye.

8. A 28-year-old man presents to his optician with a painful, red right eye. The vision has become increasingly blurred over the last 2 days. He is a soft contact lens wearer. The optician notes that the vision is reduced to 6/60 in the right eye, the conjunctiva is inflamed and there is a central opacity on the cornea. A small hypopyon is present. What is the likely diagnosis? What should the optician do?

Answer: It is likely that the man has an **infective corneal ulcer**; he requires immediate referral to an ophthalmic casualty unit. The ulcer will be scraped for culture and the contact lens and any containers cultured. Intensive broad-spectrum antibiotics are administered as an inpatient pending the result of the microbiological investigation.

9. A 27-year-old man presents with a 2-day history of a painful red right eye; the vision is slightly blurred and he dislikes bright lights. He is otherwise fit and well, but complains of some backache. He wears no glasses. What is the likely diagnosis? What would you expect to find on examination of the eye? What treatment would you give? What is the eye condition likely to be associated with?

Answer: The patient has **iritis**. Examination would reveal a reduction in visual acuity, redness of the eye that is worse at the limbus, cells in the anterior chamber and possibly on the cornea (keratic precipitate) or a collection at the bottom of the anterior chamber (hypopyon).The iris may be stuck to the lens (posterior synechiae). There may be inflammation of the vitreous and retina. The patient is treated with steroid eye drops to reduce the inflammation and dilating drops to prevent the formation of posterior synechiae. The history of backache suggests that the patient may have ankylosing spondylitis.

OCULAR AND ORBITAL INJURIES

1. Your neighbour, a 43-year-old woman, is cleaning her swimming pool. While pouring some concentrated algicide into the pool, a large dollop of this solution splashes into her right eye. You are mowing your lawn when you hear her screams. You come to her aid less than 30 seconds after the injury. What are the first aids treatments you will give to her?

Answer: This is one of the few true emergencies of all the ocular injuries that you must know'. Early and copious irrigation with whatever source of water is handy is the right approach to this problem. Even with prompt treatment, serious **ocular injuries** and visual damage may result, depending on the offending **chemical**. Time is of the essence. Do not resort to methods that will cause delay.

2. If you suspect a patient has a perforation of the eye, what signs might you expect to see?

a. Irregular shape to the pupil.
b. Shallow anterior chamber.
c. Low IOP by digital palpation.
d. Uveal tissue prolapsing.
e. Hyphema.

Answer: a, b, d, and e. If you suspect a **perforation of the eye**, digital palpation or any procedure that puts pressure on the eye should not be done.

3. You are on duty in the emergency center when an 18-year-old high school student comes in because of pain, tearing, sensitivity to light, and blurred vision in his right eye. His symptoms began sometime that afternoon. Earlier, he had been working on his car and he remembers something flying into his right eye while he was trying to knock a rivet off the chassis with a hammer and chisel. You examine his eye and take visual acuity measurements. You determine that visual acuity is 20/50 in the right eye and 20/20 in the left eye. There is some conjunctival hyperemia. The pupil of the right eye seems to be peaked and pointing to the 7-o'clock position of the limbus. There is a small, dark, slightly elevated body at the 7-o'clock position of the limbus. You cannot see fundus details of the right eye, but the left eye appears normal. Which of the following would be the appropriate initial management for this situation?

a. Irrigation of the limbal foreign body.
b. Application of a protective shield.
c. Removal of the limbal foreign body using a cotton-tipped applicator.
d. Removal of the limbal foreign body using forceps.
e. A prescription for topical anesthetic (e. g., propacaine 0.5 %) to relieve the patient's symptoms, with strict instructions that he return to see you if his blurred vision continues into the week

Answer: b. A patient whose recent activities involve striking metal on metal should be suspected of having a **foreign body**, even with minimal signs and

symptoms. However, the case illustrated includes a giveaway sign. namely, peaking of the pupil toward the 7-o'clock position. At that position, the dark body is likely to be iris or ciliary body rather than a foreign body. This indicates a penetrating ocular injury and the patient should be protected from further eye trauma by a protective shield. ACT scan will confirm the diagnosis of ocular or orbital foreign body. The patient should be referred urgently to an ophthalmologist.

4. An elderly woman falls and hits her face on the coffee table at home. She had some nose bleeding on that side after the fall. She presents to your office 2 hours later with edema and ecchymosis of the eyelids with numbness of the cheek and teeth on that side. What should be the first priority in the examination? The patient sees 20/20, the pupil is regular, and the eye sustained only a subconjunctival hemorrhage. After appropriate tests, the diagnosis of orbital fracture is made. What would treatment include?

a. lce packs to the orbit.

b. Avoiding blowing the nose.

c. Immediate surgical intervention.

d. Oral antibiotics.

Answer: Carefully open the lids and examine for a ruptured globe. The first step in any ocular or orbital trauma is to assess the status of the eye and avoid any manipulation of the eye until it is found to be intact.

a, b and d. Surgical repair of **orbital fractures** is not an emergency and can be handled over the next 1 to 2 weeks. A referral to an ophthalmologist is indicated in the next few days. Blowing the nose can cause intraorbital emphysema and should be avoided. Ice packs often decrease swelling in an acute event. Because the orbit is now exposed to bacteria in the sinuses, oral antibiotics are warranted.

AMBLYOPIA AND STRABISMUS

1. A 3-year-old girl is brought to you by her mother, who tells you that she suspects her daughter's right eye is not straight. What steps would you take to determine if a significant problem is present?

Answer: Visual acuity testing should be attempted using the rambling 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. First 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. Next. 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 there is a suspicion of intraocular disease, the patient should be referred for an urgent

ophthalmologic evaluation. If visual acuity is reduced in one or both eyes, or if misalignment of the eyes is detected, a non urgent referral should be made.

2. A 54-year-old woman has early cataracts in both eyes. With glasses, the right eye cannot be corrected to better than 20/200, whereas with the left eye she can read the 20/40 line with best correction. The amount of cataract is exactly the same in each eye. Examination of the optic disc and macula, pupillary reaction, color vision, and retinal blood vessels proved entirely normal in each eye. However, the right eye appears to be turned slightly inward when you evaluate the corneal light reflex, and the patient has not experienced diplopia. Additional questioning reveals that the patient wore a patch over one eye as a child. Why would information concerning her childhood ocular condition be relevant in this situation?

Answer: The poor vision in the right eye may be due to longstanding **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. This information is valuable in helping to determine if cataract surgery on the right eye is likely to be helpful to the patient.

3. A 4-year-old girl is brought to your office by her mother, who says that she sees her daughter's right eye «drifting». You test the patient's vision, and it is 20/20 in each eye. There is no epicanthus. The cornea light test shows no deviation, and the cover test fails to reveal strabismus. What is your next step?

Answer: Strabismus can be **intermittent**. Intermittent esodeviations are usually early manifestations of a constant deviation and may be difficult to detect during the early stages. Intermittent exodeviations are more pronounced when the patient is tired or sick but can be easily missed. If the patient has a reliable history of strabismus but you are unable to detect the deviation, referral to an ophthalmologist is recommended.

4. A 60-year-old man comes to see you because of the sudden onset of double vision. He states that when he looks straight ahead, he does not have diplopia, but on right gaze he has diplopia, with the two images horizontally displaced. He has no other abnormal neurologic symptoms. His past medical history is significant for diabetes mellitus. How would you assess this patient?

Answer: First you would check the visual acuity in each eye, and the pupillary light reflexes. Observe the corneal light reflex, and perform a cover test. It will be important to observe the patient's eye movements in all the cardinal fields of gaze. He may have an **incomitant strabismus** that can be seen in only some fields of gaze. If an incomitant strabismus is detected, he may have cranial nerve palsy. Ophthalmologic and neurologic consultation should be obtained urgently.

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