Ministry of Education and Science of Ukraine
Sumy State University

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PRACTICAL OPHTHALMOLOGY

Study guide

Recommended by the Academic Council of Sumy State University

Sumy
Sumy State University
2019

УДК 617.7(075.8)
L51

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Recommended for publication
by the Academic Council of Sumy State University
as a study guide
(minutes № 5 of 10.11.2016)

Lekishvili S. E.
L51 Practical Ophthalmology : study guide / S. E. Lekishvili. – Sumy : Sumy State University, 2019. – 392 p.
ISBN 978-966-657-763-7

The study guide is intended to train students of higher medical educational institutions of the fourth level of accreditation on the specialty "Medicine", interns, residents and masters.

The guide is a new progressive step in teaching the discipline "Ophthalmology".

УДК 617.7(075.8)

ISBN 978-966-657-763-7

CONTENTS

LIST OF ABBREVIATIONS

ADED – advanced diabetic eye disease
AIDS – acquired immune deficiency syndrome
AION – anterior ischaemic optic neuropathy
AK – allergic conjunctivitis
AMPPE – acute multifocal placoid pigment epitheliopathy
ARMD – age-related macular degeneration
ARN – acute retinal necrosis
bid – bis in die
BRVO – branch retinal vein occlusion
BRVO – branch retinal vein occlusion
BUT – tear film break-up (break-up time)
CFF – critical fusion frequency flashing
CLV – corrected loss variance – reducing variability in light sensitivity magnitudes
CME – cystoid macular edema
CME – cystoid macular edema
CMV – cytomegalovirus
COD – congestive optic disc
CRVO – central retinal vein occlusion
CRVO – occlusion of the central retinal vein and its branches
CSME – clinically significant macular edema
CSR – central serous retinopathy
D – Dioptre
DES – dry eye syndrome
DR – diabetic retinopathy
DR – diabetic retinopathy
ELISA – enzyme immunoassay
EOG – electro-oculogram
ERG – electroretinogram
ERG – electro-retinogram
GC – glucocorticoids
ICE – iridocorneal endothelial syndrome

ICSOLs – intracranial space-occupying lesions
IF – intraocular fluid
IIH – idiopathic intracranial hypertension
IOFB – intraocular foreign bodies
IOL – intraocular lens
IOP – intraocular pressure
IOP – intraocular pressure
IRMA – intraretinal microvascular abnormalities
KCS – keratoconjunctivitis sicca
LASEK – laser assisted sub-epithelium keratomileusis
LF – lacrimal film
LGN – lateral geniculate nucleus
LP – lumbar puncture
LTK – laser thermal keratoplasty
LV – loss variance (reducing variability index sensitivity)
LVA – low vision aids
MD – mean defect (the average depth of the defect)
MFA – fluorescent antibody method
MRI – magnetic resonance imaging scan
MS – mean sensitivity (mean retinal sensitivity)
NLD – nasolacrimal duct
NPDR – non-proliferative diabetic retinopathy
NSAIDs – nonsteroid anti-inflammatory drugs
NVG – neovascular glaucoma
NVM – neovascular membrane
OA – ophthalmic artery
OCRA – occlusion of the central retinal artery
OCT – optical coherence tomography
OD – right eye
OKT – optical coherence tomography
OS – left eye
PCG – primary congenital glaucoma
PCR – polymerase chain reaction
PDR – proliferative diabetic retinopathy

PDT – photodynamic therapy
PKC – protein kinase C
PORN – progressive outer retinal necrosis
PRK – photorefractive keratectomy
PRP – panretinal photocoagulation
qid – quater in die
RAPD – relative afferent pupillary defect
RF – reliability factor (considers the degree of probability study)
ROP – retinopathy of prematurity
RPE – retinal pigment epithelium
stat – immediately
STM – superior tarsal muscle
TIOP – tolerant intraocular pressure
TTT – transpupillary thermotherapy
UBM – ultrasound biomicroscopy
VECP – visually evoked cortical potential
VECP – visually evoked cortical potential
VEGF – vascular endothelial growth factors
VEP – visually evoked potential
VER – visually evoked response
VER – visually evoked response
Visus – visual acuity

TOPIC 1. OCULAR ANATOMY AND PHYSIOLOGY

The ability to see is dependent on the actions of several structures in and around the eyeball. The graphic below lists many of the essential components of the eye's optical system.

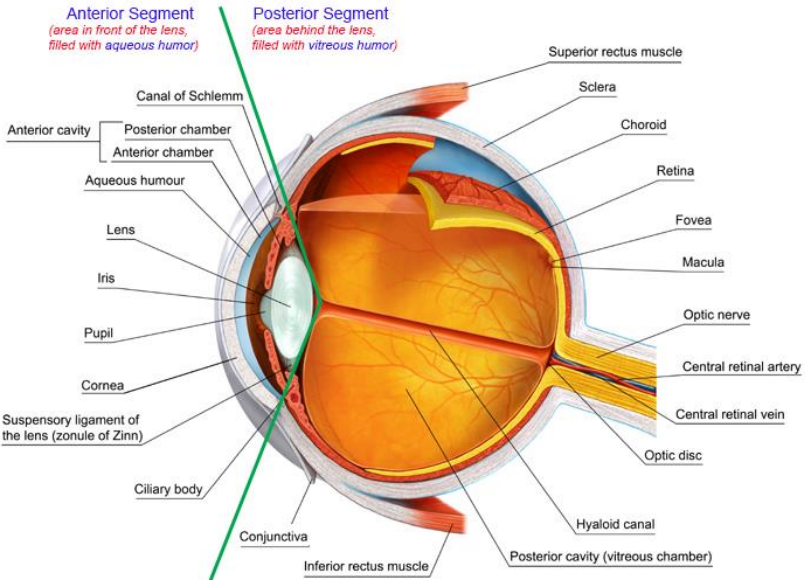

Figure 1 – Essential components of the eye's optical system [12]

When you look at an object, light rays are reflected from the object to the cornea, which is where the miracle begins. The light rays are bent, refracted and focused by the cornea, lens, and vitreous. The lens' job is to make sure the rays come to a sharp focus on the retina. The resulting image on the retina is upside-down. Here at the retina, the light rays are converted to electrical impulses which are then transmitted through the optic nerve, to the brain, where the image is translated and perceived in an upright position.

The eye (Lat. – *oculus*; Greek – *ophthalmos*) is the organ that allows us to see. It is situated in the eye socket.

An eye socket, or an orbit, is a bony cup in the skull that contains the eyeball with its auxiliary apparatus (vessels, nerves, muscles, fat, fascia, tear glands, connective membrane and lacrimal passages). The eyeball itself is a sphere spanning approximately 24 mm in diameter. The depth of the orbit in adults is 4 cm, width of the entrance to the orbit – 4 cm, height – 3.5 cm. The walls of the orbit are as follows:

– superior wall (a roof);

– inferior wall (a floor);

– medial wall;

– lateral wall.

How each of them is formed?

1. The superior wall is formed by the frontal part of the orbital bone, and the lesser wing of the sphenoid bone.

2. The inferior orbital wall is formed by a part of the orbital surface of the maxilla, zygomatic bone, orbital process of palatine bone.

3. The medial wall is formed by the frontal offshoot of the upper jaw, lacrimal bone, orbital plate lattice bones, body sphenoid bone and frontal part of the orbital bone.

4. The lateral orbital surface is formed by the greater wings of the sphenoid bone, zygomatic bone.

The orbit borders with the outer wall of the nasal cavity, namely the ethmoid labyrinth; at the top – the orbital part of the anterior cranial fossa, which houses the frontal lobes of the brain, as well as the frontal sinus of the frontal bone. Outside – the borders of the temporal fossa, at the bottom – from the top wall of the maxillary sinus of the upper jaw.

An eyeball consists of three layers and an internal optic core. There are three covers:

– external (fibrous membrane);

– middle (choroid);
– internal (retina).

The outer layer is placed around the outside of the eye, plays a mechanical role, which is protective and is the mainstay of the eye. There are two parts of the outer layer:

– anterior (cornea);
– posterior (sclera).

The eyes move symmetrically (in the same direction at the same time). These symmetrical movements are made possible through the coordination of the *extraocular muscles* (muscles outside the eye). Around the eyes there are three pairs of eye muscles. One pair of eyes turns left and right, the second – up and down, and the third pair rotates it relative to the optical axis.

Since the eyes are paired structures, the brain receives two slightly different images that overlap with one another. Interpretation of the different images is possible via coordinated eye movements achieved by complex neural mechanisms. Humans are also able to perceive three-dimensional images because they possess binocular vision, which enables the perception of depth and distance.

Conjunctiva is a mucous membrane covering the under surface of the lids and anterior part of the eyeball up to the cornea.

Parts of conjunctiva:

– palpebral (covering the lids – firmly adherent);
– forniceal (covering the fornices – loose – thrown into folds);
– bulbar (covering the eyeball – loosely attached except at limbus);
– marginal and limbal parts, and plica semilunaris.

Nerve supply

Sensory innervation:

– bulbar conjunctiva – long ciliary nerves – nasociliary n. – an ophthalmic division of the trigeminal n.;

– superior palpebral and forniceal conjunctiva – frontal and lacrimal branches of Ophthalmic division of the trigeminal n.;

– inferior palpebral and forniceal conjunctiva – laterally from lacrimal branches of the ophthalmic division of the trigeminal n. and medially to the infraorbital n. – maxillary n.;

– a division of the trigeminal n.

Sympathetic innervation: superior cervical sympathetic to blood vessels.

Blood supply

Arterial supply:

– posterior conjunctival arteries are derived from the arterial arcade of eyelids which is formed by palpebral branches of nasal and lacrimal arteries of the lids;

– anterior conjunctival arteries are derived from the anterior ciliary arteries to the muscular branch of the ophthalmic artery to rectus muscles.

Venous drainage: palpebral and ophthalmic veins.

Lymphatic drainage:

– lymph vessels are arranged as superficial and deep plexuses in the submucosa;

– ultimately as in the lids to the preauricular and sub-mandibular lymph glands.

Cornea. The transparent cornea appears from the front to be oval, as the sclera encroaches on the superior and inferior aspects. The anterior horizontal diameter is 12 mm, and the anterior vertical diameter is 11 mm.

Histological structure

The cornea is the principal refracting component of the eye. Its transparency and avascularity provide optimal light transmittance.

The anterior surface of the cornea is covered by the tear film, and the posterior surface borders the aqueous-filled anterior chamber. At its periphery, the cornea is continuous with the conjunctiva and the sclera. From anterior to posterior, the five layers that compose the cornea are:

1) epithelial layer;
2) Bowman's membrane;
3) stroma;
4) Descemet's membrane;
5) endothelium.

Corneal function

The cornea has two primary functions: to refract light and to transmit light. Factors that affect the amount of corneal refraction include:

1) the curvature of the anterior corneal surface;
2) the change in refractive index from air to cornea;
3) corneal thickness;
4) the curvature of the posterior corneal surface;
5) the change in refractive index from cornea to aqueous humor.

The total refractive power of the eye focused at infinity is between 60 and 65 diopters (D), with 43 to 48 D attributable to the cornea.

The cornea has no blood vessels of its own, it is powered through pericornealis vascular net, anterior chamber moisture and tissue respiration (oxygen from air penetrates through the epithelium towards the anterior chamber, and carbon dioxide – from the front of the camera).

Corneal innervation

The cornea is densely innervated with sensory fibers. From 70 to 80 large nerves, branches of the long and short ciliary nerves, enter the peripheral stroma. Approximately 2 to 3 mm after the nerves pass into the cornea, they lose their myelin sheath, but the covering from the Schwann cell remains.

Corneal blood supply

The cornea is avascular and obtains its nourishment by diffusion from the aqueous humor and from the conjunctival and episcleral capillary networks located in the limbus.

Sclera. The sclera forms the posterior five-sixths of the connective tissue coat of the globe. The sclera maintains the shape of the globe, offering resistance to internal and external forces, and provides an attachment for the extraocular muscle insertions. The thickness of the sclera varies from 1 mm at the posterior pole to 0.3 mm just behind the rectus muscle insertions.

Episclera. The episclera is a loose vascularized, connective tissue layer that lies just outer to the sclera.

Functions of the sclera:

1) maintains the shape of the globe;
2) stable positioning for structures resting against it, such as the choroid and retina;
3) attachment for extraocular muscle insertions.

Aqueous humor is transparent, watery fluid similar to plasma, but containing low protein concentrations, produced in the eye and filling the spaces (anterior chamber and posterior chamber) in front of the lens and its attachments. It diffuses out of the eye into the blood and is regarded as the lymph of the

eye, although its composition is different from that of the lymph in the rest of the body.

This fluid is more acidic than plasma; it contains more chlorides, lactic and ascorbic acids, hyaluronic acid, Na, K, urea, and glucose. Proteins do not exceed 0.02 %; density of aqueous humor is 1.005 g/ml.

Iris (Lat. *Iris*) – front choroid, which is viewed through the transparent cornea. Iris is the coloured ring in the front part of the eye. It is a thin layer of fine muscles. In its centre there is a hole, the *pupil*. Light passes through the pupil into the eye. The pupil becomes small (constricts) in bright light and larger (dilates) in dim light.Thus, the body serves as the aperture of the optical system of the eye.

Just behind the iris there is the ciliary body. The lens is attached to the ciliary body by very thin thread-like structures. The surface of the ciliary body secretes clear fluid that circulates through the pupil into the anterior chamber and out of the anterior chamber via chamber angle. The anterior chamber angle is the place where the iris and the cornea meet.

The front surface of the iris faces the cornea, the back – faces the lens.

The outer edge of the iris called ciliary margin (*margo ciliaris iridis*) is a continuation of the ciliary body in the transition cornea sclera (the limb). The inner edge of the iris is called pupillary margin (*margo pupillaris iridis*).

There are blood vessels in the iris stroma. Cells at the back of the stromal enriched pigment determine the colour of the iris (eye colour). With the large amount of pigment, the iris colour can be brown, gray or even black. If there is little melanin pigment, the iris will be light gray or light blue. If there is no melanin in pigment (as in albinos), the iris is red because of the translucent blood vessels.

There are two antagonistic muscles in the iris: iris sphincter muscle and iris dilator muscle. They regulate the size of the pupil according to the amount of light falling on the retina.

Iris sphincter muscle (Lat. *Musculus sphincter pupillae*) receives parasympathetic innervation from the oculomotor nerve (the 3rd pair of cranial nerves). Located near the pupillary edge of the iris, it has a circular shape. In medical practice it is called "miosis" and refers to constriction of the pupil, "mydriasis" – to expansion.

Iris sphincter muscle is located in the back of the iris around the pupil.

Iris dilator muscle (Lat. *Musculus dilatator pupilae*) receives a sympathetic innervation from sympathetic branches. It has a fan-shaped form, located at the edge of the iris ciliary.

Both muscles are composed of smooth muscle fibers.

The ciliary body (Lat. *Corpus ciliare*) is a closed circle, located between the iris and sclera, being actually a choroid. Ciliary body is not visible because it is hidden behind the iris. In the meridional section, the ciliary body looks like a triangle. Place of transition of choroid and ciliary body coincides with the transition point in the visual part of the retina called *ora serrata* (the blind and notched).

The ciliary body is a ring-shaped thickening of tissue inside the eye that divides the posterior chamber from the vitreous body. It contains the ciliary muscle, vessels, and fibrous connective tissue. Folds on the inner ciliary epithelium are called ciliary processes, which secrete aqueous humor into the posterior chamber. The aqueous humor then flows through the pupil into the anterior chamber.

The ciliary body is attached to the lens by connective tissue called the zonular fibers (fibers of Zinn). Relaxation of

the ciliary muscle puts tension on these fibers and changes the shape of the lens in order to focus light on the retina.

The inner layer is transparent and covers the vitreous body, and is continuous from the neural tissue of the retina. The outer layer is highly pigmented, continuous with the retinal pigment epithelium, and constitutes the cells of the dilator muscle. This double membrane is often considered continuous with the retina and a rudiment of the embryological correspondent to the retina. The inner layer is unpigmented until it reaches the iris, where it takes on pigment. The retina ends at the ora serrata.

The sensory innervation of the ciliary body is from one of the terminal branches of the ophthalmic division of the trigeminal nerve (*nervus trigeminus*), the nasociliary nerve (*nervus nasociliaris*). The ciliary muscle is parasympathetically innervated. The nerve fibres course in the ophthalmic nerve to the ciliary ganglion and from there to the ciliary body along with the short ciliary nerves (*nervi ciliares breves*). The ciliary body is also known to receive sympathetic innervation via long ciliary nerves.

From the muscle of the ciliary body, there are departing thin fibers, zonula ciliaris, which hold the lens. Ciliary muscle contracts and relaxes, which changes the tension and focus according to the curvature of the lens. When the ciliary muscle contracts, the lens becomes more convex, generally improving the focus for closer objects. When it relaxes, it flattens the lens, generally improving the focus for farther objects. This process is called accommodation and gives us the opportunity to see objects at the long and short distance. With age, usually after 40 years old, this property is lost and people need glasses for reading.

The vessels of the ciliary body produce intraocular fluid of the same chemical compound that circulates within the front rear camera and the eye cavity and provides nutrients to the

anterior segment of the eye. Intraocular fluid creates the intraocular pressure; the violation of its products or outflow is the cause of glaucoma.

Pain caused by spasm of the ciliary body may be conditional inflammation of the anterior eye. Inflammation of the ciliary body is called uveitis.

While a significant reduction in secretion of aqueous humor in the ciliary body decreases, the intraocular pressure and atrophy of the eyeball occur.

Choroid (choroidea) is a thin layer of vessels behind the retina; it starts from the dentate line and passes to the opening of the optic nerve, where it is firmly connected with the sclera and loosely attached to the field near the equator entrances to the choroid vessels and nerves. The rest area, adjacent to the sclera, is limited by its supra choroid space (it ends up 3 mm from the limbus in front and near the exit of the optic nerve behind; belongs to the lymph). The outer layers of the retina get their nutrition from the choroidal circulation. There are blood vessels and nerves; there is outflow of ocular fluid out of the eye.

Choroid consists of 5 layers, the basis of which is a thin connective tissue stroma of elastic fibers.

Suprachoroid lamina of choroid is situated above the choroid; applied to the layer of the choroid coat of the eyeball next to the sclerotic one. It is an exceedingly delicate layer of loose pigmented connective tissue on the outer surface of the choroids; recently, the suprachoroid laminae of the sclera and choroid have been considered to be a single suprachoroid layer.

By the middle of it there is a layer of large veins, which is the beginning of the uvea. The number of pigment in this layer determines the colour of fundus. The next layer of medium vessels (arteries and veins) has less pigment choroidea.

Choroid capillary layer is separated from the outer layers of the retina vitreous thin plate (membrana elastica), which is firmly connected with the cell pigment epithelium of the retina.

Vascular System

Arteries. The arterial input to the eye is provided by several branches from the ophthalmic artery, which is derived from the internal carotid artery. These branches include the central retinal artery, the short and long posterior ciliary arteries, and the anterior ciliary arteries. The *ophthalmic artery* is the first branch of the internal carotid artery distal to the cavernous sinus. Branches of the ophthalmic artery supply all the structures in the orbit as well as some structures in the nose, face and meninges.

Two long posterior ciliary arteries (a.ciliaris posterior longes) obliquely pierce the sclera at the posterior pole of the eye on both sides of the optic nerve and get into the supra choroidea space. Near the front end of the ciliary muscle the artery is divided into two branches, which are concentric to the limbus and anastomosing branches of the other. At the front, ciliary arteries form the root of the iris – *a major arterial circle*, the *circulus arteriosus major*, around the circumference of the iris, from which numerous converging branches run, in the substance of the iris, to its pupillary margin, where they form a second (incomplete) arterial circle, the *circulus arteriosus minor*.

Short posterior ciliary arteries (Lat. – *aa. Ciliares posterior brevis*), penetrating the sclera near the posterior pole, enter the choroid and form its vascular net. Certain meridian branches with anastomosis of the ciliary body vessels penetrate the ciliary body. At the posterior pole of the eye, the arteries make anastomosis between themselves and the branches of the central retinal artery, forming a crown around the optic nerve,

the Circle of Zinn–Haller or Circle of Zinn, which is associated with the fibrous extension of the ocular tendons (Annulus of Zinn), and feeding the branch of the nerve closest to the eye. Some branches of the short posterior ciliary arteries also supply the optic disc via an anastomotic ring.

In the distal parts of its blood supply there are front and rear branches *a. n. optici* (*branches of a. ophthalmica*). In some cases, there are anastomoses between the posterior ciliary arteries and the short central retina in a branch to the optic nerve (*a. Opticociliaris*) or near the retina nerve (*a. Cilioretinalis*).

These anastomoses are needed for disorders in blood supply of the central retinal artery or its branches, as this vision is kept by the power of the collateral central parts of the retina.

Posterior ciliary arteries perforate the sclera posteriorly in the vicinity of the optic nerve and macula to supply the posterior uveal tract. The posterior ciliary arteries arise directly from the ophthalmic artery and are end arteries, which is to say no posterior ciliary artery or any of its branches anastomose with any other artery. The ophthalmic artery continues medially the superior and inferior muscular branches to supply the extraocular muscles. The extraocular muscles are the six muscles that control movement of the eye and one muscle that controls eyelid elevation (*levator palpebrae*). The actions of the six muscles responsible for eye movement depend on the position of the eye at the time of muscle contraction.

A *vascular plexus* is found between the bulbar conjunctiva and the sclera consisting of two layers of vessels, the superficial episcleral vessels and the deep episcleral vessels. Blood to the bulbar conjunctiva is primarily derived from the ophthalmic artery. The blood supply to the palpebral conjunctiva (the eyelid) is derived from the external carotid artery. However, the circulations of the bulbar conjunctiva and palpebral conjunctiva are linked, so both bulbar conjunctival

and palpebral conjunctival vessels are supplied by both the ophthalmic artery and the external carotid artery, to varying extents.

Veins. The venous outflow from the eye is primarily via the vortex veins and the central retinal vein, which merge with the superior and inferior ophthalmic veins that drain into the cavernous sinus, the pterygoid venous plexus and the facial vein. Most of the venous drainage from the anterior segment is directed posteriorly into the choroid and then into the vortex veins. The choriocapillaris lobules drain into venules that join the larger venules of the outer conduit layer that coalesce into the 4–5 vortex veins that pierce the sclera at the equator.

The vascular supply of the optic nerve is complex. The optic nerve has three zones referenced to the lamina cribosa, the connective tissue extension of the sclera through which the optic nerve axons and the central retinal artery and vein pass. The prelaminar (i. e., inside the eye relative to the lamina cribosa) optic nerve is supplied by collaterals from the choroid and retina circulations. The laminar zone is supplied by branches from the short posterior ciliary and pial arteries. The post laminar zone is supplied by the pial arteries. Venous drainage is via the central retinal vein and pial veins.

Sensory and motor innervation of the ocular circulations is restricted to the vessels of the uvea (i. e., the choroid, ciliary body and iris) and optic nerve; the retina appears to lack sympathetic and parasympathetic nerves. The postganglionic sympathetic nerves originate in the superior cervical ganglion. Parasympathetic innervation originates in the pterygopalatine ganglion via the facial nerve.

Lens. The lens is a transparent, biconvex structure in the eye that, along with the cornea, helps to refract light to

be focused on the retina. The lens, by changing its shape, functions to change the focal distance of the eye so that it can focus on objects at various distances, thus allowing a sharp real image of the object of interest to be formed on the retina. This adjustment of the lens is known as accommodation. The lens is more flat on its anterior side than on its posterior side.

The lens is a part of the anterior segment of the human eye. The iris, which regulates the amount of light entering into the eye, is in front of the lens. The lens is suspended in place by the suspensory ligament of the lens, a ring of fibrous tissue that attaches to the lens at its equator and connects it to the ciliary body. Posterior to the lens there is the vitreous body, which, along with the aqueous humor on the anterior surface, bathes the lens. The lens has an ellipsoid, biconvex shape. The anterior surface is less curved than the posterior. The size and shape can change due to accommodation and because the lens continues to grow throughout a person's lifetime.

The lens has three main parts: the lens capsule, the lens epithelium, and the lens fibers. The lens capsule forms the outermost layer of the lens and the lens fibers form the bulk of the interior of the lens. The cells of the lens epithelium, located between the lens capsule and the outermost layer of lens fibers, are found only on the anterior side of the lens. The lens itself lacks nerves, blood vessels, or connective tissue.

There are 5 main functions of the lens:

1. Accommodation.
2. Crystalline and transparency.
3. The protective function.
4. Resolution.
5. Nourishment.

Histology is composed of 6 layers from anterior to posterior:

1. Anterior capsule: basement membrane of the anterior epithelium.

2. Anterior epithelium.
3. Anterior cortex.
4. Central nucleus.
5. Posterior cortex.
6. Posterior capsule.

Innervation and blood supply: the lens has no blood and lymph vessels and nerves. Exchange processes are carried out by intraocular liquid of the lens.

The **vitreous chamber** is positioned at the back of the eyeball. It is the largest of the chambers and takes up around 80 % of the eye. It is bounded on the front by the posterior surface of the lens and the retro-zonular portion of the posterior chamber. Peripherally and posteriorly, it is bounded by the pars plana of the ciliary body, the retina, and the optic disc.

The vitreous chamber is filled with the gelly-like vitreous body; 99 % of it consists of water and the rest is a mixture of collagen, proteins, salts and sugars. Despite the water-to-collagen ratio, the vitreous has a firm jelly-like consistency.

The vitreous performs a vital role in protecting the eye. Most importantly, it helps to hold its 'spherical' shape. The vitreous also comes in contact with the retina (the light-sensitive tissue at the back of the eye that acts like the film of a camera). The pressure of the vitreous humour helps to keep the retina in place.

All surfaces that interface with the vitreous are basement membranes. The centre of the anterior surface contains the patellar fossa, an indentation in which the lens sits.

Vitreous zones

The vitreous can be divided into zones that differ in relative density. The outermost zone is the vitreous cortex, the centre zone is occupied by Cloquet's canal, and the

intermediate zone is inner to the cortex and surrounds the centre canal.

Cloquet's canal, also called the hyaloid channel or the retrolental tract, is located in the centre of the vitreous body.

The vitreous cortex, also called the hyaloid surface, is the outer zone.

The intermediate zone contains fine fibers that are continuous and unbranched and that run anteroposterior.

Vitreous body functions:

1) providing the correct form of the eyeball;
2) refracting the light which falls on the retina (the light refractive function);
3) ensuring turgor of tissues;
4) ensuring the elasticity of the eye.

With age, the vitreous body may undergo degradation and becomes flaky. This often happens in myopic eyes (myopia) which may be due to the trauma or inflammation of the eye (uveitis).

Inflammation of the vitreous body is called vitraitis. Practically there is no separate disease, but the propagation of inflammation of the choroid of the eye (uveitis) and retina (retinitis) can develop.

Retina. The innermost neural layer of the eye, the retina, is located between the choroid and the vitreous. It includes the macula, the area at the posterior pole used for the sharpest acuity and colour vision. The retina extends from the circular edge of the optic disc, where the nerve fibers exit the eye, to the ora serrata. It is continuous with the epithelial layers of the ciliary body, with which it shares embryologic origin.

Retina is developed from the two walls of the optic cup, namely: nervous retina from the inner wall, and pigment epithelium from the outer wall.

Nervous retina: the inner wall of the optic cup is a single-layered epithelium. It divides into several layers of cells which differentiate into the following three layers (as also occurs in neural tube):

1) *matrix cell layer:*cells of this layer form the rods and cones;

2) *mantle layer:* cells of this layer form the bipolar cells, ganglion cells, other neurons of retina and the supporting tissue;

3) *marginal layer:* this layer forms the ganglion cells, axons of which form the nerve fibre layer.

Outer pigment epithelial layer: cells of the outer wall of the optic cup become pigmented. Its posterior part forms the pigmented epithelium of retina and the anterior part continues forward in the ciliary body and iris as their anterior pigmented epithelium.

Regions of retina

The retina is often described as consisting of two regions: peripheral and central. The peripheral retina is designed for detecting gross form and motion, whereas the central area is specialized for visual acuity. In area, the periphery makes up most of the retina, and rods dominate. The central retina is rich in cones, has more ganglion cells per area than elsewhere, and is a relatively small portion of the entire retina.

Central retina (*macula lutea*) – appears as a darkened region in the central retina and may seem to have a yellow hue because of the xanthophyll pigments, lutein, and zeaxanthin. These pigments are located throughout the retina, but the greatest concentration is in the macula.

The pigments are primarily located in the photoreceptor inner fibers but are also found in the rod outer segments.The newborn has little of any of these pigments, but they gradually accumulate from dietary sources.

Fovea (*fovea centralis*), or central fovea of the retina (fovea centralis retinae), is the shallow depression in the centre of the macular region. This depression is formed because the retinal neurons are displaced, leaving only photoreceptors in the centre. The fovea has a horizontal diameter of approximately 1.5 mm. The curved wall of the depression is known as the *clivus*, which gradually slopes to the floor, the *foveola*. The only photoreceptors located in the centre of the fovea are cones.

The ora serrata is the peripheral termination of the retina and lies approximately 5 mm anterior to the equator of the eye. Its name derives from the scalloped pattern of bays and dentate processes; the retina extends further anteriorly on the medial side of the eye. The ora serrata is approximately 2 mm wide and is the site of transition from the complex, multilayered neural retina to the single, nonpigmented layer of ciliary epithelium. A firm attachment between the retina and vitreous, the vitreous base, extends several millimeters posterior to the ora serrata.

The optic disc, or *an optic nerve head*, is the site where ganglion cell axons accumulate and exit the eye. It is slightly elongated vertically. The optic disc lacks all retinal elements except the nerve fiber layer and an internal limiting membrane. It is paler than the surrounding retina because there is no RPE (retinal pigment epithelium). Because the disc contains no photoreceptor cells, light incident on the disc does not elicit a response; thus, it represents the physiologic blind spot.

Retinal histologic features

Under light microscopy, the retina has a laminar appearance in which 10 layers are evident:

1. Retinal pigment epithelial layer.
2. Photoreceptor layer.
3. External limiting membrane.
4. Outer nuclear layer.

5. Outer plexiform layer.
6. Inner nuclear layer.
7. Inner plexiform layer.
8. Ganglioncell layer.
9. Nerve fiber layer.
10. Internal limiting membrane.

Retinal function

Light passes through most of the retinal layers before reaching and stimulating the photoreceptor outer segment discs. The neural flow then proceeds back through the retinal elements in the opposite direction of the incident light. The efficient and accurate performance of the retina is not hampered by this seemingly reversed situation.

Retinal blood supply

The outer retinal layers receive nutrition from the choroidal capillary bed; metabolites diffuse through the Bruch's membrane and the RPE into neural retina. The central retinal artery provides nutrients to inner retinal layers.

The artery enters the retina through the optic disc, usually slightly nasal of centre, and branches into a superior and inferior retinal artery, each of which divides further into nasal and temporal branches.

Optic nerve (**cranial nerve II**, or **CN II**) is a paired nerve that transmits visual information from the retina to the brain. It is derived from optic stalks during the seventh week of development and is composed of retinal ganglion cell axons and glial cells; it extends from the optic disc to the optic chiasma and continues as the optic tract to the lateral geniculate nucleus, pretectal nuclei, and superior colliculus.

The optic nerve transmits all visual information including brightness perception, colour perception and contrast (visual acuity). It also conducts the visual impulses that are responsible for two important neurological reflexes: the light

reflex and the accommodation reflex. The light reflex refers to the constriction of both pupils that occurs when light is shone into either eye; the accommodation reflex refers to the swelling of the lens of eye that occurs when one looks at a near object as in reading (lens adjusts to near vision).

The eye's blind spot is a result of the absence of photoreceptors in the area of the retina where the optic nerve leaves the eye.

The anatomical course of the optic nerve describes the transmission of special sensory information from the retina of the eye to the primary visual cortex of the brain. It can be divided into extracranial (outside the cranial cavity), and intracranial (the visual pathway) parts.

Extracranial – the optic nerve is formed by the convergence of axons from the retinal ganglion cells. These cells in turn receive impulses from the photoreceptors of the eye (the rods and cones). After its formation, the nerve leaves the bony orbit via the optic canal, a passageway through the sphenoid bone. It enters the cranial cavity, running along the surface of the middle cranial fossa (in close proximity to the pituitary gland).

Intracranial are the nasal retinal fibers crossing over at the optic chiasm. Within the middle cranial fossa, the optic nerves from each eye unite to form the optic chiasm. At the chiasm, fibers from the nasal (medial) half of each retina cross over, forming the optic tracts:

– left optic tract – contains fibers from the left temporal (lateral) retina, and the right nasal (medial) retina;

– right optic tract – contains fibers from the right temporal retina, and the left nasal retina.

Each optic tract travels to its corresponding cerebral hemisphere to reach the lateral geniculate nucleus (LGN), a relay system located in the thalamus; the fibers synapse here.

Axons from the LGN then carry visual information via a pathway known as the *optic radiation*. The pathway itself can be divided into:

– *upper optic radiation* – carries fibers from the superior retinal quadrants (corresponding to the inferior visual field quadrants). It travels through the parietal lobe to reach the visual cortex;

– *lower optic radiation* – carries fibers from the inferior retinal quadrants (corresponding to the superior visual field quadrants). It travels through the temporal lobe, via a pathway known as Meyers' loop, to reach the visual cortex.

Once at the visual cortex, the brain processes the sensory data and responds appropriately.

Topographically the optic nerve in its length is divided into four sections:

1. Intrabulbar department (within the eyeball to exit the sclera).
2. Retrobulbar/intraorbital division (limited sclera front, rear orbital aperture optic channel).
3. Intracanalicular department (inside the bone canal).
4. Intracranial section (from point of entry into the optic nerve to the cranial cavity chiasmi).

Full damage of the optic nerve having traumatic, ischemic, inflammatory, or other etiology leads to the loss of vision in the eye (*amaurosis*), accompanied by rectal prolapse (because of interrupted afferent reflex arc).

Reduced vision that occurs due to the damage of the optic nerve is called *amblyopia*.

Partial optic nerve damage is accompanied by constriction of the visual field loss or its individual sections (*scotoma*). Optic nerve pathology of the fundus is observed as primary atrophy. Note that the reverse image or the intersection of the eye's environment (lens, vitreous body) projected onto the retina, the reverse image is seen as objects in the right half

of the visual field, as well as perceived by the left half of the retina and vice versa.

Features of the placement of nerves in orbit have diagnostic value in conditions and diseases of the central nervous system.

The **visual field**, or **field of vision**, is the angular extent of the observable world that can be seen with eyes at any given moment.

As a result, lesions of the optic path, subcortical and cortical visual centres, and disturbed perception of visual images fall on the same half of the retina of both eyes. Thus are "blind" opposite halves of the visual fields. This pathology is called *hemianopia*, or *hemianopsia* (loss of half of the visual field of each eye). The loss of half of the visual field on the same side in both eyes is a *homonymous hemianopia*. The visual images that we see to the right side travel from both eyes to the left side of the brain, while the visual images we see to the left side in each eye travel to the right side of the brain. Therefore, damage to the right side of the posterior portion of the brain or right optic tract can cause a loss of the left field of view in both eyes. Likewise, damage to the left posterior brain or left optic radiation can cause a loss of the right field of vision.

The loss of half of the visual field on different sides in both eyes is a *heteronymous hemianopia*. It is separated into two categories:

– binasal hemianopia – the loss of the fields surrounding the nose;

– bitemporal hemianopia – the loss of the fields closest to the temples.

Other forms of hemianopia:

– superior hemianopia – the upper half of the field of vision is affected, possibly because of a tumour beginning to

compress the lower part of the chiasma, tipically one from the hypophysis;

– inferior hemianopia – the lower half of the field of vision is affected, possibly because of a tumour beginning to compress the upper part of the chiasma, tipically a craneopharyngioma.

Quadrantanopia (*quadrantanopsia*, or *quadrantic hemianopsia*) is decreased vision or blindness in one quarter of the visual field. The particular quarter of vision missing depends on whether the location of the brain damage is temporal or parietal, and the side of the lesion. For example, a lesion to the right temporal lobe with damage specifically to Meyer's loop will give rise to a left upper (superior) quadrantanopsia, while a lesion to the right parietal radiation with damage specifically to Baum's loop will result in a left lower (inferior) quadrantanopsia.

The additional structures of the eye are:

– external muscles of the eyeball (musculi externi bulbi oculi);

– eyebrows (supercilia);

– eyelids (palpebrae);

– the connective membrane, the conjunctiva (tunica conjunctiva);

– lacrimal apparatus (apparatus lacrimalis).

The external muscles of the eyeball (musculi externi bulbi oculi) are divided into:

– direct muscles (mm. recti);

– oblique muscles (mm. obliqui);

– other muscles.

The direct muscles of the eyeball include:

– upper straight muscle (m. rectus superior);

– lower straight muscle (m. rectus inferior);

– lateral rectus muscle (m. rectus lateralis);

– medial rectus muscle (m. rectus medialis).

The primary *function* of the four rectus muscles is to control the eye's movements from left to right and up and down. The two oblique muscles rotate the eyes inward and outward. All six muscles work in unison to move the eye. As one contracts, the opposing muscle relaxes, creating smooth movements. In addition to the muscles of one eye working together in a coordinated effort, the muscles of both eyes work in unison so that the eyes are always aligned:

- The superior rectus attaches to the top of the eye. It moves the eye upward.
- The inferior rectus attaches to the bottom of the eye. It moves the eye downward.
- The medial rectus attaches to the side of the eye near the nose. It moves the eye inward toward the nose.
- The lateral rectus attaches to the side of the eye near the temple. It moves the eye outward.
- The superior oblique comes from the back of the orbit. It travels through a small pulley (the trochlea) in the orbit near the nose and then attaches to the top of the eye. The superior oblique rotates the eye inward around the long axis of the eye (front to back). The superior oblique also moves the eye downward.
- The inferior oblique arises in the front of the orbit near the nose. It then travels outward and backward in the orbit before attaching to the bottom part of the eyeball. It rotates the eye outward along the long axis of the eye (front to back). The inferior oblique also moves the eye upward.

Other external muscles of the eyeball (musculi externi bulbi oculi) are:

– superior and inferior rectus muscles;

– lateral and medial rectus muscles;

– superior and inferior oblique muscle.

Ductions

Movements involving just one eye are called ductions. Rotations around the vertical axis move the anterior pole of the globe medially – *adduction*, or laterally – *abduction*. Rotations around the horizontal axis move the anterior pole of the globe up – *elevation* (*supraduction*), or down – *depression* (*introduction*).

Innervation

Oculomotor nerve (N. III): superior rectus muscle, inferior rectus muscle**,** medial rectus muscle, inferior oblique muscle, levator palpebrae superior muscle.

Trochlear nerve (N. IV): superior oblique muscle.

Abducens nerve (N. VI): lateral rectus muscle, retractor bulbi muscle.

The extraocular muscles are supplied mainly by branches of the ophthalmic artery. This is done either directly or indirectly, as in the lateral rectus muscle, via the lacrimal artery, a main branch of the ophthalmic artery. Additional branches of the ophthalmic artery include the ciliary arteries which branch into the anterior ciliary arteries. Each rectus muscle receives blood from two anterior ciliary arteries, except for the lateral rectus muscle which receives blood from only one. The exact number and arrangement of these ciliary arteries may vary. Branches of the infraorbital artery supply the inferior rectus and inferior oblique muscles.

The internal muscles of the eyeball (*musculi interni bulbi oculi*), which are smooth muscle fibers (myofibrae glabrae), include:

1) ciliary muscle (m. ciliaris), which has:
   – meridian fibers (fibrae meridionales);

– longitudinal fibers (fibrae longitudinales);
– radial fibers (fibrae radiales);
– circular fibers (fibrae circulares);

2) iris sphincter muscle (m. sphincter pupillae);
3) iris dilator muscle (m. dilatator pupillae).

The superior tarsal muscle (*STM*, or *Müller's muscle*) is a smooth muscle adjoining the levator palpebrae superioris muscle that helps raise the upper eyelid. It originates on the underside of the levator palpebrae superioris muscle and inserts onto the superior tarsal plate of the eyelid. The STM receives its innervation from the sympathetic nervous system arising from fibers that enter the cranial skull base encircling the carotid artery (pericarotid plexus), then traverse ventrally the cavernous sinus, and access the orbit wherein the nerve fibers form a tight plexus around the ophthalmic artery. These postganglionic sympathetic fibers originate in the superior sympathetic cervical ganglion. The STM acts synergistically with the levator palpebrae superioris muscle to raise the upper eyelid. Damage to some elements of the sympathetic nervous system can inhibit this muscle, causing a drooping eyelid phenomenon (blepharoptosis).

Eyelids, or palpebrae, are folds of skin and tissue that when closed cover the globe. The eyelids have four major functions:

– cover the globe for protection;
– move the tears toward drainage at the medial canthus on closure, spread the tear film over the anterior surface of the eye on opening;
– contain structures that produce the tear film.

The upper eyelid extends to the eyebrow and is divided into the tarsal and the orbital (or preseptal) parts. The tarsal portion lies closest to the lid margin, rests on the globe, and

contains the tarsal plate. The skin is thin, and the underlying loose connective tissue is devoid of adipose tissue. The orbital portion extends from the tarsus to the eyebrow and a furrow – the superior palpebral sulcus – separates the tarsal portion from the orbital portion.

In the lower eyelid the inferior palpebral sulcus which separates the lower lid into tarsal and orbital parts is often not very distinct. The tarsal portion rests against the globe and the orbital portion extends from the lower border of the tarsus onto the cheek extending just past the inferior orbital margin to the nasojugal and malar sulci.

Eyelid structures

Eyelid skin is very thin, soft, and poor in fat. It is loosely connected to the tissues located deeper. Features of the structure cause lung edema spread (in terms of local inflammation, venous stasis, some common diseases, hemorrhage). Thin and elastic skin of this area is convenient for plastic surgery.

The skin of the eyelids are two horizontal folds – the upper and lower (sulcus orbitopalpebralis superior et inferior), corresponding limits cartilage. The upper, depending on muscle tone, lifts the upper lid and the bottom corresponds to the lower orbital edge.

Musclus orbicularis oculi are the striated fibers of the orbicularis oculi muscle located below the subcutaneous connective tissue layer; encircle the palpebral fissure from the eyelid margin to overlap onto the orbital margin. The muscle can be divided into two regions: palpebral and orbital.

The palpebral portion of the orbicularis oculi muscle occupies the area of the eyelid that rests on the globe and is closest to the eyelid margin. It sometimes is divided further into pretarsal and preseptal parts.

The orbital portion of the orbicularis oculi muscle is attached superiorly to the orbital margin, medial to the supraorbital notch.

The orbicularis oculi muscle is innervated by the cranial nerve VII (the facial nerve).

Muscle levator palpebral superiori is the superior palpebral levator muscle, the retractor of the upper eyelid; located within the orbit above the globe and extends into the upper lid. It originates on the lesser wing of the sphenoid bone above and in front of the optic foramen and its sheath blends with the sheath of the superior rectus muscle.

Levator aponeurosis: as the levator enters the eyelid, it becomes a fan-shaped tendinous expansion, or the levator aponeurosis. Unlike a typical tendon, the aponeurosis spreads out into an extensive sheet posterior to the orbital septum. The fibers of the aponeurosis penetrate the orbital septum and extend into the upper lid fanning out across its entire width.

The tarsal plates are composed of dense connective tissue. The collagen fibrils of this tissue are of uniform size and run both vertically and horizontally to surround the meibomian glands.

The palpebral conjunctiva is composed of two layers, a stratified epithelial layer and a connective tissue stromal layer, the submucosa.

Glands of the lids

The meibomian glands (*tarsal glands*) are sebaceous glands embedded in the tarsal plate. These long multilobed glands resemble a large bunch of grapes and are arranged vertically so that their openings are located in a row along the lid margin posterior to the cilia.

The glands of Zeis are unilobar sebaceous glands located on the margin of the eyelid. These glands produce an

oily substance (sebum) into the hair follicle of the cilia, coating the eyelash shaft to keep it from becoming brittle.

The glands of Moll have been called modified sweat glands but are more accurately described as specialized apocrine glands. They are located near the lid margin and their ducts empty into the hair follicle, into the Zeis gland duct or directly onto the lid margin.

The Krause's glands are the accessory lacrimal glands located in the stroma of the conjunctival fornix and *the accessory lacrimal glands of Wolfring* are located along the orbital border of the tarsal plate.

Innervation of the eyelids

The ophthalmic and maxillary divisions of the trigeminal nerve provide sensory innervation of the eyelids.

The upper lid is supplied by the supraorbital, supratrochlear, infratrochlear, and lacrimal nerves, branches of the ophthalmic division.

The supply to the lower lid is by the infratrochlear branch of the ophthalmic nerve and the infraorbital nerve, a branch of the maxillary division.

Motor control of the orbicularis muscle is through the temporal and zygomatic branches of the facial nerve and that of the levator muscle is through the superior division of the oculomotor nerve. The tarsal smooth muscles are innervated by sympathetic fibers from the superior cervical ganglion.

Blood supply of the eyelids

The blood vessels are located in a series of arcades or arches in each eyelid. The marginal palpebral arcade lies near the lid margin and the peripheral palpebral arcade lies near the orbital edge of the tarsal plate.

The vessels forming these arcades are branches from the medial and lateral palpebral arteries. The medial and lateral

palpebral arteries are branches of the ophthalmic and lacrimal arteries, respectively. Normal variations occur in the blood supply and the most common variation is a lack of the peripheral arcade in the lower lid.

Conjunctiva is a thin, translucent mucous membrane that runs from the limbs over the anterior sclera, forms a cul-de-sac at the superior and inferior fornixes, and turns anteriorly to line the eyelids. It ensures smooth movement of the eyelids over the globe. The conjunctiva can be divided into three sections that are continuous with one another:

1) the tissue lining the eyelids is the palpebral conjunctiva or tarsal conjunctiva;

2) the bulbar conjunctiva covers the sclera;

3) the conjunctival fornix is the cul-de-sac connecting palpebral and bulbar sections.

Blood vessels

The palpebral conjunctiva receives its blood supply from the palpebral arcades. Branches from the arcades anastomose on both sides of the tarsal plate; vessels from the posterior network supply the palpebral conjunctiva in both upper and lower lids.

The fornices are supplied by branches from the peripheral arcades, which then branch again and enter the bulbar conjunctiva, forming a plexus of vessels, the posterior conjunctival arteries. These anastomose with the plexus of anterior conjunctival arteries formed by branches from the anterior ciliary arteries.

Conjunctival veins parallel the arteries but are more numerous. They drain into the palpebral and ophthalmic veins.

The conjunctival lymphatic vessels are arranged in superficial and deep networks within the submucosa.

These vessels drain into the lymphatics of the eyelids; those from the lateral aspect empty into the parotid lymph node, and those from the medial aspect empty into the submandibular lymph node.

Conjunctival innervation

Sensory innervation of the bulbar conjunctiva is through the long ciliary nerves. Sensory innervation of the superior palpebral conjunctiva is provided by the frontal and lacrimal branches of the ophthalmic nerve. Innervation of the inferior palpebral conjunctiva is provided by the lacrimal nerve and the infraorbital branch of the maxillary nerve. All sensory information is carried in the trigeminal nerve.

Tenon's capsule

Below the conjunctival stroma is a thin, fibrous sheet called Tenon's capsule (fascia bulbi). Tenon's capsule serves as a fascial cavity within which the globe can move. It protects and supports the globe and attaches it to the orbital connective tissue.

Tear film

The tear film which covers the anterior surface of the globe has several functions:

– keeps the surface moist and serves as a lubricant between the globe and eyelids;

– traps debris and helps remove sloughed epithelial cells and debris;

– is the primary source of atmospheric oxygen for the cornea;

– provides a smooth refractive surface necessary for optimum optical function;

– contains antibacterial substances (lysozyme, beta-lysin, lactoferrin, immunoglobulins) to help protect against infection;

– helps to maintain corneal hydration by changes in tonicity that occur with evaporation;

– contains various growth factors and peptides that can regulate ocular surface wound repair.

The tear film is composed of three layers. The outermost is a lipid layer, the middle or aqueous layer, the innermost or mucous layer.

Functions:

1. Protective: mechanical (protects from exposure to dust, pollutants, small foreign bodies), barrier (protects from penetration of microorganisms), moisturizing (protects from drying out).
2. Aspiration.
3. Nutritious.

Lacrimal system

The secretory system includes the main lacrimal gland, accessory lacrimal glands, meibomian glands, and conjunctival goblet cells. The main lacrimal gland is located in a fossa on the temporal side of the orbital plate of the frontal bone, just posterior to the superior orbital margin.

The lacrimal gland is divided into two portions, *palpebral* and *orbital,* by the aponeurosis of the levator muscle.

The superior orbital portion is larger and almond shaped. The superior surface lies against the periorbita of the lacrimal fossa, the inferior surface rests against the aponeurosis, the medial edge lies against the levator, and the lateral edge lies on the lateral rectus muscle. The palpebral lobe is one third to one half the size of the orbital lobe and is subdivided into two or three sections.

The lacrimal gland is supplied by the lacrimal artery, a branch of the ophthalmic artery. Sensory innervation is through the lacrimal nerve, a branch of the ophthalmic division of the

trigeminal nerve. The gland receives vasomotor sympathetic innervation and secretomotor parasympathetic innervation.

A small aperture, the *lacrimal punctum*, is located in a slight tissue elevation, the lacrimal papilla, at the junction of the lacrimal and ciliary portions of the eyelid margin. Both upper and lower lids have a punctum. The puncta are turned toward the globe and normally can be seen only if the eyelid edge is everted slightly. Each punctum opens into a tube, the lacrimal canaliculus.

The lacrimal sac lies within a fossa in the anterior portion of the medial orbital wall. This fossa is formed by the frontal process of the maxillary bone and the lacrimal bone. The sac is surrounded by fascia, continuous with the periorbita, which runs from the anterior to the posterior lacrimal crests. The two limbs of the medial palpebral ligament straddle the sac to attach to the posterior and anterior crests. The orbital septum and the check ligament of the medial rectus muscle lie behind the lacrimal sac.

The lacrimal sac empties into the nasolacrimal duct just as it enters the nasolacrimal canal in the maxillary bone. The duct is approximately 15 mm long and terminates in the inferior meatus of the nose. At this point the valve of Hasner is found. This fold of mucosal tissue prevents retrograde movement of fluid up the duct from the nasal cavity.

The supply area of the lacrimal is going through the eye artery (branch of the internal carotid artery) and through the foreign maxillary artery (branch of the external carotid artery). Branches of these arteries form an anastomosis well together. The largest of vessels are the angular artery (a. angularis) and nasal artery (a. dorsalis nasi); veins accompany the arteries.

The main source of blood supply to the orbit is by the ophthalmic artery, the first branch of the internal carotid artery. The external carotid artery normally contributes only to a small

extent to the orbital blood supply via the infraorbital artery and orbital branch of the middle meningeal artery.

Throughout its rather tortuous course, there are many branches of the ophthalmic artery:

- central retinal artery;
- lacrimal artery;
- ciliary arteries (usually two, sometimes three);
- ethmoid arteries (usually two);
- supraorbital artery;
- muscular arteries (usually two);
- medial palpebral arteries (superior and inferior);
- supratrochlear artery;
- arteria dorsalis nasi.

The orbital venous system is complex, highly variable, and confusing. In the orbit, in contrast to other parts of the body, there is no direct correspondence between the arteries and veins, except for the superior ophthalmic vein that has some correspondence with the ophthalmic artery. Also, the orbital veins, unlike the orbital arteries, have a highly variable and inconstant pattern and formation of venous networks at several places, resulting in marked uncertainty and controversy on their number, nomenclature and pattern.

The complex, highly variable and confusing orbital venous system can be divided into:

- main orbital veins (superior and inferior ophthalmic veins);
- inconstant orbital veins (middle and medial ophthalmic veins and four collateral veins);
- orbital venous networks;
- various venous tributaries.

Cranial Innervation of Ocular Structures

The orbital structures are innervated by cranial nerves (CNs) II, III, IV, V, VI, and VII. Motor functions of the striated

muscles are controlled by CN III, the oculo-motor nerve; CN IV, the trochlear nerve; CN VI, the abducens nerve; and CN VII, the facial nerve. CN V, the trigeminal nerve, carries the sensory supply from the orbital structures. CN II, the optic nerve, carries visual information and is discussed. This chapter discusses sensory and motor innervation of the orbit, including pathways.

Trigeminal nerve

The trigeminal nerve is the largest and most complex of the cranial nerves (CNs). It supplies sensations to the face, mucous membranes, and other structures of the head. It contains proprioceptive fibers. The fibers of the trigeminal nerve (CN V) serving ocular structures are sensory and originate in the innervated structures.

Ophthalmic Division of the Trigeminal Nerve

Nasociliary Nerve

Sensory fibers from the structures of the medial central area – caruncle, canaliculi, lacrimal sac, medial aspect of the eyelids, and skin at the side of the nose – join to form the infratrochlear nerve.

Sensory fibers from the skin along the centre of the nose, the nasal mucosa, and the ethmoid sinuses form the *anterior ethmoid nerve*; fibers from the ethmoid sinuses and the sphenoid sinus form the *posterior ethmoid nerve*.

Some of these branches join with nerves from other anterior segment structures to form two long ciliary nerves.

Thus, the nasociliary nerve is formed by the joining of the infratrochlear nerve, anterior and posterior ethmoid nerves, long ciliary nerves, and sensory root of the ciliary ganglion.

Frontal Nerve

Sensory fibers from the skin and muscles of the forehead and upper eyelid come together and form the

supratrochlear nerve. This nerve enters the orbit by piercing the superior medial corner of the orbital septum.

Sensory fibers from the skin and muscles of the forehead and upper eyelid form a second nerve, the *supraorbital nerve*, and lateral to the supratrochlear nerve.

Lacrimal Nerve

Sensory fibers from the lateral aspect of the upper eyelid and temple area come together and enter the lacrimal gland; they join the sensory fibers that serve the gland itself to form the lacrimal nerve.

After exiting the orbit, the nasociliary, lacrimal, and frontal nerves join and form the ophthalmic division of the trigeminal nerve.

Maxillary Division of the Trigeminal Nerve

Infraorbital Nerve

The infraorbital nerve, formed by sensory fibers from the cheek, upper lip, and lower eyelid, enters the maxillary bone through the infraorbital *foramen*.

It runs posteriorly through the infraorbital *canal* and *groove;* while it is in the maxillary bone, branches join from the upper teeth and maxillary sinus. As the nerve leaves the infraorbital groove it exits the orbit through the inferior orbital fissure and joins other fibers in forming the maxillary nerve.

Zygomatic Nerve

Sensory fibers from the lateral aspect of the forehead enter the orbit through a foramen in the zygomatic bone as the zygomatico temporal nerve.

Maxillary Nerve Formation

Having been formed by the joining of the infraorbital nerve, zygomatic nerve, and nerves from the roof of the mouth, upper teeth and gums, and mucous membranes of the cheek, the maxillary nerve traverses the area between the maxilla and the sphenoid bone. As it passes near the fossa pterygopalatine,

it receives some autonomic fibers from the ganglion pterygopalatine.

Mandibular Division of the Trigeminal Nerve

The mandibular nerve innervates the lower face and contains both sensory and motor fibers. It enters the skull via the foramen ovale.

Efferent pathway motor nerves: the cranial nerves that supply striated muscles of the orbit and adnexa are the oculomotor, trochlear, abducens, and facial nerves.

Oculomotor nerve: cranial nerve III

The oculomotor nerve innervates the superior rectus, medial rectus, inferior rectus, inferior oblique, and superior palpebral levator muscles. It also provides a route along which the autonomic fibers travel to innervate the iris sphincter muscle, the ciliary muscle, and the smooth muscles of the eyelid.

Trochlear nerve: cranial nerve IV

The trochlear nerve innervates the superior oblique muscle. IV pair of cranial nerves has its own motor core – *nucleus block nerve* (*nucleus nervi trochlearis*), located in the gray matter of the midbrain roof.

Axons of neurons in this nucleus forming block nerve emanating from the brain (encephalon) of the upper brain sails (velum medullare superius), based on the brain arises from the lateral surface of the leg of the brain (crura cerebri), and from the cranial cavity (cavitas cranii) – through the upper orbital fissure (fissura orbitalis superior).

Abducens nerve: cranial nerve VI

The abducens nerve innervates the lateral rectus muscle.

VI pair of cranial nerves has its own motor *nucleus abducens nerve* (*nucleus nervi abducentis*), that resides in the gray matter of the roof of the bridge (substantia grisea tegmenti pontis) on top of the mound facial rhomboid fossa (apex colliculi facialis fossae rhomboideae).

Facial nerve: cranial nerve VII

The facial nerve has two roots: the large motor root innervates the facial muscles, and the smaller root contains sensory and parasympathetic fibers. The sensory fibers carry taste sensations from the tongue. The parasympathetic nerves supply secretomotor fibers to various glands of the face; those supplying the lacrimal gland are discussed.

Ciliary node (ganglion ciliare) measuring about 2 mm is placed under the external rectus (between it and the optic nerve) at a distance of 10–18 mm of the posterior pole of the eye. It includes:

– *sensitive fibers* – long spine of nasal ciliary nerve, separated from the trunk of the trigeminal nerve deep in orbit; movement is from the lower branches of the oculomotor nerve;

– *plexus of the internal carotid artery* – from ciliary nerve, branch off 4–6 short ciliary nerves, which combine with a branch of the sympathetic nerve, enter the vascular tract through the sclera, form 20–30 small nerves distributed in the tissues of the eye, usually in the vascular tract.

Thus, short cilia perform motor nerves, sensory and vasomotor innervation of the eye.

Oculomotor abducens nerve blocks and reaches the muscles, getting to orbit cavernous sinus. Motor nerves are sympathetic fibers that branch off from the carotid plexus.

Trauma or inflammation in the area of the upper orbital fissure due to violations of the neurovascular bundle with a current venous (v. ophthalmic superior) function and damage of motor nerves (n. oculomotorius and n. abducens) and the first branch of the trigeminal nerve (n. ophthalmicus) characterizes the following symptoms: retrobulbar hemorrhage, loss of muscle tone and sensitivity of the eyes and trophic broken cornea. The result is that later appears neuro paralytic keratitis.

TOPIC 2. EYE EXAMINATION

Vision is the most important of all senses. Approximately 80 % of information from the outside world is incorporated through the visual pathway. Loss of vision has a profound effect on the quality of life.

Basic examination:

- observation of external structures;
- extraocular movements and cranial nerves;
- dilating the pupil;
- visual acuity;
- children's visual acuity;
- slit-lamp examination;
- intraocular pressure (IOP);
- visual field (confrontation) testing;
- colour vision;
- ophthalmoscopy.

Other tests carried out in the ophthalmology departments are:

- visual field assessment;
- ultrasound;
- exophthalmometery;
- keratometry;
- Hess chart fluorescein angiography;
- optical coherence tomography (OCT);
- visually evoked potential (VEP), also called visually evoked response (VER) and visually evoked cortical potential (VECP).

Observation of External Structures

1. *Ocular symmetry.* Occasionally, one of the muscles that control eye movement will be weak or foreshortened, causing one eye to appear deviated medially or laterally compared with the other.

2. *Eyelid symmetry*. Both eyelids should cover approximately the same amount of eyeball. Damage to the nerves controlling these structures (Cranial Nerves III and VII) can cause the upper or lower eyelids on one side to appear lower than the other.

3. *Sclera.* The normal sclera is white and surrounds the iris and pupil. In the setting of liver or blood disorders that cause hyperbilirubinemia, the sclera may appear yellow, referred to as icterus. This can be easily confused with a muddy-brown discolouration common among older African Americans among whom it is a normal variant.

4. *Conjunctiva.* Normally, it's invisible except for the fine blood vessels that run through it. When infected or otherwise inflamed, this layer can appear quite red, a condition known as conjunctivitis. Blood can also accumulate underneath the conjunctiva when one of the small blood vessels within it ruptures. This may be the result of relatively minor trauma (cough, sneeze, or direct blow), a bleeding disorder or idiopathic. The resulting collection of blood is called a subconjunctival hemorrhage.

5. *Pupil and Iris*. Normally, both of these structures are round and symmetric.

Extraocular Movements and Cranial Nerves

Normally, the eyes move in concert (e. g. when the left eye moves left, the right eye moves left to a similar degree). The brain takes the input from each eye and puts it together to form a single image. This coordinated movement depends on the 6 extraocular muscles inserted around the eye balls, allowing them to move in all directions. Each muscle is innervated by one of the Cranial Nerves (CNs): CN III (Oculomotor), IV (Trochlear) and VI (Abducens). Movements are described as: elevation (pupil directed upwards), depression (pupil directed downwards), abduction (pupil directed

laterally), adduction (pupil directed medially), extorsion (top of eye rotating away from the nose), and intorsion (top of eye rotating towards the nose).

Dilating the Pupil

Mydriatic drops. Dilate one eye when you start your H&P (history and physical) and by the time you are done you will have a good look. In general *tropicamide* is considered the safest.

Parasympathetic antagonists. These paralyze the circular muscle of iris (mydriasis) and the ciliary muscle (loss of accommodation):

– *tropicamide*: 1–2 drops (0.5 %) 15–20 minutes before exam; may repeat every 30 minutes as necessary. Individuals with heavily pigmented eyes may require larger doses;

– *cyclopentolate*: 1 drop of 1 % followed by another drop in 5 min; 2 % solution in heavily pigmented iris;

– *atropine*: (1 % solution): instill 1–2 drops 1 hour before the procedure;

– *homatropine*: 1 drop of 2 % solution immediately before the procedure; repeat at 10 min intervals as necessary.

Sympathetic agonist:

– *phenylephrine*: 1 drop of 2.5 % or 10 % solution, may repeat in 10–60 min as necessary.

Pupillary reactions: these should be tested in a dimly lit room (to avoid pupillary constriction from the room light over-riding that from your torch). Tell the patient to look at a far wall to overcome the accommodation reflex. Use a bright light source directed from below (to avoid the shadow from the nose).

Direct response to light*:* light directly shone on the eye for three seconds should elicit a prompt pupillary constriction

of the pupil. Failure to do so is known as an afferent pupillary defect:

– if there is also failure of the pupil to constrict, this indicates severe optic nerve pathology (e. g., transected nerve);

– if there is no pupillary constriction to light but the pupil does constrict, consider a traumatic iris paresis.

The swinging flashlight test. This may elicit a relative afferent pupillary defect (RAPD). Shine the light source from one eye to the other in rapid succession. Stimulation of the normal eye should elicit a brisk constriction of both pupils but when the light is shone on the diseased eye, both pupils dilate. This is because the dilatation produced by withdrawing the light from the normal eye outweighs the weak constriction produced by shining light on the diseased eye. It can be difficult to elicit this sign if there are dark irises and sluggish, dilated, or miotic pupils.

Light-near dissociation. If the reactions to light are normal, proceed to the accommodation reflex. The room light should be turned on again and the patients asked to look at a far wall. Tell them that as soon as they see your pen (or other object), they should focus straight on it. As they gaze to the distance, hold your object above the level of their eyes, then drop it into their line of view and observe the pupillary reactions as they look at it. There should be a brisk constriction. Absence of this is known as light-near dissociation.

If all pupillary tests are normal, the patient can be said to have **P**upils **E**qual and **R**eactive to **L**ight and **A**ccommodation (PERLA).

In the normal clinical settings, we measure only one of these functions – central resolution at high contrast (visual acuity).

Visual Acuity

This essential examination should be carried out on every patient presented with an eye problem.

Snellen chart: this comprises random letters arranged in rows, decreasing in size in each row. Charts are designed to be read at three or six meters. The number indicated at the side of the row corresponds to the distance at which a normal eye could read that row. For example, the top row (marked 60) could be read by the normal eye 60 meters away. The patient should be tested one eye at a time using their normal distance glasses (or distance portion of their bifocals) and then using a pinhole – you cannot assume that their glasses are of the correct prescription and the pinhole will correct any refractory errors, unless there is media opacity – e. g., corneal edema.

The reading is recorded as 6/60 – this means that the patient was tested at 6 meters (or equivalent if you used a reversed three-meter chart and a mirror) and was able to read the top row only.

If they score 6/4 (i. e., read the lowest row), they were tested at 6 meters but their eyesight was so good that they actually saw what a 'normal' person would usually need to be four meters away to read.

If the patient is unable to read the top row, try counting fingers (CF) at 1 meter in a well-lit room, then hand movements (HM), then perception of light (PL).

If the patient sees nothing at all, they are said to be NPL (no perception of light).

The patient reads most of a line right but gets one or two wrong at the 12-metre row, for example, and this is recorded as 6/12–2.

If he or she gets more than two wrong, assume that the patient can only read the line above. Similarly, if he or she could manage a couple of letters in the line below but not the whole line, it is recorded as 6/12+2.

There are variations of the Snellen chart for patients who are illiterate: capital ‘E’s are rotated in different directions which the patient has to identify.

Children’s Visual Acuity

Children may use the Sheridan–Gardiner test where they have to match up letters or pictures of different sizes with those presented on a card in front of them.

Very young children are examined by assessing their preferential looking at cards of various pictorial complexity (Cardiff card test) and babies may be assessed by their ability to pick up very small objects such as the ‘hundreds and thousands’ cake decorations. Infants are watched for tracking of a light source.

There exist many other visual acuity tests which take other factors into account, such as contrast sensitivity and the crowding phenomenon (where the spacing between the letters affects the acuity), but these are the area of activity of ophthalmology departments.

Slit-Lamp Examination

The slit-lamp is one of the important examining tools of ophthalmologists. Clinical ophthalmologists all over the world routinely use a slit-lamp to examine their patients.

Clinical procedure

Before using the slit-lamp, it is important to ensure that the instrument is correctly set up. The following points should be checked:

– the eyepieces should be focused for the observer for his/her own refractive error; often a little more minus correction is required than the observer’s actual refractive error due to proximal accommodation and convergence;

– the pupillary distance (PD) is adjusted for the observer (perhaps the PD should be slightly less than the usually measured to account for proximal convergence);

– check that the slit-lamp is parallel on the runners of the table;

– check that the observation and illumination systems are coupled, and the slit-beam is of even illumination and has sharply demarcated edge (otherwise irregularity of the beam may be falsely interpreted as irregularity of tissues);

– the locations of the controls are known;

– the observer and patient are comfortable in the mid-travel of the slit-lamp. Mid-travel is the location of the slit-lamp when it is half-way up or down.

Clinical application*:* slit-lamp biomicroscopy is very useful in the diagnosis of eye diseases. It should routinely be performed in almost all diseases of the eye.

Intraocular Pressure (IOP)

This needs to be measured where glaucoma is suspected.

Quick examination

Very low IOP may manifest itself as a soft eyeball at palpation of the globe over the closed eyelids, and very high IOP may feel hard. However, this is a very crude measure (notoriously unreliable) and a globe thought to be soft on account of perforation should not be palpated. It is not a substitute for proper tonometry where there is a concern over IOP.

Tonometry

IOP can be very easily measured using a tonometer (normal readings should be between 10 mm Hg and 21 mm Hg). There are many types of tonometers, most of which make contact with the eye surface, so that the eye is first anaesthetized.

Goldmann tonometry has long been considered the gold-standard method. It uses a prism pressed against the cornea.

Non-contact (‘puff of air’) tonometers were not considered to be an accurate way to measure IOP, although they were a fast and simple way to screen for high IOP. However, modern non-contact tonometers correlate well with Goldmann tonometry measurements and are particularly useful for measuring IOP in children and other non-compliant patient groups.

Although tonometry is completely painless, many patients find it very difficult.

Visual Fields

Perimetry is the systematic measurement of a visual field function. The two most commonly used types of perimetry are Goldmann kinetic perimetry and Threshold Static Automated Perimetry. With Goldmann or “kinetic” perimetry, a trained perimetrist moves the stimulus; the stimulus brightness is held constant. The limits of the visual field are mapped to lights of different sizes and brightness.

With Threshold Static Automated Perimetry, a computer program is selected. The most commonly used one tests the central 30° of the visual field using a six degree spaced grid. This is accomplished by keeping the size and location of a target constant and varying the brightness until the dimmest target the patient can see at each of the test locations is found.

These maps of visual sensitivity, made by either of these methods, are very important in diagnosing diseases of the visual system. Different patterns of visual loss are found with diseases of the eye, optic nerve, central nervous system.

Colour Vision

The Ishihara plates provide a popular and effective method of screening for colour vision defects. The patient is presented with a series of plates with numerous printed

coloured dots. The normal-sighted subject will see numbers on the majority of the plates, whereas the colour-defective patient will fail to see many of the numbers. The test is easy to do and will effectively screen out the more common red-green deficiency found in 8 % of the male population.

Ophthalmoscopy

Ophthalmoscopy is an examination of the back part of the eye (fundus), which includes the retina, optic disc, choroid and blood vessels.

There are different types of ophthalmoscopy:

Direct ophthalmoscopy. You will be seated in a darkened room. The health care provider performs this exam by shining a beam of light through the pupil using an instrument called an ophthalmoscope. An ophthalmoscope is about the size of a flashlight. It has light and different tiny lenses that allow the provider to view the back of the eyeball.

Indirect ophthalmoscopy. You will either lie or sit in a semi-reclined position. The provider holds your eye open while shining a very bright light into the eye using an instrument worn on the head. (The instrument looks like a miner's light). The provider views the back of the eye through a lens held close to your eye. Some pressure may be applied to the eye using a small, blunt probe. You will be asked to look in various directions.

Slit-lamp ophthalmoscopy. You will sit in a chair with the instrument placed in front of you. You will be asked to rest your chin and forehead on a support to keep your head steady. The provider will use the microscope part of the slit lamp and a tiny lens placed close to the front of the eye. The provider can see about the same with this technique as with indirect ophthalmoscopy, but with higher magnification.

The ophthalmoscopy examination takes about 5 to 10 minutes.

Other Tests Carried Out in the Ophthalmology Department

Other tests that are routinely performed in specialist units include:

Visual field assessment is using static and kinetic perimeters. Perimetry or campimetry systematically tests the visual field through the detection of the presence of test targets on a defined background. Perimetry maps and quantifies the visual field, especially at the extreme periphery. Automated perimeters are used widely.

Ultrasound is used to visualize the structures of lens, vitreous and retina.

Exophthalmometery is used to assess proptosis (e. g., thyroid eye disease). There are several types of exophthalmos meters, some of which measure the distance of the corneal apex from the level of the lateral orbital rim while others measure the relative difference between each eye.

Keratometry is the measurement of the corneal curvature, which determines the power of the cornea. Differences in power across the cornea result in astigmatism. Keratometry can be done manually or using automated devices. Keratometry allows visualization of the pre-corneal tear film and a dynamic view of the surface of the cornea and of the tear film. You can recognize areas of corneal surface irregularity or compromise. If the tear film is oily or disrupted, or the cornea has subtle dystrophy or degeneration, it will be reflected in the quality of the measurements.

Hess chart: this maps extraocular muscle movement and assesses diplopia. In the Hess test the patient's left and right eyes see two similar grids superimposed by angled mirrors. They are then asked to point out the grid's intersection points with a marker.

In a normal patient, the results would be centreed on each chart. Distortion in caused by uncoordinated movements of the eye muscles.

Fluorescein angiogram lets the doctor see how well blood moves in your retina. It helps diagnose diabetic retinopathy, retinal detachment, and macular degeneration. The doctor will inject a special dye, called fluorescein, into a vein in your arm. It travels quickly to blood vessels inside your eye. Once it gets there, the doctor uses a camera with special filters to highlight the dye. He takes pictures of the dye as it goes through the blood vessels in the back of your eye. This helps him spot circulation problems, swelling, leaking, or abnormal blood vessels.

Optical coherence tomography (OCT) uses light waves to take detailed cross-section images of the retina. Imaging of retinal layers helps with diagnosis and provides treatment guidance for glaucoma and retinal disease, such as age-related macular degeneration and diabetic retinopathy. The OCT machine scans the eye without touching it, through a dilated pupil. Scanning takes about 5–10 minutes.

Visually evoked potential (VEP), also called ***visually evoked response (VER)*** and ***visually evoked cortical potential (VECP)***: this measures electrical potentials, initiated by brief visual stimuli, recorded from the scalp overlying the visual cortex. VEPs are used primarily to measure the functional integrity of the visual pathways from retina via the optic nerves to the visual cortex. Any abnormality that affects the visual pathways or visual cortex can affect the VEP, for example, cortical blindness due to meningitis or anoxia, optic neuritis as a consequence of demyelination, optic atrophy, stroke and compression of the optic pathways. Myelin plaques (found in multiple sclerosis) tend to slow the speed of VEP wave peaks. Compression of the optic pathways reduces amplitude of wave peaks.

TOPIC 3. REFRACTION AND ACCOMMODATION

Errors of Refraction

Refractive error means that the shape of your eye does not bend light correctly, resulting in a blurred image. The main types of refractive errors are:

- myopia (nearsightedness);
- hyperopia (farsightedness);
- presbyopia (loss of near vision with age);
- astigmatism.

Emmetropia (optically normal eye) can be defined as a state of refraction, where in the parallel rays of light coming from infinity are focused at the sensitive layer of retina with the accommodation being at rest.

At birth, the eyeball is relatively short, having +2 D to +3 D hypermetropia.

This is gradually reduced until by the age of 5–7 years the eye is emmetropic and remains so till the age of about 50 years. After this, there is tendency to develop hypermetropia again, which gradually increases until at the extreme of life the eye has the same +2 D to +3 D with which it started. This senile hypermetropia is due to changes in the crystalline lens.

Ametropia (a condition of refractive error) is defined as a state of refraction, when the parallel rays of light coming from infinity (with accommodation at rest), are focused either in front or behind the sensitive layer of retina, in one or both of the meridians. The *ametropia* includes *myopia, hypermetropia* and *astigmatism.*

Myopia

Myopia or short-sightedness is a type of refractive error in which parallel rays of light coming from infinity are focused in front of the retina when accommodation is at rest.

Etiological classification:

– axial myopia;
– curvature myopia;
– positional myopia;
– index myopia results;
– myopia due to excessive accommodation.

Clinical varieties of myopia:

– congenital myopia;
– simple or developmental myopia;
– pathological or degenerative myopia;
– acquired myopia which may be:
  · post-traumatic;
  · post-keratitis;
  · drug-induced;
  · pseudo myopia;
  · space myopia;
  · night myopia;
  · consecutive myopia.

Classification according to the degree of myopia:

– low myopia (< 3.00 D);
– medium myopia (3.00–6.00 D);
– high myopia (> 6.00 D).

Congenital myopia (present at birth and persisting through infancy):

– youth-onset myopia (< 20 years of age);
– early adult-onset myopia (20–40 years of age);
– late adult-onset myopia (> 40 years of age).

Myopia occurs when the eyeball is too long, relative to the focusing power of the cornea and lens of the eye. This causes light rays to focus at a point in front of the retina, rather than directly on its surface.

Nearsightedness also can be caused by the cornea and/or lens being too curved for the length of the eyeball. In some cases, myopia is due to a combination of these factors.

Myopia typically begins in childhood and you may have a higher risk if your parents are nearsighted. In most cases, nearsightedness stabilizes in early adulthood but sometimes it continues to progress with age.

Symptoms

1. *Poor vision for distance* (short-sightedness) is the main symptom of myopia.

2. *Asthenopic symptoms* may occur in patients with small degree of myopia.

3. *Half shutting* of the eyes may be complained by parents of the child. The child does so to achieve the greater clarity of stenopaeic vision.

Signs

1. *Prominent eyeballs.* The myopic eyes typically are large and somewhat prominent.

2. *Anterior chamber* is slightly deeper than normal.

3. *Pupils* are somewhat large and a bit sluggishly reacting.

4. *Fundus* is normal; rarely temporal myopic crescent may be seen.

5. *Magnitude of refractive error.* Simple myopia usually occurs between 5 and 10 year of age and it keeps on increasing till about 18–20 years of age at a rate of about –0.5 ± 0.30 every year. In simple myopia, usually the error does not exceed 6 to 8.

Degenerative Myopia

Pathologic myopia represents a subgroup of myopia and affects up to 3 % of the world population. Vision loss related to pathologic myopia is of great clinical significance as it can be progressive, irreversible and affects individuals during their most productive years. High myopia is defined as refractive error of at least –6.00 D or an axial length of 26.5 mm or more. Pathologic or degenerative myopia is defined as “high myopia

with any posterior myopia-specific pathology from axial elongation".

Etiology

Biomechanical forces related to axial elongation of the eye result in stretching of the ocular layers and progressive thinning of the retina, choroid and sclera.

Risk factors

Primary risk factors for pathologic myopia include greater axial length and age. Additional possible risk factors such as female gender, larger optic disc area and family history of myopia have been suggested by additional studies.

Symptoms

1. *Defective vision.*

2. *Muscae volitantes,* i. e., floating black opacities in front of the eyes are also complained of by many patients. These occur due to degenerated liquefied vitreous.

3. *Night blindness* may be complained by very high myopes having marked degenerative changes.

Signs

1. *Prominent eye balls.*

2. *Cornea* is large.

3. *Anterior chamber* is deep.

4. *Pupils* are slightly large and react sluggishly to light.

5. *Fundus examination* reveals following characteristic signs:

– *optic disc* appears large and pale and at its temporal edge a characteristic myopic crescent is present;

– *degenerative changes in retina and choroid* are common in progressive myopia. These are characterized by white atrophic patches at the macula with a little heaping up of pigment around them;

– *Foster–Fuchs' spot* (dark red circular patch due to sub-retinal neovascularization and choroid hemorrhage) may be present at the macula;

– *cystoid degeneration* may be seen at the periphery. In an advanced case there occurs total retinal atrophy, particularly in the central area;

– *posterior staphyloma* due to ectasia of sclera at posterior pole may be apparent as an excavation with the vessels bending backward over its margins;

– *degenerative changes in vitreous* include: liquefaction, vitreous opacities, and posterior vitreous detachment (PVD) appearing as Weiss' reflex.

6. *Visual fields* show contraction and in some cases ring scotoma may be seen.

7. *ERG* reveals subnormal electroretinogram due to chorioretinal atrophy.

Complications:

– retinal detachment;
– complicated cataract;
– vitreous hemorrhage;
– choroid hemorrhage;
– strabismus fixus convergence.

Treatment of myopia

Optical treatment of myopia constitutes prescription of appropriate concave lenses, so that clear image is formed on the retina.

Surgery to correct refractive errors has become very popular. It should be performed after the error has stabilized; preferably after 20 years of age. Various surgical techniques in vogue are described below:

1. *Radial keratotomy (RK)* refers to making deep (90 percent of corneal thickness) radial incisions in the peripheral part of cornea leaving the central 4 mm optical. These incisions flatten the central cornea thereby reducing its refractive power. This procedure gives very good correction in low to moderate myopia (2 to 6 D).

2. *Photorefractive keratectomy (PRK)*. In this technique, to correct myopia a central optical zone of anterior corneal stroma is photo-ablated using *excimer laser*. Like RK, the PRK also gives very good correction for –2 to –6 D of myopia.

3. *Laser in-situ keratomileusis (LASIK)*. In this technique first a flap of 130–160 micron thickness of anterior corneal tissue is raised. After creating a corneal flap midstromal tissue is ablated directly with an excimer laser beam, ultimately flattening the cornea. Currently this procedure is being considered the refractive surgery of choice for myopia of up to –12 D.

Patient selection criteria are:

– patients above 20 years of age;
– stable refraction for at least 12 months;
– motivated patient;
– absence of corneal pathology; presence of ectasia or any other corneal pathology and a corneal thickness less than 450 mm is an *absolute contraindication* for LASIK.

Advantages of LASIK:

– minimal or no postoperative pain;
– recovery of vision is very early as compared to PR;
– no risk of perforation during surgery and later rupture of globe due to trauma unlike RK;
– no residual haze unlike PRK where sub-epithelial scarring may occur;
– LASIK is effective in correcting myopia of –12 D.

Disadvantages:

1) LASIK is much more expensive;
2) it requires greater surgical skill than RK and PRK;
3) there is potential risk of flap related complications, which include:

– intraoperative flap amputation;
– wrinkling of the flap on repositioning;

– postoperative flap dislocation/subluxation;
– epithelization of flap-bed interface;
– irregular astigmatism.

4. *Extraction of clear crystalline lens* (Fucala's operation) has been advocated for myopia of –16 to –18 D. Refractive surgery for myopia of more than 12 D.

5. *Phakic intraocular lens or intraocular contact lens* (ICL) implantation is also being considered for correction of myopia of >12 D. In this technique, a special type of intraocular lens is implanted in the anterior chamber or posterior chamber anterior to the natural crystalline lens.

6. *Intercorneal ring* (ICR) implantation into the peripheral cornea at approximately 2/3 stromal depth is being considered. It results in a vaulting effect that flattens the central cornea, decreasing myopia. The ICR procedure has the advantage of being reversible.

7. *Orthokeratology* – a non-surgical reversible method of molding the cornea with overnight wear unique rigid gas permeable contact lenses, is also being considered for correction of myopia up to –5D. It can be used even in the patients below 18 year of age.

8. *Prophylaxis* (genetic counselling). As the pathological myopia has a strong genetic basis, the hereditary transfer of disease may be decreased by advising against marriage between two individuals with progressive myopia.

However, if they do marry, they should not produce children.

Hypermetropia

Hypermetropia (***hyperopia***) or long-sightedness is the refractive state of the eye wherein parallel rays of light coming from infinity are focused behind the retina with accommodation being at rest.

Thus, the posterior focal point is behind the retina, which therefore receives a blurred image. Hypermetropia may be axial, curvatural, index, positional and due to absence of lens.

Classification of hyperopia

Hyperopia is typically classified according to its clinical appearance, its severity, or how it relates to the eye's accommodative status.

Classification by clinical appearance:

– simple hyperopia;
– pathological hyperopia;
– functional hyperopia.

Classification by severity

Hyperopia is often categorized by the amount of refractive error:

– *low hyperopia* is a refractive error of +2.00 diopters (D) or less;

– *moderate hyperopia* is a refractive error from +2.25 to +5.00 D;

– *high hyperopia* is a refractive error of +5.25 D or more.

Classification by accommodative status:

– facultative hyperopia;
– absolute hyperopia;
– manifest hyperopia;
– latent hyperopia.

Symptoms

The main symptom is a difficulty with near vision. 'Tiring' of the eyes (*asthenopia*) is common and long-sighted people may have headaches and uncomfortable vision.

There may be difficulties with seeing with both eyes (binocular vision), as the brain will tend to ignore signals coming from the most long-sighted eye. Lazy eye (*amblyopia*) or squint (*strabismus*) can therefore also occur in long sight.

Long-sighted people may have difficulty with depth perception (3-dimensional vision), as this needs two eyes to work together, more or less equally.

Signs

1. *Size of eyeball* may appear small as a whole.

2. *Cornea* may be slightly smaller than the normal.

3. *Anterior chamber* is comparatively shallow.

4. *Fundus examination* reveals a small optic disc which may look more vascular with ill-defined margins and even may simulate papillitis (though there is no swelling of the disc, and so it is called pseudopapillitis). The retina as a whole may shine due to greater brilliance of light reflections (shot silk appearance).

5. *A-scan ultrasonography* (*biometry*) may reveal a short anteroposterior length of the eyeball.

Complications

If hypermetropia is not corrected for a long time, the following complications may occur:

1. *Recurrent sties, blepharitis* or *chalazia.*

2. *Accommodative convergent squint* may develop in children (usually by the age of 2–3 years) due to excessive use of accommodation.

3. *Amblyopia* may develop in some cases. It may be anisometropic (in unilateral hypermetropia), strabismus (in children developing accommodative squint) or ametropic (seen in children with uncorrected bilateral high hypermetropia).

Predisposition to develop primary narrow angle glaucoma

The eye in hypermetropes is small with a comparatively shallow anterior chamber.

Due to regular increase in the size of the lens with increasing age, these eyes become prone to an attack of narrow angle glaucoma. This point should be kept in mind while instilling mydriatics in elderly hypermetropes.

Optical treatment

Basic principle of treatment is to prescribe convex (plus) lenses (contact lenses and eyeglasses), so that the light rays are brought to focus on the retina.

Surgery treatment

PRK: the removal of a minimal amount of the corneal surface.

LASIK: laser eye surgery to reshape the cornea, so that glasses or contact lenses are no longer needed.

Refractive Lens Exchange: a variation of cataract surgery; the difference is the existence of abnormal ocular anatomy which causes a high refractive error.

LASEK: resembles PRK, but uses alcohol to loosen the corneal surface.

Aphakia

Aphakia literally means absence of crystalline lens from the eye. However, from the optical point of view, it may be considered a condition in which the lens is absent from the pupillary area. Aphakia produces a high degree of hypermetropia.

Causes

1. *Congenital* absence of lens. It is a rare condition.

2. *Surgical aphakia* occurring after removal of lens is the most common presentation.

3. *Aphakia due to absorption of lens matter* is noticed rarely after trauma in children.

4. *Traumatic extrusion* of lens from the eye also constitutes a rare cause of aphakia.

5. *Posterior dislocation* of lens in vitreous causes optical aphakia.

Optics of aphakic eye

The following optical changes occur after removal of crystalline lens:

1. Eye becomes highly hypermetropic.

2. Total power of eye is reduced to about +44 D from +60 D.

3. The anterior focal point becomes 23.2 mm in front of the cornea.

4. The posterior focal point is about 31 mm behind the cornea, i. e. about 7 mm behind the eyeball. The anteroposterior length of eyeball is about 24 mm).

5. There occurs total loss of accommodation.

Symptoms

1. *Defective vision.* The main symptom in aphakia is marked defective vision for both far and near due to high hypermetropia and absence of accommodation.

2. *Erythropsia and cyanopsia*, i. e., seeing red and blue images. This occurs due to excessive entry of ultraviolet and infrared rays in the absence of crystalline lens.

Signs

1. *Limbal scar* may be seen in surgical aphakia.

2. *Anterior chamber* is deeper than normal.

3. *Iridodonesis* i. e., tremulousness of iris can be demonstrated.

4. *Pupil* is jet black in colour.

5. *Purkinje's image test* shows only two images.

6. *Fundus examination* shows hypermetropic small disc.

7. *Retinoscopy* reveals high hypermetropia.

Treatment

Optical principle is to correct the error by convex lenses of appropriate power so that the image is formed on the retina.

Modalities for correcting aphakia include:

– contact lens;

– intraocular lens;

– refractive corneal surgery (*keratophakia, epikeratophakia, hyperopic LASIK).*

Pseudophakia

The condition of aphakia when corrected with an intraocular lens implant (IOL) is referred to as *pseudophakia* or *artephakia*.

Refractive status of a pseudophakic eye depends upon the power of the IOL implanted as follows:

1. *Emmetropia* is produced when the power of the implanted IOL is exact. It is the most ideal situation. Such patients need plus glasses for near vision only.

2. *Consecutive myopia* occurs when the implanted IOL overcorrects the refraction of eye. Such patients require glasses to correct the myopia for distance vision and may or may not need glasses for near vision depending upon the degree of myopia.

3. *Consecutive hypermetropia* develops when the underpower IOL is implanted. Such patients require plus glasses for distance vision and additional +2 to +3 D for near vision.

Note. Varying degree of surgically-induced astigmatism is also present in pseudophakia

Signs of pseudophakia (with posterior chamber IOL):

1. *Surgical scar* may be seen near the limbus.

2. *Anterior chamber* is slightly deeper than normal.

3. *Mild iridodonesis* (tremulousness) of iris may be demonstrated.

4. *Purkinje image test* shows four images.

5. *Pupil* is blackish in colour but when light is thrown in pupillary area shining reflexes are observed. When examined under magnification after dilating the pupil, the presence of IOL is confirmed.

6. *Visual status and refraction* will vary depending upon the power of IOL implanted as described above.

Astigmatism

Astigmatism is a type of refractive error wherein the refraction varies in the different meridian. Consequently, the rays of light entering in the eye cannot converge to a point focus but form focal lines. Broadly, there are two types of astigmatism: *regular and irregular.*

Causes

It's totally natural and most people are born with it. We don't know the exact cause. You can also get it after an eye injury, eye disease, or surgery. There's a myth that you can get it if you read in low light or sit too close to the TV, but that isn't true.

Regular astigmatism

The astigmatism is regular when the refractive power changes uniformly from one meridian to another (i. e., there are two principal meridians).

Etiology

1. *Corneal astigmatism* is the result of abnormalities of curvature of cornea. It constitutes the most common cause of astigmatism.

2. *Lenticular astigmatism* is rare. It may be:

– *curvatural;*
– *positional;*
– *index astigmatism.*

3. *Retinal astigmatism* due to oblique placement of macula may also be seen occasionally.

Types of regular astigmatism

Depending upon the axis and the angle between the two principal meridians, regular astigmatism can be classified into the following types:

1. *With-the-rule astigmatism.* Thus, correction of this astigmatism will require the concave cylinders at 180° ± 20° or convex cylindrical lens at 90° ± 20°.

2. *Against-the-rule astigmatism*. Therefore, correction of this astigmatism will require the prescription of convex cylindrical lens at 180° ± 20° or concave cylindrical lens at 90° ± 20° axis.

3. *Oblique astigmatism* is a type of regular astigmatism where the two principal meridians are not the horizontal and vertical.

4. *Bi-oblique astigmatism.* In this type of regular astigmatism the two principal meridians are not at right angle to each other, e. g., one may be at 30° and other at 100°.

Refractive types of regular astigmatism

Depending upon the position of the two focal lines in relation to retina, the regular astigmatism is further classified into three types:

1. *Simple astigmatism*, wherein the rays are focused on the retina in one meridian and either in front or behind the retina in the other meridian.

2. *Compound astigmatism.* In this type the rays of light in both the meridians are focused either in front or behind the retina and the condition is labelled as *compound myopic* or *compound hypermetropic astigmatism*, respectively.

3. *Mixed astigmatism* refers to a condition wherein the light rays in one meridian are focused in front and in other meridian behind the retina. Thus, in one meridian the eye is myopic and in another – hypermetropic. Such patients have comparatively less symptoms as '*circle of least diffusion*' is formed on the retina.

Symptoms of regular astigmatism include:

– defective vision;

– blurring of objects;

– depending upon the type and degree of astigmatism, objects may appear proportionately elongated;

– asthenopic symptoms, which are marked especially in small amount of astigmatism, consist of a dull ache in the

eyes, headache, early tiredness of eyes and sometimes nausea and even drowsiness.

Investigations

Basic examination: observation of external structures, extraocular movements and cranial nerves, dilating the pupil, visual acuity, refractometry, slit-lamp examination, intraocular pressure (IOP), visual field testing, ophthalmoscopy.

Keratometry and *computerized corneal topotograpy* reveal different corneal curvature in two different meridians in corneal astigmatism.

Astigmatic fan test and *Jackson's cross cylinder test.* These tests are useful in confirming the power and axis of cylindrical lenses.

Treatment

1. *Optical treatment* of regular astigmatism comprises the prescribing appropriate cylindrical lens, discovered after accurate refraction.

Spectacles with full correction of cylindrical power and appropriate axis should be used for distance and near vision.

Contact lenses: rigid contact lenses may correct up to 2–3 of regular astigmatism, while soft contact lenses can correct only little astigmatism. For higher degrees of astigmatism toric contact lenses are needed. In order to maintain the correct axis of toric lenses, ballasting or truncation is required.

2. *Surgical correction of astigmatism* is quite effective.

Irregular astigmatism

It is characterized by an irregular change of refractive power in different meridians. There are multiple meridians which admit no geometrical analysis.

Etiology

Irregular astigmatism can result from conditions such as keratoconus or other naturally occurring corneal degenerations, corneal injuries, refractive surgery.

Symptoms of irregular astigmatism include:

– distortion or blurring of images at all distances;
– headache and fatigue;
– squinting and eye discomfort or irritation.

Investigations

Basic examination: observation of external structures, extra ocular movements and cranial nerves, dilating the pupil, visual acuity, refractometry, slit-lamp examination, intraocular pressure (IOP), visual field testing, ophthalmoscopy.

Photokeratoscopy and computerized corneal topography give photographic record of irregular corneal curvature.

Treatment

1. *Optical treatment* of irregular astigmatism consists of contact lens which replaces the anterior surface of the cornea for refraction.

2. *Phototherapeutic keratectomy* (PTK) performed with excimer laser may be helpful in patients with superficial corneal scar responsible for irregular astigmatism.

3. *Surgical treatment* is indicated in extensive corneal scarring (when vision does not improve with contact lenses) and consists of penetrating keratoplasty.

Anisometropia

The optical state with equal refraction in the two eyes is termed *isometropia.* When the total refraction of the two eyes is unequal, the condition is called *anisometropia.* Small degree of anisometropia is of no concern.

A difference of 1 D in two eyes causes a 2 percent difference in the size of the two retinal images. A difference up to 5 percent in retinal images of two eyes is well tolerated. In other words, an anisometropia up to 2.5 is well tolerated and that between 2.5 and 4 D can be tolerated depending upon the

individual sensitivity. However, if it is more than 4 D, it is not tolerated and is a matter of concern.

Etiology

1. *Congenital and developmental anisometropia* occurs due to differential growth of the two eyeballs.

2. *Acquired anisometropia* may occur due to uniocular aphakia after removal of cataractous lens or due to implantation of IOL of wrong power.

Clinical types

1. *Simple anisometropia*. In this, one eye is normal (emmetropic) and the other either myopic (simple myopic anisometropia) or hypermetropic (simple hypermetropic anisometropia).

2. *Compound anisometropia.* Wherein both eyes are either hypermetropic (compound hypermetropic anisometropia) or myopic (compound myopic anisometropia), but one eye is having higher refractive error than the other.

3. *Mixed anisometropia*. In this, one eye is myopic and the other is hypermetropic. This is also called *anti-metropia.*

4. *Simple astigmatic anisometropia*. When one eye is normal and the other has either simple myopic or hypermetropic astigmatism.

5. *Compound astigmatic anisometropia*. Both eyes are astigmatic but of unequal degree.

Anomalies of Accommodation

Anomalies of accommodation are not uncommon. These include:

- presbyopia;
- insufficiency of accommodation;
- paralysis of accommodation;
- spasm of accommodation.

Presbyopia

Presbyopia (eye sight of old age) is not an error of refraction but a condition of physiological insufficiency of accommodation leading to a progressive fall in near vision.

Causes

Decrease in the accommodative power of crystalline lens with increasing age, leading to presbyopia, occurs due to:

1. *Age-related changes in the lens* which include:

– decrease in the elasticity of lens capsule;

– progressive increase in size and hardness (sclerosis) of lens substance which is less easily molded.

2. *Age related decline in ciliary muscle power* may also contribute in causation of presbyopia.

Causes of premature presbyopia are:

1. Uncorrected hypermetropia.

2. Premature sclerosis of the crystalline lens.

3. General debility causing pre-senile weakness of ciliary muscle.

4. Chronic simple glaucoma.

Symptoms

1. *Difficulty in near vision.* Patients usually complaint of difficulty in reading small prints (to start with, in the evening and in dim light, and later even in good light). Another important complaint of the patient is difficulty in threading a needle, etc.

2. *Asthenopic symptoms* due to fatigue of the ciliary muscle are also complained after reading or doing any near work.

Investigations

Basic examination: observation of external structures, dilating the pupil, visual acuity, slit-lamp examination, intraocular pressure (IOP), visual field testing, and ophthalmoscopy.

Treatment

Optical treatment: the treatment of presbyopia is the prescription of appropriate convex glasses for near work. A rough guide for providing presbyopic glasses in an emmetrope can be made from the age of the patient about:

1) +1 D is required at the age of 40–45 years;
2) +1.5 D at 45–50 years;
3) + 2 D at 50–55 years;
4) +2.5 D at 55–60 years.

However, the presbyopic add should be estimated individually in each eye in order to determine how much is necessary to provide a comfortable range.

Basic principles for presbyopic correction are:

1. Always find out refractive error for distance and first correct it.

2. Find out the presbyopic correction needed in each eye separately and add it to the distant correction.

3. Near point should be fixed by taking due consideration for profession of the patient.

4. The weakest convex lens with which an individual can see clearly at the near point should be prescribed, since overcorrection will also result in asthenopic symptoms.

Presbyopic spectacles may be unifocal, bifocal or varifocal.

Surgical treatment of presbyopia is still in infancy.

Insufficiency of Accommodation

The term insufficiency of accommodation is used when the accommodative power is significantly less than the normal physiological limits for the patient's age. Therefore, it should not be confused with presbyopia in which the physiological insufficiency of accommodation is normal for the patient's age.

Causes

1. Premature sclerosis of lens.

2. Weakness of ciliary muscle due to systemic causes of muscle fatigue such as debilitating illness, anemia, toxemia, malnutrition, diabetes mellitus, pregnancy, stress and so on.

3. Weakness of ciliary muscle associated with primary open-angle glaucoma.

Clinical features

All the symptoms of presbyopia are present, but those of asthenopia are more prominent than those of blurring of vision.

Treatment

1. The treatment is essentially that of the systemic cause.

2. *Near vision spectacles* in the form of weakest convex lens which allows adequate vision should be given till the power of accommodation improves.

3. *Accommodation exercises* help in recovery, if the underlying debility has passed.

Paralysis of Accommodation

Paralysis of accommodation also known as *cycloplegia* refers to complete absence of accommodation.

Causes

1. *Drug induced cycloplegia* results due to the effect of atropine, tropicamide, homatropine or other parasympatholytic drugs.

2. *Internal ophthalmoplegia* (paralysis of ciliary muscle and sphincter pupillae) may result from neuritis associated with diphtheria, syphilis, diabetes, alcoholism, cerebral or meningeal diseases.

3. *Paralysis of accommodation as a component of complete third nerve paralysis* may occur due to intracranial or orbital causes. The lesions may be traumatic, inflammatory or neoplastic in nature.

Clinical features

1. *Blurring of near vision.* It is the main complaint in previously emmetropic or hypermetropic patients. Blurring of near vision may not be marked in myopic patients.

2. *Photophobia* (glare) due to accompanying dilatation of pupil (mydriasis) is usually associated with blurring of near vision.

3. Examination reveals abnormal receding of near point and markedly decreased range of accommodation.

Treatment

1. Self-recovery occurs in drug-induced paralysis and in diphtheric cases (once the systemic disease is treated).

2. Dark-glasses are effective in reducing the glare.

3. Convex lenses for near vision may be prescribed if the paralysis is permanent.

Spasm of Accommodation

Spasm of accommodation refers to exertion of abnormally excessive accommodation.

Causes

1. *Drug induced spasm* of accommodation is known to occur after use of strong miotics such as echothiophate and DFP.

2. *Spontaneous spasm* of accommodation. It usually occurs when the eyes are used for excessive near work in unfavourable circumstances such as bad illumination, bad reading position, lowered vitality, state of neurosis, mental stress or anxiety.

Clinical features

1. Defective vision due to induced myopia.

2. Asthenopic symptoms are more marked than the visual symptoms.

Diagnosis is made with refraction under atropine.

Treatment

1. Relaxation of ciliary muscle by atropine for a few weeks and prohibition of near work allow prompt recovery from spasm of accommodation.

2. Correction of associated causative factors prevents recurrence.

TOPIC 4. DISEASES OF EYELIDS AND ORBIT

Classification of eyelid diseases:

– inflammatory as *stye*, *chalazion*, etc;
– eyelid deformity as *entropion*, *ectropion*, etc;
– ptosis;
– miscellaneous subjects;
– tumours of the eyelid;
– congenital eyelid anomalies.

The inflammatory eyelid diseases are:

1. Stye.
2. Chalazion.
3. Infected chalazion.
4. Blepharitis.
5. Meibomianitis.

Stye (Hordeolum Externum)

A *hordeolum* (i. e. *stye*) is an infection or inflammation of the eyelid margin involving hair follicles of the eyelashes (i. e. external hordeolum) or meibomian glands (i. e. internal hordeolum).

Etiology

Staphylococcus aureus is the agent of infection in 90–95 % of hordeolum cases.

Risk factors

– chronic diseases;
– health or poor endurance;
– chronic inflammation of the eyelids, such as blepharitis;
– diabetes;
– hyperlipidemia, including hypercholesterolemia;
– history of previous hordeolum;
– hygiene and clean environment;
– skin conditions such as seborrheic dermatitis.

Symptoms and signs

Symptoms: swelling, pain in the eyelid, feelings of discomfort and burning sensation in the eyelid, history of the same disease.

Signs: erythema, edema, pain when pressed near the base of the eyelashes, as small abscess picture.

Treatment: usually hordeolum can heal itself within 5–7 days.

Generally: warm compresses 4–6 times a day for 15 minutes each time to help the drainage. Do it with your eyes closed. Clean your eyelids with clean water or with soap or shampoo that does not cause irritation, such as baby soap. It can speed up the healing process.

Do not press or puncture hordeolum, this can lead to more serious infections. Avoid using makeup on the eyes, because of the possibility that it can cause infection.

Do not wear contact lenses because it can spread the infection to the cornea.

Topical antibiotics: bacitracin ointment or eye tobramicin should be given every 4 hours for 7–10 days. You can also give erythromycin eye ointment for external hordeolum.

Systemic antibiotics: given if there are signs of bacteremia or the signs of enlarged lymph nodes in the pre-auricular.

In the case of hordeolum internum with moderate to severe cases: Dicloxacilin can be given or cephalexin 500 mg orally four times daily for 7 days. If allergic to penicillin or cephalosporin, clindamycin 300 mg can be administered orally four times daily for 7 days, or claritromycin 500 mg 2 times daily for 7 days.

Surgery: if the treatment does not respond well, then a surgical procedure may be required to make drainage of hordeolum.

At incision of hordeolum, a topical anesthetic is given prior to ophthalmic alcaine.

External incisions may lead to scarring, so making external eyelid incisions or punctures is less desirable. In this case, the incision should be made through a lid crease if possible.

Internal incisions may be made vertically to reduce the area of the cornea swept by the irregular healing tissue during blinking and prevent lid contracture that may contribute to malposition; external incisions may be made horizontally for optimal post-operative cosmesis.

The incision is left open with clean margins to allow for drainage of any residual material within the lesion.

Complications: the most common is one lash trichiasis and the most serious is cavernous sinus thrombosis.

Differential diagnosis: infected chalazion (see below) and marginal chalazion. It is a chronic painless swelling behind the grey line, while stye is an acute painful swelling in front of the grey line.

Chalazion (Meibomian Cyst)

Chalazion or *meibomian cyst* is chronic lipogranulomatous inflammation of one of the meibomian glands.

Etiology

Chalazion occurs after gland blockage, which can be associated with the following: poor eyelid hygiene, seborrheic dermatitis, rosacea, chronic blepharitis, high blood lipid concentrations, leishmaniasis, tuberculosis, immunodeficiency, viral infection, carcinoma, stress (casualty has not been proven, and the mechanism by which it might act is unknown), trachoma, eyelid trauma and eyelid surgery.

Symptoms and signs

Symptoms: painless localized swelling of the eyelid of accidental onset and gradual increase in size.

Signs

1. Tarsal chalazion:

– single or multiple well localized non tender swellings can be easily palpated in the lid substance (it is easier to be felt than seen);

– it is not attached to skin and disappears by contraction of orbicularis oculi;

– from the conjunctival side, the chalazion appears as a pale area surrounded by hyperemia.

2. Marginal chalazion:

– it is a small swelling in the lid margin behind the grey line.

Complications:

– infection leading to acute chalazion (painful);

– opening into the conjunctiva with incomplete evacuation will lead to the formation of chalazion granuloma; this will lead to chronic conjunctivitis;

– pressure on the cornea by a large chalazion causing astigmatism.

Differential diagnosis:

– adenoma or adenocarcinoma of Meibomian gland; there is recurrence in the same site. In such cases, histopathological diagnosis should be done;

– sebaceous cyst: it is attached to skin and becomes more apparent by contraction of orbicularis oculi;

– marginal chalazion should be differentiated from stye (see above).

Treatment

Small chalazion: local steroid ointment for 1 week (usually not effective).

Tarsal chalazion: it can be removed by the following ways:

– local anaesthesia by topical novosine eye drops plus infiltration of novocain in submuscular layer;

– hold the chalazion with the chalazion forceps; it is important for fixation of the chalazion, hemostasis, protection of the globe and lid evertion;

– excise the conjunctival wall of chalazion to prevent recurrence;

– curettage to ensure complete evacuation to prevent chalazion granuloma formation;

– bandage for few hours and eye ointment that contains both steroids and antibiotics should be used for few days.

Multiple chalazions are removed in different sessions and vitamin A is used for support.

Recurrent chalazion: in the same site we do excision biopsy. In different sites we give vitamin A.

Marginal chalazion: shaving by scalpel while the eye is protected by the lid spatula.

Infected chalazion: antibiotics, and the drainage of pus is performed.

Infected Chalazion (Hordiolum Internum)

It is a suppurative inflammation of the meibomian gland associated with blockage of the duct.

Etiology

It may occur as primary staphylococcal infection of the meibomian gland or due to secondary infection in a chalazion (infected chalazion).

Symptoms and signs

Symptoms: dull aching pain and diffuse swelling of the lid; throbbing pain (pus under tension), and a localized lid swelling.

Signs: lid swelling that disappears by lid closure and is not related to lashes, maximal tenderness over the infected gland, the underlying chalazion can be felt, pus points under the conjunctiva in the localized stage.

Complications:

– opens spontaneously into the conjunctiva leading to incomplete evacuation of its contents so that chalazion granuloma develops;

– cavernous sinus thrombosis.

Treatment

It is similar to hordeolum externum, except when the pus is formed, it should be drained by a vertical incision from the tarsal conjunctiva.

Molluscum Contagiosum

It is a viral infection of the lids, commonly affecting children. It is caused by a large poxvirus. Its typical lesions are multiple, pale, waxy, umbilicated swellings scattered over the skin near the lid margin.

These may be *complicated* by chronic follicular conjunctivitis and superficial keratitis.

Treatment

The skin lesions should be incised and the interior cauterized with tincture of iodine or pure carbolic acid.

Blepharitis

It is a subacute or chronic inflammation of the lid margins. It is an extremely common disease which can be divided into the following clinical types:

– seborrheic or squamous blepharitis;
– staphylococcal or ulcerative blepharitis;
– mixed staphylococcal with seborrheic blepharitis;
– posterior blepharitis or meibomitis;
– parasitic blepharitis.

Seborrheic (simple) blepharitis

It is the chronic inflammation of the 4 lid margins characterized by the formation of scales formed of dried sebaceous secretion.

Etiology

It is usually associated with seborrhea of scalp (dandruff). Some constitutional and metabolic factors play a part in its etiology. The glands of Zeiss secrete abnormal excessive neutral lipids which are split by *Corynebacterium acne* into irritating free fatty acids.

Symptoms and signs:

– itching, discomfort and scanty discharge;

– small scales like dandruff are seen on and in-between the lashes;

– removal of these scales leaves un ulcerated hyperemic lid margin;

– dandruff in the scalp and eyebrows confirm the diagnosis.

Treatment

1. *Lid hygiene:* the lid margins are scrubbed with neutral pH shampoo or special preparations. Also seborrhea of scalp and brows should be treated.

2. *Antibiotic drops* – 1 % oxytetracycline, 0.3 % tobramycin, 0.3 % gentamycin, 0.3 % floxal eye ointment and eye drops 2–3 times a day for 9–14 days will eradicate the infection.

3. *Local steroids* (dexamethasone or betamethasone) are embrocated with the lashes only in severe cases.

4. *NSAID-diclofenac, nimesil, ibuprofen or voltaren* may be given orally for 2–3 days.

Ulcerative blepharitis

It is a chronic staphylococcal infection of the lid margin usually caused by coagulase positive strains. The disorder usually starts in childhood and may continue throughout life.

Etiology

Chronic conjunctivitis and dacryocystitis may act as predisposing factors.

Symptoms and signs

Symptoms: hyperemia, red shedding of lashes, crusts, severe itching, burning sensation, lacrimation, photophobia and blepharospasm.

Signs:

– the lid margins are congested and edematous with crustations;

– the crusts may form collarettes surrounding the lashes;

– removal of the crusts leaves bleeding ulcers;

– lashes are glued together.

Complications and sequelae

These are seen in longstanding (non-treated) cases and include chronic conjunctivitis, madarosis (sparseness or absence of lashes), trichiasis, poliosis (greying of lashes), tylosis (thickening of lid margin) and eversion of the punctum leading to epiphora. Eczema of the skin and ectropion may develop due to prolonged watering. Recurrent styes are very common.

Treatment

It should be treated promptly to avoid complication and sequelae. *Crusts should be removed* after softening and hot compresses with solution of 3 percent soda bicarb.

Antibiotic ointment (tobramycin, ofloxacin, gentamicin, erythromycin or tetracyclines) should be applied at the lid margin, immediately after removal of crusts, at least twice daily.

Antibiotic eyedrops – tobradex (tobramycin), tobrex-2x (tobramycin), ciprofarm (ciprofloxacin) gentamicin or erythromycin should be instilled 3–4 times a day. Avoid rubbing of the eyes or fingering of the lids.

Oral anti-inflammatory drugs (diclofenac, indomethacin, voltaren, nimesil, ibuprofen) can be used to help in reducing the inflammation.

Posterior blepharitis (meibomitis)

1. *Chronic meibomitis* is a meibomian gland dysfunction, seen more commonly in middle-aged persons with acne rosacea and seborrhoeic dermatitis.

It is characterized by white frothy (foam-like) secretion on the eyelid margins and canthi (meibomian seborrhea). On eversion of the eyelids, vertical yellowish streaks shining through the conjunctiva are seen. At the lid margin, openings of the meibomian glands become prominent with thick secretions.

2. *Acute meibomitis* occurs mostly due to staphylococcal infection.

Treatment of meibomitis consists of *expression* of the glands by repeated vertical lid massage, followed by embrocating the lid margin with *antibiotic-steroid ointment*.

Antibiotic ointment (tobramycin, ofloxacin, gentamicin, erythromycin or tetracycline) should be applied at the lid margin, immediately after removal of crusts, at least twice daily.

Antibiotic eyedrops – tobradex (tobramycin), tobrex-2x (tobramycin), ciprofarm (ciprofloxacin) gentamicin or erythromycin should be instilled 3–4 times a day. Avoid rubbing of the eyes or fingering of the lids.

Oral anti-inflammatory drugs (diclofenac, indomethacin, voltaren, nimesil, and ibuprofen) help in reducing the inflammation.

Parasitic blepharitis

Blepharitis acrica refers to a chronic blepharitis associated with Demodex folliculorum infection and Phthiriasis palpebram due to the crab-louse, very rarely – to the head-louse. In addition to features of chronic blepharitis, it is characterized by presence of nits at the lid margin and at roots of eyelashes

Symptoms and signs: itching and lacrimation; the parasite is seen on the lashes and the black nests of eggs are also seen.

Treatment

Demodicosis of the eyelids as a rule lasts for a few months. The use of yellow mercurial ointment, sulphur ointment, camphorated oil, crotamiton, choline esterase inhibitors, sulfacetamide, steroids, antibiotics, as well as antimycotic drugs offers some improvement. A good response has been observed after oral application of ivermectin along with topical application of cream permethrin. However, the best results were obtained after applying 2 % metronidazole gel or ointment treatment.

Deformities of the Lid

This group of diseases includes:

– trichiasis;
– entropion;
– ectropion;
– symblepharon;
– ankyloblepharon;
– blepharophimosis;
– lagophthalmos;
– blepharospasm.

Trichiasis

It refers to inward misdirection of cilia (which rub against the eyeball) with normal position of the lid margin. The inward turning of lashes along with the lid margin (seen in entropion) is called *pseudotrichiasis.*

Etiology

Common causes of trichiasis are: cicatrizing trachoma, ulcerative blepharitis, healed membranous conjunctivitis, hordeolum externum, mechanical injuries, burns, and operative scars on the lid margin.

Symptoms and signs

These include foreign body sensation and photophobia. Patient may feel troublesome irritation, pain and lacrimation.

Complications

These include recurrent corneal abrasions, superficial corneal opacities, corneal vascularization and non-healing corneal ulcer.

Treatment

A few misdirected cilia may be treated by any of the following methods:

1. *Epilation* (mechanical removal with forceps): it is a temporary method, as recurrence occurs within 3–4 weeks.

2. *Electrolysis*: it is a method of destroying the lash follicle by electric current. In this technique, infiltration anesthesia is given to the lid and a current of 2 mA is passed for 10 seconds through a fine needle inserted into the lash root. The loosened cilia with destroyed follicles are then removed with epilation forceps.

3. *Cryoepilation*: it is also an effective method of treating trichiasis. After infiltration anaesthesia, the cryoprobe (–20 °C) is applied for 20–25 seconds to the external lid margin. Its main disadvantage is depigmentation of the skin.

4. *Surgical correction*: when many cilia are misdirected operative treatment similar to cicatricial entropion should be employed.

Entropion

It is inturning of the lid margin.

Types of entropion:

– cicatricial entropion;
– spastic entropion;
– senile (involutional) entropion;
– congenital entropion;
– mechanical entropion.

Symptoms and signs

Occur due to rubbing of cilia against the cornea and conjunctiva and are thus similar to trichiasis. These include foreign body sensation, irritation, lacrimation and photophobia. On examination, lid margin is found inturned. Depending upon the degree of its turning, it can be divided into three grades:

Grade I – only the posterior lid border is enrolled.

Grade II – entropion includes inturning up to the inter-marginal strip.

Grade III – the whole lid margin including the anterior border is inturned.

Complications

These are similar to trichiasis

Treatment

Surgical treatment

Ectropion

It is outward rolling of the lid margin, so that the posterior lid margin will not be in contact with the surface of the eyeball. This malposition will disturb tear spreading over the surface of cornea and conjunctiva. Also the lacrimal

punctum will not be in contact with tears, so that epiphora will occur.

Types of ectropion:

– senile ectropion;
– paralytic ectropion;
– cicatricial ectropion;
– mechanical ectropion;
– spastic ectropion.

Symptoms

1. Watering of the eye (epiphora).

2. Burning sensation due to incomplete lid closure (lagophthalmos) in severe cases.

3. Cosmetic disfigurement in severe cases.

Signs

1. The posterior lid margin is seen to be rolled outward away from the globe, so that the punctum can be seen.

2. Assessment of the degree of ectropion:

– in mild ectropion the punctum can be seen;
– in moderate ectropion the palpebral conjunctiva can be seen;
– in severe ectropion the lower fornix can be seen.

Complication

Prolonged exposure may cause dryness and thickening of the conjunctiva and corneal ulceration (exposure keratitis). Eczema and dermatitis may occur due to prolonged epiphora.

Treatment

Surgical treatment

Symblepharon

In this condition lids become adherent with the eyeball as a result of adhesions between the palpebral and bulbar conjunctiva.

It is characterised by difficulty in lid movements, diplopia (due to restricted ocular motility), inability to close the lids (lagophthalmos) and cosmetic disfigurement.

Fibrous adhesions between palpebral conjunctiva and the bulbar conjunctiva and/or cornea may be present only in the anterior part (*anterior symblepharon*), or fornix (*posterior symblepharon*) or the whole lid (*total symblepharon*).

Treatment

1. *Prophylaxis.* During the stage of raw surfaces, the adhesions may be prevented by sweeping a glass rod coated with lubricant around the fornices several times a day. A large-sized, therapeutic, soft contact lens also helps in preventing the adhesions.

2. *Curative treatment* consists of symblepharectomy. The raw area created may be covered by mobilising the surrounding conjunctiva in mild cases. Conjunctival or buccal mucosal graft is required in severe cases.

Ankyloblepharon

It refers to the adhesions between margins of the upper and lower lids. It may occur as a congenital anomaly or may result after healing of chemical burns, thermal burns, ulcers and traumatic wounds of the lid margins. Ankyloblepharon may be complete or incomplete. It is usually associated with symblepharon.

Treatment

Lids should be separated by excision of adhesions between the lid margins and kept apart during the healing process. When adhesions extend to the angles, epithelial grafts should be given to prevent recurrences.

Blepharophimosis

In this condition the extent of the palpebral fissure is decreased. It appears contracted at the outer canthus.

Etiology

It may be congenital or acquired, due to formation of a vertical skin fold at the lateral canthus (epicanthus lateralis) following eczematous contractions.

Treatment

Usually no treatment is required. In marked cases, canthoplasty operation is performed.

Lagophthalmos

This condition is characterized by inability to voluntarily close the eyelids.

Etiology

It occurs in patients with paralysis of orbicularis oculi muscle, cicatricial contraction of the lids, symblepharon, severe ectropion, proptosis, following over-resection of the levator muscle for ptosis, and in comatose patients. Physiologically some people sleep with their eyes open (nocturnal lagophthalmos)

Clinical picture

It is characterized by incomplete closure of the palpebral aperture associated with features of the causative disease.

Complications include conjunctival and corneal xerosis and exposure keratitis.

Treatment

To prevent exposure keratitis artificial tear drops should be instilled frequently and the open palpebral fissure should be filled with an antibiotic eye ointment during sleep and in comatose patients. Soft bandage contact lens may be used to prevent exposure keratitis.

Blepharospasm

It refers to the involuntary, sustained and forceful closure of the eyelids.

Etiology

Blepharospasm occurs in two forms:

1. *Essential (spontaneous) blepharospasm.* It is a rare idiopathic condition involving patients between 45 and 65 years of age.

2. *Reflex blepharospasm.* It usually occurs due to reflex sensory stimulation through branches of the fifth nerve, in conditions such as: phlyctenular keratitis, interstitial keratitis, corneal foreign body, corneal ulcers and iridocyclitis. It is also seen in excessive stimulation of retina by dazzling light, stimulation of facial nerve due to central causes and in some hysterical patients.

Clinical features

Persistent epiphora may occur due to spasmodic closure of the canaliculi which may lead to eczema of the lower lid. Oedema of the lids is of frequent occurrence. Spastic entropion (in elderly people) and spastic ectropion (in children and young adults) may develop in long-standing cases.

Blepharophimosis may result due to contraction of the skin folds following eczema.

Treatment

In essential blepharospasm botulinum toxin, injected subcutaneously over the orbicularis muscle, blocks the neuromuscular junction and relieves the spasm. Facial denervation may be required in severe cases. *In reflex blepharospasm*, the causative disease should be treated to prevent recurrences. Associated complications should also be treated.

Ptosis

Abnormal drooping of the upper eyelid is called *ptosis*.

Normally, upper lid covers about upper one-sixth of the cornea, i. e. about 2 mm. Therefore, in ptosis it covers more than 2 mm.

Types and etiology

I. *Congenital ptosis*

It is associated with congenital weakness (maldevelopment) of the levator palpebrae superioris. It may occur in the following forms:

1. *Simple congenital ptosis* (not associated with any other anomaly).

2. Congenital ptosis with associated weakness of superior rectus muscle.

3. As a part of *blepharophimosis syndrome*, comprising congenital ptosis, blepharophimosis, telecanthus and epicanthus inversus.

4. *Congenital synkinetic ptosis* (Marcus Gunn jaw-winking ptosis). In this condition there occurs retraction of the ptotic lid with jaw movements, i. e., with stimulation of ipsilateral pterygoid muscle.

II. *Acquired ptosis*

Depending upon the cause it can be neurogenic, myogenic, aponeurotic or mechanical.

1. *Neurogenic ptosis:* it is caused by innervational defects such as third nerve palsy, Horner's syndrome, ophthalmoplegic migraine and multiple sclerosis.

2. *Myogenic ptosis:* it occurs due to acquired disorders of the levator palpebrae superioris muscle or of the myoneural junction. It may be seen in patients with myasthenia gravis, dystrophia myotonica, ocular myopathy, oculo-pharyngeal muscular dystrophy and following trauma to the levator palpebrae superioris muscle.

3. *Aponeurotic ptosis:* it develops due to defects of the levator aponeurosis in the presence of a normal functioning muscle. It includes involutional (senile) ptosis, postoperative ptosis (which is rarely observed after cataract and retinal detachment surgery), ptosis due to aponeurotic weakness

associated with blepharochalasis, and in traumatic dehiscence or disinsertion of the aponeurosis.

4. *Mechanical ptosis:* it may result due to excessive weight on the upper lid as seen in patients with lid tumours, multiple chalazia and lid oedema. It may also occur due to scarring (cicatricial ptosis) as seen in patients with ocular pemphigoid and trachoma.

Measurement of amount (degree) of ptosis

In unilateral cases, difference between the vertical heights of the palpebral fissures of the two sides indicates the degree of ptosis. In bilateral cases it can be determined by measuring the amount of cornea covered by the upper lid and then subtracting 2 mm.

Depending upon its amount the ptosis is graded as:

- mild 2 mm;
- moderate 3 mm;
- severe 4 mm.

Treatment

I. *Congenital ptosis:* it almost always needs surgical correction. In severe ptosis, surgery should be performed at the earliest to prevent stimulus deprivation amblyopia. However, in mild and moderate ptosis, surgery should be delayed until the age of 3–4 years, when accurate measurements are possible.

II. *Acquired ptosis:* efforts should be made to find out the underlying cause and if possible treat it. In neurogenic ptosis conservative treatment should be carried out and surgery deferred at least for 6 months. Surgical procedures (when required) are essentially the same as described for congenital ptosis. However, the amount of levator resection required is always less than the congenital ptosis of the same degree.

Tumours of the Lids

Almost all types of tumours arising from the skin, connective tissue, glandular tissue, blood vessels, nerves and muscles can involve the lids. A few common tumours are listed and only the important ones are described here.

Classification

1. *Benign tumours:* these include simple papilloma, nevus, angioma, haemangioma, neurofibroma and sebaceous adenoma.

2. *Pre-cancerous conditions:* these are solar keratosis, carcinoma-in-situ and xeroderma pigmentosa.

3. *Malignant tumours:* commonly observed tumours include squamous cell carcinoma, basal cell carcinoma, malignant melanoma and sebaceous gland adenocarcinoma.

Congenital Anomalies

1. *Congenital ptosis:* it is a common congenital anomaly. It is described in detail in the section of ptosis.

2. *Congenital coloboma:* it is a rare condition characterized by a full thickness triangular gap in the tissues of the lids.

Treatment consists of plastic repair of the defect.

3. *Epicanthus:* it is a semicircular fold of skin which covers the medial canthus. It is the most common congenital anomaly of the lids.

Treatment consists of plastic repair of the deformity.

4. *Distichiasis.* *Congenital distichiasis* is a rare anomaly in which an extra row of cilia occupies the position of Meibomian glands which open into their follicles as ordinary sebaceous glands. These cilia are usually directed backwards and when rubbing the cornea, should be electro epilated or cryoepilated.

5. *Cryptophthalmos:* it is a very rare anomaly in which lids fail to develop and the skin passes continuously from the eyebrow to the cheek hiding the eyeball

6. *Microblepharon:* in this condition, eyelids are abnormally small. It is usually associated with microphthalmos or anophthalmos. Occasionally the lids may be very small or virtually absent and the condition is called *ablepharon.*

Proptosis

Proptosis is protrusion of the eyeball. *Exophthalmos* means the same thing, and this term is usually used when describing proptosis due to Graves. Disorders that may cause changes in the appearance of the face and eyes that resemble proptosis but are not, include hyperthyroidism without infiltrative eye disease, Cushing disease, and severe obesity.

Classification

– unilateral proptosis;
– bilateral proptosis;
– acute proptosis;
– intermittent proptosis;
– pulsating proptosis.

Etiology

Important causes of proptosis in each clinical group are listed here:

A. *Causes of unilateral proptosis* include:

1. *Congenital conditions.* These include: dermoid cyst, congenital cystic eyeball, and orbital teratoma.

2. *Traumatic lesions*. These are: orbital haemorrhage, retained intraorbital foreign body, traumatic aneurysm and emphysema of the orbit.

3. *Inflammatory lesions.* Acute inflammations are orbital cellulitis, abscess, thrombophlebitis, panophthalmitis, and cavernous sinus thrombosis (proptosis is initially unilateral but ultimately becomes bilateral). Chronic inflammatory

lesions include: pseudotumours, tuberculoma, gumma and sarcoidosis.

4. *Circulatory disturbances and vascular lesions.* These are: angioneurotic oedema, orbital varix and aneurysms.

5. *Cysts of orbit.* These include: haematic cyst, implantation cyst and parasitic cyst (hydatid cyst and cysticercus cellulosae).

6. *Tumours of the orbit.* These can be primary, secondary or metastatic.

7. *Mucoceles of paranasal sinuses,* especially frontal (most common), ethmoidal and maxillary sinus are common causes of unilateral proptosis.

B. *Causes of bilateral proptosis* include:

1. *Developmental anomalies of the skull*: craniofacial dysostosis, e. g., oxycephaly (tower skull).

2. *Osteopathies*: osteitis deformans, rickets and acromegaly.

3. *Inflammatory conditions*: Mikulicz's syndrome and late stage of cavernous sinus thrombosis.

4. *Endocrinal exophthalmos*: it may be thyrotoxic or thyrotropic.

5. *Tumours*: these include symmetrical lymphoma or lymphosarcoma, secondary form neuroblastoma, nephroblastoma, Ewing's sarcoma and leukemic infiltration.

6. *Systemic diseases*: histiocytosis, systemic amyloidosis, xanthomatosis and Wegener's granulomatosis.

C. *Causes of acute proptosis.* It develops with extreme rapidity (sudden onset). Its common causes are: orbital emphysema fracture of the medial orbital wall, orbital haemorrhage and rupture of ethmoidal mucocele.

D. *Cause of intermittent proptosis.* This type of proptosis appears and disappears of its own. Its common causes are: orbital varix, periodic orbital oedema, recurrent orbital haemorrhage and highly vascular tumours.

E. ***Causes of pulsating proptosis.*** It is caused by pulsating vascular lesions such as caroticocavernous fistula and saccular aneurysm of ophthalmic artery. Pulsating proptosis also occurs due to transmitted cerebral pulsations in conditions associated with deficient orbital roof. These include congenital meningocele or meningoencephalocele, neurofibromatosis and traumatic or operative hiatus.

Investigation of a case of proptosis

I. ***Clinical evaluation***

A. *History*. It should include: age of onset, nature of onset, duration, progression, chronology of orbital signs and symptoms and associated symptoms.

B. *Basic examination:* observation of external structures, extra ocular movements and cranial nerves, dilating the pupil, visual acuity, slit-lamp examination, intraocular pressure (IOP), visual field testing, ophthalmoscopy.

Exophthalmometry

Normal values vary between 10 and 21 mm and are symmetrical in both eyes.

C. *Systemic examination*. A thorough examination should be conducted to rule out systemic causes of proptosis such as thyrotoxicosis, histiocytosis, and primary tumours elsewhere in the body (secondaries in orbits). Otorhinolaryngological examination is necessary when the paranasal sinus or a nasopharyngeal mass appears to be a possible etiological factor.

II. ***Laboratory investigations*** should include:

– thyroid function tests;
– haematological studies;
– Casoni's test (to rule out hydatid cyst);
– stool examination for cysts and ova;
– urine analysis for Bence Jones proteins for multiple myeloma.

III. *Imaging Technique*

A. Non-invasive techniques:

– *plain X-rays;*
– *computed tomography scanning;*
– *ultrasonography;*
– *magnetic resonance imaging (MRI).*

B. Invasive procedures:

– *orbital venography;*
– *carotid angiography;*
– *radioisotope studies*.

Enophthalmos

It is the inward displacement of the eyeball. About 50 percent cases of mild enophthalmos are misdiagnosed as having ipsilateral ptosis or contralateral proptosis.

Common causes are:

1. *Congenital:*

– microphthalmos and maxillarynhypoplasia.

2. *Traumatic*:

– blow out fractures of floor of the orbit.

3. *Post-inflammatory:*

– cicatrization of extraocular muscles as in the pseudotumour syndromes.

4. *Paralytic enophthalmos:*

– it is seen in Horner's syndrome (due to paralysis of cervical sympathetic).

5. *Atrophy of orbital contents:*

– senile atrophy of orbital fat, atrophy due to irradiation of malignant tumour, following cicatrizing metastatic carcinoma and due to scleroderma.

Orbital Inflammations

Classification

A. *Acute orbital and related inflammations*

1. Pre-septal cellulitis.
2. Orbital cellulitis and intraorbital abscess.

3. Orbital osteoperiostitis.
4. Orbital thrombophlebitis.
5. Tenonitis.
6. Cavernous sinus thrombosis.

B. *Chronic orbital inflammations*

I. *Specific inflammations*

1. Tuberculosis.
2. Syphilis.
3. Actinomycosis.
4. Mycotic infections (e. g., mucormycosis).
5. Parasitic infestations.

II. *Chronic non-specific inflammations*

1. Idiopathic orbital inflammatory disease (inflammatory pseudotumours).
2. Tolosa–Hunt syndrome.
3. Chronic orbital periostitis.

Salient features of some orbital inflammations of interest are described here.

Preseptal Cellulitis

Preseptal (or *periorbital*) *cellulitis* refers to infection of the subcutaneous tissues anterior to the orbital septum. Strictly speaking it is not an orbital disease but is included hereunder because the facial veins are valveless and preseptal cellulitis may spread posteriorly to produce orbital cellulitis.

Causes

Causative organisms are usually staphylococcus aureus or sreptococcus pyogenes.

Modes of infection

The organisms may invade the pre-septal tissue by any of the following modes:

1. *Exogenous infection* may result following skin laceration or insect bites.

2. *Extension from local infections* such as from an acute hordeolum or acute dacryocystitis.

3. *Endogenous infection* may occur by haematogenous spread from remote infection of the middle ear or upper respiratory tract.

Clinical features

Preseptal cellulitis presents as inflammatory oedema of the eyelids and periorbital skin with no involvement of the orbit. Thus, *characteristic features* are: painful acute periorbital swelling, erythema and hyperaemia of the lids. There may be associated fever and leukocytosis.

Treatment

It consists of *oral antibiotics* and *anti-inflammatory* drugs, with close follow-up care.

Orbital Cellulitis and Intraorbital Abscess

Orbital cellulitis refers to an acute infection of the soft tissues of the orbit behind the orbital septum. Orbital cellulitis may or may not progress to a sub-periosteal abscess or orbital abscess.

Etiology

Orbits may be infected by the following modes:

1. *Exogenous infection*. It may result from penetrating injury especially when associated with retention of intraorbital foreign body, and following operations like evisceration, enucleation, dacryocystectomy and orbitotomy.

2. *Extension of infection from neighbouring structures*.

3. *Endogenous infection*.

Causative organisms

Those commonly involved are: Streptococcus pneumonia, Staphylococcus aureus, Streptococcus pyogenic and Homophiles influenza.

Pathology

Pathological features of orbital cellulitis are similar to suppurative inflammations of the body in general.

Symptoms include: swelling and severe pain which is increased by movements of eye or pressure. Other associated symptoms may be fever, nausea, vomiting, prostrations and sometimes loss of vision.

Signs of orbital cellulitis are:

– a marked *swelling of lids* characterized by woody hardness and redness;

– a marked *chemosis of conjunctiva*, which may protrude and become desiccated or necrotic;

– the eyeball is *proptosed axially*;

– frequently, there is mild to severe *restriction of the ocular movements;*

– *fundus examination* may show congestion of retinal veins and signs of papillitis or papilledema.

Complications

These are quite common if not treated promptly:

1. *Ocular complications* are usually blinding and include exposure keratopathy, optic neuritis and central retinal artery occlusion.

2. *Orbital complications* are progression of orbital cellulitis into subperiosteal abscess and/or orbital abscess.

3. *Temporal or parotid abscesses* may occur due to spread of infection around the orbit.

4. *Intracranial complications* include cavernous sinus thrombosis, meningitis and brain abscesses.

5. *General septicemia or pyaemia* may occur eventually in few cases.

Treatment

1. *Intensive antibiotic therapy* to overcome the infection; intravenous antibiotics (oxacillin, chloramphenicol, cefotaxime, ciprofloxacin or vancomycin).

2. *Analgesic and anti-inflammatory drugs* are helpful in controlling pain and fever.

3. *Surgical intervention*. Its indications include unresponsiveness to antibiotics, decreasing vision and presence of an orbital or sub-periosteal abscess.

Orbital Periostitis

Orbital periostitis, i. e. inflammation of the periorbita is not very common. It may rarely involve the surrounding bones producing orbital osteoperiostitis.

Cavernous Sinus Thrombosis

Septic thrombosis of the cavernous sinus is a disastrous sequela, resulting from spread of sepsis travelling along its tributaries.

General features

Patient is seriously ill having high grade fever with rigors, vomiting and headache.

Ocular features

Patient develops:

– *severe pain* in the eye and forehead on the affected side;

– *conjunctiva* is swollen and congested;

– *proptosis* develops rapidly;

– *palsy* of the third, fourth and sixth cranial nerves occurs frequently;

– *oedema in mastoid region* is a pathognomonic sign;

– *fundus* may be normal with unimpaired vision in early cases. In advanced cases, retinal veins show congestion and there may appear papilloedema.

Complications

At any stage, the hyperpyrexia and signs of meningitis or pulmonary infarction may precede death.

Treatment

1. *Antibiotics* (oxacillin, chloramphenicol, cefotaxime, ciprofloxacin or vancomycin) are the sheet anchor of treatment. Massive doses of modern potent broad spectrum antibiotics should be injected intravenously.

2. *Analgesics* and *anti-inflammatory* drugs control pain and fever.

3. *Anticoagulants*' *role* is controversial.

Idiopathic Orbital Inflammatory Disease (Pseudotumours)

The term "pseudotumour" was coined for those conditions of the orbit which clinically presented as tumours but histopathologically proved to be chronic inflammations. Presently, *idiopathic orbital inflammatory disease (IOID)* is a term being preferred to denote this condition.

Graves' Ophthalmopathy

This term is coined to denote typical ocular changes which include lid retraction, lid lag, and proptosis. These changes have also been labelled as: endocrine exophthalmos, malignant exophthalmos, dysthyroid ophthalmopathy, ocular Graves' disease (OGD), and thyroid eye disease (TED).

TOPIC 5. DISEASES OF LACRIMAL SYSTEM

The nasolacrimal system, with its many intertwined and interrelated mechanisms, is a vital component of ocular homeostasis. This important system enables the spreading, distributing and elimination of the tear constituents across the ocular surfaces (palpebra, fornix and bulbar conjunctiva and cornea). The system supports the tear layer of the eye, which is essential to ocular surface health.

Lacrimal system disorders:

– tear film disorders;
– the dry eye;
– the wet eye;
– drainage obstruction;
– lacrimal inflammation;
– lacrimal gland tumours;
– lacrimal sac tumours.

The Watering Eye

It is characterized by overflow of tears from the conjunctival sac. The condition may occur either due to excessive secretion of tears (hyperlacrimation) or may result from obstruction to the outflow of normally secreted tears (epiphora).

Etiology:

– causes of hyperlacrimation;
– primary hyperlacrimation;
– reflex hyperlacrimation;
– causes of epiphora;
– physiological cause is ‘lacrimal pump’ failure due to lower lid laxity or weakness of orbicularis muscle;
– mechanical obstruction in lacrimal passages may lie at the level of punctum, canaliculus, lacrimal sac or nasolacrimal duct.

The Dry Eye

The dry eye per se is not a disease entity, but a symptom complex occurring as a consequence to deficiency or abnormalities of the tear film.

Etiology:

– systemic disease with lacrimal gland involvement;
– sarcoidosis;
– rheumatoid arthritis (Sjögren's syndrome);
– trachoma (chlamydial conjunctivitis and keratitis – *see next chapter*);
– neuroparalytic keratitis;
– exposure keratitis;
– old age;
– other rare causes.

Symptoms suggestive of dry eye include: irritation, foreign body (sandy) sensation, feeling of dryness, itching, non-specific ocular discomfort and chronically sore eyes not responding to a variety of drops instilled earlier.

Signs of dry eye include: presence of stringy mucus and particulate matter in the tear film, lusterless ocular surface, conjunctival xerosis, reduced or absent marginal tear strip and corneal changes in the form of punctate epithelial erosions and filaments.

Tear film tests

These include tear film break-up time (BUT), vital staining with Rose Bengal, Schirmer-I test, tear levels of lysozyme and lactoferrin, tear osmolality and conjunctival impression cytology. Out of these BUT, Rose Bengal staining and Schirmer-I tests are most important and when any two of these are positive, diagnosis of dry eye syndrome is confirmed.

1. *Tear film break-up (BUT):* it is the interval between a complete blink and appearance of the first randomly distributed dry spot on the cornea. It is noted after instilling a drop of fluorescein and examining in a cobalt-blue light of a slit-lamp.

BUT is an indicator of adequacy of mucin component of tears. Its normal values range from 15 to 35 seconds. Values less than 10 seconds imply an unstable tear film.

2. *Schirmer-I test*: it measures total tear secretions. It is performed with the help of a 5×35 mm strip of Whatman–41 filter paper which is folded 5 mm from one end and kept in the lower fornix at the junction of lateral one-third and medial two-thirds. The patient is asked to look up and not to blink or close the eyes. After 5 minutes wetting of the filter paper strip from the bent end is measured. Normal values of Schirmer-I test are more than 15 mm. Values of 5–10 mm are suggestive of moderate to mild keratoconjunctivitis sicca (KCS) and less than 5 mm of severe KCS.

3. *Rose Bengal staining:* it is a very useful test for detecting even mild cases of KCS. Depending upon the severity of KCS three staining patterns A, B and C have been described: "C" pattern represents mild or early cases with fine punctate stains in the interpalpebral area; "B" the moderate cases with extensive staining; and "A" the severe cases with confluent staining of conjunctiva and cornea.

Treatment

At present, there is no cure for dry eye. The following treatment modalities have been tried with variable results:

1. *Supplementation with tear substitutes:* artificial tears remain the mainstay in the treatment of dry eye.

2. *Topical cyclosporine* (0.05 %, 0.1 %) is reported to be very effective drug for dry eye in many recent studies. It helps by reducing the cell-mediated inflammation of the lacrimal tissue.

3. *Mucolytics*, such as 5 percent acetylcystine used 4 times a day, help by dispersing the mucus threads and decreasing tear viscosity.

4. *Topical retinoids* have recently been reported to be useful in reversing the cellular changes (squamous metaplasia) occurring in the conjunctiva of dry eye patients.

5. *Preservation of existing tears* by reducing evaporation and decreasing drainage.

Evaporation can be reduced by decreasing room temperature, use of moist chambers and protective glasses.

Punctal occlusion to decrease drainage can be carried out by collagen implants, cyanoacrylate tissue adhesives, electrocauterisation, argon laser occlusion and surgical occlusion to decrease the drainage of tears in patients with very severe dry eye.

Dacryocystitis

Inflammation of the lacrimal sac is not an uncommon condition. It may occur in two forms:

- congenital dacryocystitis;
- adult dacryocystitis.

Congenital dacryocystitis

It is an inflammation of the lacrimal sac occurring in newborn infants; and thus also known as *dacryocystitis neonatorum.*

Etiology

It follows stasis of secretions in the lacrimal sac due to congenital blockage in the nasolacrimal duct. It is of very common occurrence. Other causes of congenital nasolacrimal duct (NLD) block are: presence of epithelial debris, membranous occlusion at its upper end near lacrimal sac, complete non canalization and rarely bony occlusion. Common bacteria associated with congenital dacryocystitis are staphylococci, pneumococci and streptococci.

Clinical picture

Congenital dacryocystitis usually presents as a mild grade chronic inflammation. It is characterized by:

1. *Epiphora*, usually developing after seven days of birth. It is followed by copious *mucopurulent discharge* from the eyes.

2. *Regurgitation test* is usually positive, i. e., when pressure is applied over the lacrimal sac area, purulent discharge regurgitates from the lower punctum.

3. S*welling* on the sac area may appear eventually.

Differential diagnosis

Congenital dacryocystitis needs to be differentiated from other causes of watering in early childhood especially *ophthalmia neonatorum* and *congenital glaucoma*.

Complications

When not treated in time it may be complicated by recurrent conjunctivitis, acute on chronic dacryocystitis, lacrimal abscess and fistulae formation.

Treatment

It depends upon the age at which the child is brought. The treatment modalities employed are as follows:

1. *Massage over the lacrimal sac area and topical antibiotics* constitute the treatment of congenital NLD block, up to 6–8 weeks of age. This conservative treatment cures obstruction in about 90 percent of the infants.

2. *Lacrimal syringing (irrigation) with normal saline and antibiotic solution*. Syringing may be carried out once or twice a week.

3. *Probing of NLD with Bowman's probe*. It should be performed, in case the condition is not cured by the age of 3–4 months. It is usually performed under general anesthesia. While performing probing, care must be taken not to injure the canaliculus. In case of failure, it may be repeated after an interval of 3–4 weeks.

4. *Intubations with silicone tube* may be performed if repeated probings are failure.

5. *Dacryocystorhinostomy (DCR) operations:* when the child is brought very late or repeated probing is a failure, then conservative treatment by massaging, topical antibiotics and intermittent lacrimal syringing should be continued till the age of 4 years. After this, DCR operation should be performed.

Adult dacryocystitis

Adult dacryocystitis may occur in an *acute or a chronic form*.

Chronic dacryocystitis

Chronic dacryocystitis is more common than the acute dacryocystitis.

Etiology

The etiology of chronic dacryocystitis is multifactorial. The well-established fact is a vicious cycle of *stasis and mild infection* of long duration. The etiological factors can be grouped as follows:

A. *Predisposing factors*

1. *Age*: it is more common between 40 and 60 years of age.

2. *Sex:* the disease is predominantly seen in females (80 %) probably due to comparatively narrow lumen of the bony canal.

3. *Race:* it is rarer among Negroes than in Whites; as in the former NLD is shorter, wider and less sinuous.

4. *Heredity:* it plays an indirect role. It affects the facial configuration as well as the length and width of the bony canal.

5. *Socio-economic status:* it is more common in low socio-economic group.

6. *Poor personal hygiene*: it is also an important predisposing factor.

B. *Factors responsible for stasis of tears in lacrimal sac*

1. *Anatomical factors*, which retard drainage of tears include: comparatively narrow bony canal, partial canalization of membranous NLD and excessive membranous folds in NLD.

2. *Foreign bodies* in the sac may block opening of NLD.

3. *Excessive lacrimation*, primary or reflex, causes stagnation of tears in the sac.

4. *Mild grade inflammation* of lacrimal sac due to associated recurrent conjunctivitis may block the NLD by epithelial debris and mucus plugs.

5. *Obstruction of lower end of the NLD* by nasal diseases such as polyps, hypertrophied inferior concha, marked degree of deviated nasal septum, tumours and atrophic rhinitis causing stenosis may also cause stagnation of tears in the lacrimal sac.

C. *Source of infection*

Lacrimal sac may get infected from the conjunctiva, nasal cavity (retrograde spread), or paranasal sinuses.

D. *Causative organisms*

These include: staphylococci, pneumococci, streptococci and *Pseudomonas pyocyanea*. Rarely chronic granulomatous infections like tuberculosis, syphilis, leprosy and occasionally rhinosporiodosis may also cause dacryocystitis.

Clinical picture of chronic dacryocystitis may be divided into four stages:

1. *Stage of chronic catarrhal dacryocystitis:* it is characterized by mild inflammation of the lacrimal sac associated with blockage of NLD, watering eye and sometimes mild redness in the inner canthus.

2. *Stage of lacrimal mucocoele:* chronic stagnation causing distension of lacrimal sac. It is characterized by

constant epiphora associated with a swelling just below the inner canthus.

3. *Stage of chronic suppurative dacryocystitis:* due to pyogenic infections, the mucoid discharge becomes purulent, converting the mucocele into pyocele. The condition is characterized by epiphora, associated recurrent conjunctivitis and swelling at the inner canthus with mild erythema of the overlying skin.

4. *Stage of chronic fibrotic sac:* low grade repeated infections for a prolonged period ultimately result in a small fibrotic sac due to thickening of mucosa, which is often associated with persistent epiphora and discharge. Dacryocystography at this stage reveals a very small sac with irregular folds in the mucosa.

Complications:

– chronic intractable conjunctivitis, acute on chronic dacryocystitis;

– ectropion of lower lid, maceration and eczema of lower lid skin due to prolonged watering;

– simple corneal abrasions may become infected leading to hypopyon ulcer;

– if an intraocular surgery is performed in the presence of dacryocystitis, there is high risk of developing endophthalmitis. Because of this, syringing of lacrimal sac is always done before attempting any intraocular surgery.

Treatment

1. *Conservative treatment* by repeated lacrimal syringing.

2. *Dacryocystorhinostomy* (DCR): it should be the operation of choice as it re-establishes the lacrimal drainage.

3. *Dacryocystectomy* (DCT): it should be performed only when DCR is contraindicated. *Indications of DCT* include:

– too young (less than 4 years) or too old (more than 60 years) patient;

– markedly shrunken and fibrosed sac;

– tuberculosis, syphilis, leprosy or mycotic infections of sac;

– tumours of sac;

– gross nasal diseases like atrophic rhinitis;

– an unskilled surgeon, because it is said that "a good DCT" is always better than "a badly done DCR".

4. *Conjunctivodacryocystorhinostomy* (CDCR): it is performed in the presence of blocked canaliculi.

Acute dacryocystitis

Acute dacryocystitis is an acute suppurative inflammation of the lacrimal sac, characterized by presence of a painful swelling in the region of sac.

Etiology

It may develop in two ways:

1. *As an acute exacerbation of chronic dacryocystitits.*

2. *As an acute peridacryocystitis* due to direct involvement from the neighbouring infected structures such as: paranasal sinuses, surrounding bones and dental abscess or caries teeth in the upper jaw.

Causative organisms

Commonly involved are *Streptococcus haemolyticus*, *Pneumococcus* and *Staphylococcus*.

Clinical picture of acute dacryocystitis can be divided into 3 stages:

1. *Stage of cellulitis:* it is characterized by a *painful swelling* in the region of lacrimal sac associated with *epiphora* and constitutional symptoms such as *fever* and *malaise.* The swelling is red, hot, firm and tender. Redness and edema also spread to the lids and cheek.

2. *Stage of lacrimal abscess:* continued inflammation causes occlusion of the canaliculi due to edema. The sac is filled with pus, distends and its anterior wall ruptures forming a pericystic swelling. In this way, a large fluctuant swelling the lacrimal abscess is formed.

3. *Stage of fistula formation:* when the lacrimal abscess is left unattended, it discharges spontaneously, leaving an *external fistula* below the medial palpebral ligament.

Complications:

– acute conjunctivitis;

– corneal abrasion which may be converted to corneal ulceration;

– lid abscess;

– osteomyelitis of lacrimal bone;

– orbital cellulitis;

– facial cellulitis and acute ethmoiditis;

– rarely cavernous sinus thrombosis and very rarely generalized septicaemia may also develop.

Treatment

1. *During cellulitis stage:* it consists of systemic and topical antibiotics to control infection; and systemic anti-inflammatory analgesic drugs and hot fomentation to relieve pain and swelling.

2. *During stage of lacrimal abscess:* in addition to the abovementioned treatment when pus starts pointing on the skin, it should be drained with a small incision.

The pus should be gently squeezed out, the dressing done with *betadine* soaked roll gauze. Later on depending upon condition of the lacrimal sac either DCT or DCR operation should be carried out, otherwise recurrence will occur.

3. *Treatment of external lacrimal fistula.* After controlling the acute infection with systemic antibiotics, fistulectomy along with DCT or DCR operation should be performed.

Dacryocystorhinostomy (DCR) operation can be performed by two techniques: conventional external approach DCR, and endonasal DCR.

Dacryoadenitis

Dacryoadenitis is inflammation of the lacrimal glands; may be *acute or chronic.*

Acute dacryoadenitis

Etiology

It may develop as a primary inflammation of the gland or secondary to some local or systemic infection. Dacryoadenitis secondary to local infections occurs in trauma, erysipelas of the face, conjunctivitis (especially gonococcal and staphylococcal) and orbital cellulitis. Dacryoadenitis secondary to systemic infections is associated with mumps, influenza, infectious mononucleosis and measles.

Clinical picture

Acute inflammation of the palpebral part is characterized by a painful swelling in the lateral part of the upper lid which becomes red and swollen with a typical S-shaped curve of its margin. Acute orbital dacryoadenitis produces some painful proptosis in which the eyeball moves down and in. A fistula in the upper and lateral quadrant of the upper lid may develop as a complication of suppurative dacryoadenitis.

Treatment

It consists of a course of appropriate systemic antibiotic, analgesic and anti-inflammatory drugs along with hot fomentation. When pus is formed, incision and drainage should be carried out.

Chronic dacryoadenitis

It is characterized by engorgement and simple hypertrophy of the gland.

Etiology

Chronic dacryoadenitis may occur:

– as sequelae to acute inflammation;

– in association with chronic inflammations of conjunctiva;

– due to systemic diseases such as tuberculosis, syphilis and sarcoidosis.

Clinical features:

– a painless swelling in upper and outer part of lid associated with ptosis;

– eyeball may be displaced down and in;

– diplopia may occur in up and out gaze.

On palpation, a firm lobulated mobile mass may be felt under the upper and outer rim of the orbit.

Differential diagnosis: from other causes of lacrimal gland swellings is best made after fine needle aspiration biopsy or incisional biopsy.

Treatment consists of treating the cause.

Tumours of the Lacrimal Gland

These are not so common and in a simplified way can be classified as follows:

1. *Lymphoid tumours and inflammatory pseudotumours*.
2. *Benign epithelial tumours.*
3. *Malignant epithelial tumours*.

TOPIC 6. STRABISMUS AND NYSTAGMUS

Binocular Single Vision

Definition

When a normal individual fixes his/her visual attention on an object of regard, the image is formed on the fovea of both eyes separately; but the individual perceives a single image. This state is called *binocular single vision.*

Anomalies of binocular vision

Anomalies of binocular vision include *suppression*, *amblyopia*, *abnormal retinal correspondence (ARC)*, *confusion* and *diplopia*.

Diplopia

Binocular diplopia occurs due to formation of image on dissimilar points of the both retinae.

Causes of binocular diplopia are:

– *paralysis or paresis* of the extraocular muscles (the most common cause);

– *displacement of one eye ball*;

– *mechanical restriction of ocular movements*;

– *deviation of ray of light in one eye* as caused by decentred spectacles;

– *anisometropia*, i. e. disparity of image size between two eyes.

Types

Binocular diplopia may be *crossed* or *uncrossed*.

In *uncrossed* (harmonious) diplopia the false image is on the same side as deviation. It occurs in convergent squint.

In *crossed* (unharmonious) diplopia the false image is seen on the opposite side. It occurs in divergent squint.

*Uniocular diplopia**:*** an object appears double from the affected eye even when the normal eye is closed.

Causes of uniocular diplopia are:

– *subluxated clear lens*;

– *subluxated intraocular lens*;
– *double pupil*;
– *incipient cataract*;
– *keratoconus.*

Treatment of diplopia

Treat the causative disease. Temporary relief from annoying diplopia can be obtained by occluding the affected eye.

Strabismus

Normally visual axis of the two eyes is parallel to each other in the "primary position of gaze" and this alignment is maintained in all positions of gaze.

A misalignment of the visual axes of the two eyes is called *squint* or *strabismus.*

Classification of strabismus:

I. Apparent squint (pseudostrabismus).

II. Latent squint (heterophoria).

III. Manifest squint (heterotropia):

1. Concomitant squint.
2. Incomitant squint.

Pseudostrabismus

In pseudostrabismus (apparent squint), the visual axes are in fact parallel, but the eyes seem to have a squint:

1. *Pseudoesotropia* or *apparent convergent squint* may be associated with a prominent epicanthal fold (which covers the normally visible nasal aspect of the globe and gives a false impression of esotropia) and negative angle kappa.

2. *Pseudoexotropia* or *apparent divergent squint* may be associated with *hypertelorism,* a condition of wide separation of the two eyes, and positive angle kappa.

Heterophoria

Heterophoria, also known as "*latent strabismus*", is a condition in which the tendency of the eyes to deviate is kept latent by fusion. Therefore, when the influence of fusion is removed the visual axis of one eye deviates away.

O*rthophoria* is a condition of perfect alignment of the two eyes which is maintained even after the removal of influence of fusion. However, orthophoria is a theoretical ideal. Practically a small amount of heterophoria is of universal occurrence and is known as "*physiological heterophoria*".

Types of heterophoria

1. *Esophoria:* it is a tendency to converge. It may be:

– *convergence excess type* (esophoria greater for near than distance);

– *divergence weakness type* (esophoria greater for distance than near);

– *non-specific type* (esophoria which does not vary significantly in degree for any distance).

2. *Exophoria:* it is a tendency to diverge. It may be:

– *convergence weakness type* (exophoria greater for near than distance);

– *divergence excess type* (exophoria greater on distant fixation than the near);

– *non-specific type* (exophoria which does not vary significantly in degree for any distance).

3. *Hyperphoria:* it is a tendency to deviate upwards, while hypophoria is a tendency to deviate downwards. However, in practice it is customary to use the term right or left hyperphoria depending on the eye which remains up as compared to the other.

4. *Cyclophoria:* it is a tendency to rotate around the anteroposterior axis. When the 12 o'clock meridian of cornea rotates nasally, it is called *incyclophoria* and when it rotates temporally it is called *excyclophoria.*

Etiology

A. *Anatomical factors*

1. Orbital asymmetry.

2. Abnormal interpupillary distance (IPD). A wide IPD is associated with exophoria and small with esophoria.

3. Faulty insertion of extraocular muscle.

4. A mild degree of extra ocular muscle weakness.

5. Anomalous central distribution of the tonic innervation of the two eyes.

6. Anatomical variation in the position of the macula in relation to the optical axis of the eye.

B. *Physiological factors*

1. *Age.* Esophoria is more common in younger age group as compared to exophoria which is more often seen in the elderly.

2. *Role of accommodation.* Increased accommodation is associated with esophoria (as seen in hypermetropes and individuals doing excessive near work) and decreased accommodation with exophoria (as seen in simple myopes).

3. *Role of convergence.* Excessive use of convergence may cause esophoria (as occurs in bilateral congenital myopes) while decreased use of convergence is often associated with exophoria (as seen in presbyopes).

4. *Dissociation factor* such as prolonged constant use of one eye may result in exophoria (as occurs in individuals using uniocular microscope and watch makers using uniocular magnifying glass).

Factors predisposing to decompensation

– inadequacy of fusional reserve;

– general debility and lowered vitality;

– psychosis, neurosis, mental stress, precision of job and advancing age.

Symptoms

Depending upon the symptoms heterophoria can be divided into *compensated* and *decompensated*.

Compensated heterophoria: it is associated with no subjective symptoms. Compensation of heterophoria depends upon the reserve neuro-muscular power to overcome the muscular imbalance and individual's desire for maintenance of binocular vision.

Decompensated heterophoria: it is associated with multiple symptoms which may be grouped as follows:

1. *Symptoms of muscular fatigue* resulting due to continuous use of the reserve neuromuscular power. These include:

– *headache* and *ophthalmalgia* after prolonged use of eyes, which is relieved when the eyes are closed;

– *difficulty in changing the focus* from near to distant objects of fixation or vice-versa;

– *photophobia* caused by muscular fatigue is not relieved by using dark glasses, but relieved by closing one eye.

2. *Symptoms of failure to maintain binocular single vision* are:

– *blurring* or crowding of words while reading;

– *intermittent diplopia* due to temporary manifest deviation under conditions of fatigue;

– *intermittent squint* (without diplopia) which is usually noticed by the patient's close relations or friends.

3. *Symptoms of defective postural sensations* cause problems in judging distances and positions especially of the moving objects. This difficulty may be experienced by cricketers, tennis players and pilots during landing.

Treatment is indicated in decompensated heterophoria:

1. Correction of refractive error (when detected) is most important.

2. Orthoptic treatment.

3. Prescription of prism in glasses.
4. Surgical treatment.

Concomitant strabismus

It is a type of manifest squint in which the amount of deviation in the squinting eye remains constant (unaltered) in all the directions of gaze; and there is no associated limitation of ocular movements.

Etiology

It is not clearly defined. As we know, the binocular vision and coordination of ocular movements are not present since birth but are acquired in the early childhood. Any obstacle to the development of these processes may result in concomitant squint. These obstacles can be arranged into three groups, namely: *sensory*, *motor* and *central*.

1. *Sensory obstacles.* These are the factors which hinder the formation of a clear image in one eye. They include:

– refractive errors;
– prolonged use of incorrect spectacles;
– anisometropia;
– corneal opacities;
– lenticular opacities;
– diseases of macula (e. g., central chorioretinitis);
– optic atrophy;
– obstruction in the pupillary area due to congenital ptosis.

2. *Motor obstacles.* These factors hinder the maintenance of two eyes in the correct positional relationship in primary gaze and/or during different ocular movements. Some of such factors are:

– congenital abnormalities of the shape and size of the orbit;
– abnormalities of extra ocular muscles such as faulty insertion, faulty innervation and mild paresis;

– abnormalities of accommodation, convergence and AC/A ratio.

3. *Central obstacles.* These may be in the form of:

– deficient development of fusion faculty;

– abnormalities of cortical control of ocular movements as occurs in mental trauma, and hyper excitability of the central nervous system during teething.

Clinical features

The cardinal features of different clinic-etiological types of concomitant strabismus are described separately. However, the clinical features of concomitant strabismus (in general) are as follows:

1. *Ocular deviation.* Characteristics of ocular deviation are:

– unilateral (monocular squint) or alternating (alternate squint);

– inward deviation (esotropia), outward deviation (exotropia) or vertical deviation (hypertropia);

– primary deviation (of squinting eye) is equal to secondary deviation (deviation of normal eye under cover when patient fixes with squinting eye);

– ocular deviation is equal in all the directions of gaze.

2. *Ocular movements* are not limited in any direction.

3. *Refractive error* may or may not be associated.

4. *Suppression* and *amblyopia* may develop as sensory adaptation to strabismus. Suppression may be monocular (in monocular squint) and alternating (in alternating strabismus). Amblyopia develops in monocular strabismus only and is responsible for poor visual acuity.

5. *A–V patterns* may be observed in horizontal strabismus. When A–V patterns are associated, the horizontal concomitant strabismus becomes vertical.

Types of concomitant squint:

1. Convergent squint (esotropia).

2. Divergent squint (exotropia).
3. Vertical squint (hypertropia).

Convergent squint

Concomitant convergent squint or esotropia denotes inward deviation of one eye. It can be *unilateral* (the same eye always deviates inwards and the second normal eye takes fixation) or *alternating* (either of the eyes deviates inwards and the other eye takes up fixation, alternately).

Treatment of Concomitant Strabismus

Goals of treatment

These are to achieve good cosmetic correction, to improve visual acuity and to maintain binocular single vision. However, sometimes it is not possible to achieve all the goals in every case.

Treatment modalities:

1. *Spectacles with full refractive correction.*
2. *Occlusion therapy.* It is indicated in the presence of amblyopia. Occlusion helps to improve the vision in children below the age of 10 years.
3. *Preoperative orthoptic exercises.* These are given after the correction of amblyopia to overcome suppression.
4. *Squint surgery.* It is required in most of the cases to correct the deviation.
5. *Postoperative orthoptic exercises.*

Paralytic Strabismus

It refers to ocular deviation resulting from complete or incomplete paralysis of one or more extraocular muscles.

Etiology

The lesions may be neurogenic, myogenic or at the level of neuromuscular junction.

I. *Neurogenic lesions*

1. *Congenital.* Hypoplasia or absence of nucleus is a known cause of the third and sixth cranial nerve palsies. Birth injuries may mimic congenital lesions.

2. *Inflammatory lesions.* These may be in the form of encephalitis, meningitis, neurosyphilis or peripheral neuritis (commonly viral). Nerve trunks may also be involved in the infectious lesions of cavernous sinus and orbit.

3. *Neoplastic lesions.* These include brain tumours.

4. *Vascular lesions.* These are known in patients with hypertension, diabetes mellitus and atherosclerosis.

5. *Traumatic lesions.* These include head injury and direct or indirect trauma to the nerve trunks.

6. *Toxic lesions.* These include carbon monoxide poisoning, effects of diphtheria toxins (rarely), alcoholic and lead neuropathy.

7. *Demyelinating lesions.* Ocular palsy may occur in multiple sclerosis and diffuse sclerosis.

II. *Myogenic lesions*

1. *Congenital lesions.* These include absence, hypoplasia, malinsertion, weakness and musculo-facial anomalies.

2. *Traumatic lesions.* These may be in the form of laceration, disinsertion, hemorrhage into the muscle substance or sheath and incarceration of muscles in fractures of the orbital walls.

3. *Inflammatory lesions.* Myositis is usually viral in origin and may occur in influenza, measles and other viral fevers.

4. *Myopathies.* These include thyroid myopathy, carcinomatous myopathy and that associated with certain drugs.

5. *Progressive external ophthalmoplegia* is a bilateral myopathy of extra ocular muscles; which may be sporadic or inherited as an autosomal dominant disorder.

III. *Neuromuscular junction lesion.* It includes *myasthenia gravis*.

Symptoms

1. *Diplopia.*

2. *Confusion.*

3. *Nausea* and *vertigo.* These result from diplopia and confusion.

4. *Ocular deviation.* It is of sudden onset.

Treatment

Therapy for incomitant paralytic strabismus is aimed at aligning the eyes in positions of gaze in which a deviation exists without disturbing single binocular vision elsewhere in the field of fixation.

While surgery is necessary to achieve this goal in most instances, conservative methods should be considered in suitable cases.

When conservative therapy fails or the deviation is of such magnitude that it may not even be considered, surgery becomes necessary. The timing of an operation depends on the nature of the underlying paralysis. If the paralysis is longstanding, surgery may be performed as soon as the diagnosis is established. If the paralysis is of recent onset, a 6 to 8-month waiting period is mandatory for the condition to be considered stable; spontaneous recovery of function rarely occurs after that length of time. During the waiting period the patient should be examined at frequent intervals and visual comfort maintained with prisms or unilateral occlusion.

Nystagmus

It is defined as regular and rhythmic to-and-fro involuntary oscillatory movements of the eyes.

Etiology

It occurs due to disturbance of the factors responsible for maintaining normal ocular posture. These include disorders of sensory visual pathway, vestibular apparatus, semicircular canals, mid-brain and cerebellum.

Features of nystagmus

Nystagmus may be characterized by any of the following ***features***:

1. It may be *pendular* or *jerk nystagmus.* In pendular nystagmus the movements are of equal velocity in each direction. It may be horizontal, vertical or rotatory. In jerk nystagmus, the movements have a slow component in one direction and a fast component in the other direction. The direction of jerk nystagmus is defined by direction of the fast component (phase). It may be right, left, up, down or rotatory.

2. Nystagmus movements may be *rapid* or *slow*, *fine* or *coarse*, *latent* or *manifest*.

Different kinds of nystagmus include:

– congenital nystagmus;
– manifest nystagmus;
– latent nystagmus;
– manifest-latent nystagmus;
– acquired nystagmus.

Congenital nystagmus is present at birth. With this condition, your eyes move together as they oscillate (swing like a pendulum).

As mentioned above, most people with nystagmus are born with the condition or develop it early in life. Unless induced by trauma or disease, nystagmus almost always is caused by neurological problems.

The two basic types of nystagmus are:

– optokinetic (eye related);
– vestibular (inner ear related).

People with inner ear problems can develop something called “jerk nystagmus”, when the eyes drift slowly in one direction and then jerk back in the other direction. Because of the motion of the eyes, people with this condition can develop nausea and vertigo. This type of nystagmus, usually temporary, also can occur in people with Meniere’s disease (inner ear disorder) or when water settles into one ear. Taking a decongestant sometimes can clear up this type of nystagmus.

Children with nystagmus may need extra help in learning to adapt to schoolwork and social situations.

Manifest nystagmus is present at all times, whereas *latent nystagmus* occurs when one eye is covered.

Manifest-latent nystagmus is continually present, but worsens when one eye is covered.

Acquired nystagmus can be caused by a disease (multiple sclerosis, brain tumour, diabetic neuropathy), an accident (head injury), or a neurological problem (side effect of a medication). Hyperventilation, a flashing light in front of one eye, nicotine and even vibrations have been known to cause nystagmus in rare cases.

Some acquired nystagmuses can be treated with medications or surgeries.

All forms of nystagmus are involuntary, which means people with the condition cannot control their eyes. Nystagmus improves slightly as a person reaches adulthood; however, it worsens with tiredness and stress.

Having nystagmus affects both vision and self-concept. Most people with nystagmus have some sort of vision limitations because the eyes continually sweep over what they are viewing, making it impossible to obtain a clear image.

Some people with nystagmus have so many vision problems that they can be considered legally blind.

TOPIC 7. DISEASES OF THE CONJUNCTIVA

Inflammation of the conjunctiva (conjunctivitis) is defined as conjunctival hyperemia associated with a discharge which may be watery, mucoid, mucopurulent or purulent.

Etiological classification:

– infective conjunctivitis: bacterial, chlamydial, viral, fungal, rickettsia, spirochetal, protozoal, parasitic, etc;
– allergic conjunctivitis;
– irritative conjunctivitis;
– keratoconjunctivitis associated with diseases of skin and mucous membrane;
– traumatic conjunctivitis;
– keratoconjunctivitis of unknown etiology.

Clinical classification

Depending upon clinical manifestation, conjunctivitis can be classified as follows:

– acute catarrhal or mucopurulent conjunctivitis;
– acute purulent conjunctivitis;
– serous conjunctivitis;
– chronic simple conjunctivitis;
– angular conjunctivitis;
– membranous conjunctivitis;
– pseudomembranous conjunctivitis;
– papillary conjunctivitis;
– follicular conjunctivitis;
– ophthalmia neonatorum;
– granulomatous conjunctivitis;
– ulcerative conjunctivitis;
– cicatrizing conjunctivitis.

To describe different types of conjunctivitis, a mixed approach has been adopted, i. e. some varieties of conjunctivitis are described by their etiological names and

others – by their clinical names. Only common varieties of clinical interest are described here.

Bacterial Conjunctivitis

It is a common type of pink eye, caused by bacteria that infect the eye through various sources of contamination. The bacteria spread through contact with an infected individual, exposure to contaminated surfaces or through other means such as sinus or ear infections.

Etiology

Predisposing factors for bacterial conjunctivitis, especially epidemic forms, are flies, poor hygienic conditions, hot dry climate, poor sanitation and dirty habits. These factors help the infection to establish, as the disease is highly contagious.

Causative organisms: it may be caused by a Staphylococcus aureus, Staphylococcus epidermidis, Streptococcus pneumoniae (pneumococcus), Streptococcus pyogenes (Hemolytic), Hemophilus influenzae (Koch–Weeks bacillus), Moraxella lacunae (Morax–Axenfeld bacillus), Pseudomonas pyocyanea, Neisseria gonorrhoeae, Neisseria meningitidis (meningococcus), Corynebacterium diphtheriae.

Symptoms:

- discomfort and foreign body sensation;
- mild photophobia;
- mucopurulent discharge;
- sticking together of lid margins;
- slight blurring of vision;
- coloured halos;
- conjunctival congestion;
- chemosis;
- petechial;
- flakes of mucopus;
- cilia are usually matted together with yellow crusts.

Treatment

No bandage should be applied in patients with mucopurulent conjunctivitis!!!

Topical antibiotics: treatment may be started with 0.3 %, ciprofloxacin, 0.3 %, ofloxacin, 1 % chloramphenicol, 0.3 % gentamycin or framycetin eye drops every 3–4 hours during a day and ointment used at night.

Anti-inflammatory and *analgesic drugs* (ibuprofen, voltaren, nimesil and paracetamol) may be given orally for 2–3 days to provide symptomatic relief from mild pain, especially in sensitive patients.

No steroids should be applied, otherwise infection will flare up and bacterial corneal ulcer may develop.

Acute Purulent Conjunctivitis

Acute purulent conjunctivitis, also known as *acute blenorrhea* or *hyper acute conjunctivitis*, is characterized by a violent inflammatory response. It occurs in two forms:

1. Adult purulent conjunctivitis.
2. Ophthalmia neonatorum in newborns.

Acute purulent conjunctivitis of adults

Etiology

The disease affects adults, predominantly males. Commonest causative organism is *Gonococcus*; but rarely, it may be *Staphylococcus aureus* or *Pneumococcus.* Gonococcal infection spreads directly from genitals to eyes. Presently, incidence of gonococcal conjunctivitis has markedly decreased.

Clinical picture

It can be divided into three stages:

1. *Stage of infiltration.* It lasts for 4–5 days and is characterized by:

– considerably painful and tender eyeball;
– bright red velvety chemosed conjunctiva;
– lids are tense and swollen;

– discharge is watery or sanguineous;

– pre-auricular lymph nodes are enlarged.

2. *Stage of blennorrhea.* It starts at about fifth day, lasts for several days and is characterized by:

– frankly purulent, copious, thick discharge trickling down the cheeks;

– other symptoms are increased but tension in the lids is decreased.

3. *Stage of slow healing.* During this stage, pain is decreased and swelling of the lids subsides. Conjunctiva remains red, thickened and velvety. Discharge diminishes slowly and in the end resolution is complete.

Associations

Gonococcal conjunctivitis is usually associated with *urethritis* and *arthritis*.

Complications

1. *Corneal involvement* is quite frequent, such as the diffuse haze and corneal edema, central necrosis, ulceration or even perforation.

2. *Iridocyclitis* may also occur, but is not as common as corneal involvement.

3. *Systemic complications*, though rare, include gonorrhea arthritis, endocarditis and septicemia.

Treatment

1. *Systemic therapy* is far more critical than the topical therapy for the infections caused by *N. gonorrhoeae* and *N. meningitidis*. Because of the resistant strains penicillin and tetracycline are no longer adequate as first-line treatment. Any of the following regimes can be adopted:

– *oral antibiotics:* norfloxacin 1.2 g, cefoxitin 1.0 g or cefotaxime 500 mg orally for 5 days;

– *IM injections:* ceftriaxone 1.0 g, spectinomycin 2.0 g for 3–5 days.

All of the above regimes should then be followed by a one week course of either doxycycline 100 mg bid or erythromycin 250–500 mg orally qid (four times a day).

2. *Topical antibiotic therapy* presently recommended includes ofloxacin, ciprofloxacin or tobramycin eye drops or bacitracin or erythromycin eye ointment every 2 hours for the first 2–3 days and then 5 times daily for 7 days.

3. *Irrigation* of the eyes frequently with sterile saline is very therapeutic in washing away infected debris.

4. *Other general measures* are similar to acute mucopurulent conjunctivitis.

5. *Topical atropine* 1 % eye drops should be instilled once or twice a day if cornea is involved.

6. *Patient and the sexual partner* should be referred for evaluation of other sexually transmitted diseases.

Acute Membranous Conjunctivitis

It is an acute inflammation of the conjunctiva, characterized by formation of a true membrane on the conjunctiva. Nowadays, it is rare because of markedly decreased incidence of diphtheria. It is because of the fact that immunization against diphtheria is very effective.

Etiology

The disease is typically caused by *Corynebacterium diphtheriae* and occasionally by virulent type of *Streptococcus haemolyticus*.

Clinical features

Children between 2–8 years of age who are not immunized against diphtheria are usually affected. The disease may have a mild or very severe course. The child is intoxicated and febrile. The clinical picture of the disease can be divided into three stages:

1. *Stage of infiltration* is characterized by pain in the eye, conjunctival discharge, lids are swollen and hard,

conjunctiva is red, swollen and covered with a thick grey-yellow membrane. The membrane is tough and firmly adherent to the conjunctiva, which on removing bleeds and leaves behind a raw area. Pre-auricular lymph nodes are enlarged.

2. *Stage of suppuration*. There is copious outpouring of purulent discharge.

3. *Stage of cicatrisation.* In this stage, the raw surface covered with granulation tissue is epithelized. Healing occurs by cicatrisation, which may cause trichiasis and conjunctival xerosis.

Complications

1. *Corneal ulceration* is a frequent complication in acute stage.

2. *Delayed complications* due to cicatrization include symblepharon, trichiasis, entropion and conjunctival xerosis.

Diagnosis

It is made from typical clinical features and is confirmed by bacteriological examination.

Treatment

A. *Topical therapy*

1. *Penicillin eye drops* (1:10 000 units per ml) should be instilled half hourly.

2. *Antidiphtheric serum* (ADS) should be instilled every hour.

3. *Atropine sulfate* 1 % ointment should be added if cornea is ulcerated.

4. *Broad spectrum antibiotic* ointment should be applied at bed time.

B. *Systemic therapy*

1. *Crystalline penicillin* 500 thousand units should be injected intramuscularly twice a day for 10 days.

2. *Antidiphtheric serum* (ADS) 50 thousand units should be given intramuscularly stat (immediately).

C. *Prevention of symblepharon*

Once the membrane is sloughed off, the healing of raw surfaces will result in symblepharon, which should be prevented by applying contact shell or sweeping the fornices with a glass rod smeared with ointment.

Prophylaxis

Isolation of patient will prevent family members from being infected.

Proper immunization against diphtheria is very effective and provides protection to the community.

Pseudomembranous Conjunctivitis

It is a type of acute conjunctivitis, characterized by formation of a pseudo membrane (which can be easily peeled off leaving behind intact conjunctiva epithelium) on the conjunctiva.

Etiology: *bacterial infection, viral infections and chemical irritants.*

Clinical picture

Pseudomembranous conjunctivitis is characterized by:

– *acute mucopurulent conjunctivitis*;

– *pseudo membrane* formation which is thin yellowish-white membrane seen in the fornices and on the palpebral conjunctiva. Pseudo membrane can be peeled off easily and does not bleed.

Treatment

It is similar to that of mucopurulent conjunctivitis.

Chronic Catarrhal Conjunctivitis

It is characterized by mild catarrhal inflammation of the conjunctiva.

Etiology

A. *Predisposing factors*

1. *Chronic exposure* to dust, smoke, and chemical irritants.

2. *Local cause of irritation* such as trichiasis, concretions, foreign body and seborrheic scales.

3. *Eye strain* due to refractive errors, phorias or convergence insufficiency.

4. *Abuse of alcohol*, *insomnia* and *metabolic disorders*.

B. *Causative organisms**:*** *Staphylococcus aureus*, also *gram-negative rods* such as *Proteus mirabilis*, *Klebsiella pneumoniae*, *Escherichia coli* and *Moraxella lacunata*.

Clinical picture

Symptoms of simple chronic conjunctivitis include:

– burning and grittiness in the eyes, especially in the evening;

– mild chronic redness in the eyes;

– feeling of heat and dryness on the lid margins;

– difficulty in keeping the eyes open;

– mild mucous discharge especially in the canthi;

– off and on lacrimation;

– feeling of sleepiness and tiredness in the eyes;

– mild papillary hypertrophy of the palpebral conjunctiva.

Treatment

Predisposing factors when associated should be treated and eliminated.

Oral antibiotics: norfloxacin 1.2 g, cefoxitin 1.0 g or cefotaxime 500 mg orally for 5 days.

IM injections: ceftriaxone 1.0 g, spectinomycin 2.0 g for 3–5 days.

All of the above regimes should then be followed by a one week course of either doxycycline 100 mg bid or erythromycin 250–500 mg orally qid.

Topical antibiotic therapy presently recommends ofloxacin, ciprofloxacin or tobramycin eye drops; bacitracin or

erythromycin eye ointment every 2 hours for the first 2–3 days and then 5 times daily for 7 days.

Angular Conjunctivitis

It is a type of chronic conjunctivitis characterized by mild degree of inflammation confined to the conjunctiva and lid margins near the angles (hence the name) associated with maceration of the surrounding skin.

Etiology

1. *Predisposing factors* are same as for the simple chronic conjunctivitis.

2. *Causative organisms.* Morax–Axenfeld bacillus is the commonest causative organism. Rarely, *staphylococci* may also cause angular conjunctivitis.

3. *Source of infection* is usually nasal cavity.

4. *Mode of infection.* Infection is transmitted from nasal cavity to the eyes by contaminated fingers or handkerchief.

Clinical picture

– irritation, smarting sensation and feeling of discomfort in the eyes;

– history of collection of dirty-white foamy discharge at the angles;

– redness in the angles of eyes;

– hyperemia of bulbar conjunctiva near the canthi;

– hyperemia of lid margins near the angles;

– excoriation of the skin around the angles;

– presence of foamy mucopurulent discharge at the angles.

Complications include: blepharitis and shallow marginal catarrhal corneal ulceration.

Treatment

Prophylaxis includes treatment of associated nasal infection and good personal hygiene.

Curative treatment consists of:

1. *Antibiotics* – 1 % oxytetracycline, 0.3 % tobramycin, 0.3 % gentamycin eye ointment 2–3 times a day for 9–14 days will eradicate the infection.

2. *Zinc lotion* instilled in day time and *zinc oxide ointment* at bed time inhibits the proteolytic ferment and thus helps in reducing the maceration.

Chlamydial Conjunctivitis

Chlamydiae lie between bacteria and viruses, sharing some of the properties of both. Like viruses, they are obligate, intracellular and filterable, whereas like bacteria they contain both DNA and RNA, divided by binary fission and are sensitive to antibiotics.

The chlamydiae form the PLT group (psittacosis, lymphogranuloma venereum and trachomatis group).

Classification

- class 1 blinding trachoma;
- class 2 non-blinding trachoma;
- class 3 paratrachoma.

Trachoma

The word "trachoma" comes from the Greek word for "rough" which describes the surface appearance of the conjunctiva in chronic trachoma.

Trachoma (previously known as *Egyptian ophthalmia*) is a chronic keratoconjunctivitis, primarily affecting the superficial epithelium of conjunctiva and cornea simultaneously.

Etiology

It is caused by *Chlamydia trachomatis serotypes A, B* and *D*. Chlamydia is similar to viruses as it can diffuse through membranes and forms inclusion bodies made of carbohydrates and proteins.

Predisposing factors

These include age, sex, race, climate, socioeconomic status and environmental factors.

1. *Age.* The infection is usually contracted during infancy and early childhood.

2. *Sex.* As far as sex is concerned, there is general agreement that preponderance exists in the females both in number and in severity of disease.

3. *Race.* No race is immune to trachoma, but the disease is very common in Jews and comparatively less common among Africans.

4. *Climate.* Trachoma is more common in areas with dry and dusty weather.

5. *Socioeconomic status.* The disease is more common in poor classes owing to unhygienic living conditions, overcrowding, unsanitary conditions, abundant fly population, paucity of water, lack of materials like separate towels and handkerchiefs, and lack of education and understanding about spread of contagious diseases.

6. *Environmental factors*, e. g., exposure to dust/smoke, irritants, sunlight, increase the risk of contracting disease. Therefore, outdoor workers are more affected in comparison to office workers.

Source of infection

In trachoma endemic zones the main source of infection is the conjunctival discharge of the affected person.

Modes of infection

Infection may spread from eye to eye by any of the following modes:

1. Spread of infection occurs through conduct by airborne or waterborne modes.

2. Vector transmission of trachoma is common through flies.

3. Material transfer. It can occur through contaminated fingers of doctors, nurses and contaminated tonometers. Other sources of material transfer of infection are: using common towels, handkerchiefs, bedding, etc.

Prevalence

Trachoma is a worldwide disease but it is highly prevalent in North Africa, Middle East and certain regions of South-East Asia. It is believed to affect some 500 million people in the world.

Clinical profile of trachoma

Incubation period of trachoma varies from 5–21 days. Onset of disease is usually insidious (sub-acute), however, rarely it may present in acute form. But, mostly the picture is complicated by secondary infection and may start with typical symptoms of acute conjunctivitis.

Grading of trachoma:

Stage I (incipient trachoma or stage of infiltration). It is characterized by *hyperemia of palpebral conjunctiva and immature follicles.*

Stage II (established trachoma or stage of florid infiltration). It is characterized by *appearance of mature follicles, papillae and progressive corneal pannus.*

Stage III (cicatrizing trachoma or stage of scarring). It includes *obvious scarring of palpebral conjunctiva.*

Stage IV (healed trachoma or stage of sequelae). The disease is cured but sequelae due to cicatrisation give rise to symptoms.

Complications of trachoma

1. *Lid complications (more in the upper lid)* – trichiasis (usually multiple), cicatricial entropion, mild ptosis due to paralysis of the Muller's muscle, chronic meibomianitis.

2. *Conjunctival complications* – posterior symblepharon (shallow fornix). It may obliterate ducts of the main lacrimal

gland leading to loss of reflex, xerosis due to atrophy of goblet cells.

3. *Corneal complications* – corneal ulcers, corneal opacities, complications of xerosis, Kera ectasia (very rare).

4. *Lacrimal complications* – fibrosis of lacrimal puncti, canaliculi or NLD leading to chronic dacryocystitis; dacryoadenitis.

Diagnosis

A. *The clinical diagnosis* of trachoma is made from its typical signs; at least two sets of signs should be present out of the following:

1. Conjunctival follicles and papillae.
2. Pannus progressive or regressive.
3. Epithelial keratitis near superior limbus.
4. Signs of cicatrisation or its sequelae.

B. *Laboratory diagnosis*. Laboratory diagnosis of trachoma includes:

1. *Conjunctival cytology.* Presence of plasma cells and Lieber cells is suggestive of trachoma.

2. *Detection of inclusion bodies* in conjunctival smear.

3. *Enzyme-linked immunosorbent assay* (ELISA) for chlamydial antigens.

4. *Polymerase chain reaction* (PCR) is also useful.

5. *Serotyping of TRIC agents* is done by detecting specific antibodies using micro immunofluorescence (micro-IF) method.

Direct monoclonal fluorescent antibody microscopy of conjunctival smear is rapid and affordable.

Treatment of active trachoma

Antibiotics for treatment of active trachoma may be given locally or systemically.

1. *Topical therapy regimes.* It is 1 % tetracycline, 1 % erythromycin, 0.3 % tobramycin eye ointment 4 times a day for 6 weeks; 20 % sulfacetamide, 0.3 % ofloxacin, 0.3 %

gentamicin, 0.3 % tobramycin eye drops three times a day for 6 weeks.

The *continuous treatment* for active trachoma should be followed by an *intermittent treatment* especially in endemic or hyperendemic area.

2. *Systemic therapy regimes:* tetracycline or erythromycin 250 mg orally, four times a day for 3–4 weeks, or doxycycline 100 mg orally twice daily for 3–4 weeks.

Treatment of trachoma sequelae

1. *Concretions* should be removed with a hypodermic needle.

2. *Trichiasis* may be treated by epilation, electrolysis or cryolysis.

3. *Entropion* should be corrected surgically.

4. *Xerosis* should be treated by artificial tears.

Prophylaxis

1. Hygienic measures.

2. Early treatment of conjunctivitis.

3. Blanket antibiotic therapy (intermittent treatment).

Viral Conjunctivitis

Viral conjunctivitis, or *pinkeye*, is a common, self-limiting condition that is typically caused by adenovirus. Other viruses that can be responsible for conjunctival infection include: herpes simplex virus (HSV), varicella-zoster virus (VZV), picornavirus (enterovirus 70, Coxsackie virus A24), poxvirus (molluscum contagiosum, vaccinia), and human immunodeficiency virus (HIV).

Viral infections of conjunctiva include:

– adenovirus conjunctivitis;
– herpes simplex keratoconjunctivitis;
– herpes zoster conjunctivitis;
– pox virus conjunctivitis;
– myxovirus conjunctivitis;

– paramyxovirus conjunctivitis;
– arbor virus conjunctivitis.

Clinical presentations may be presented in three clinical forms:

1. Acute serous conjunctivitis.
2. Acute hemorrhagic conjunctivitis.
3. Acute follicular conjunctivitis (see *follicular conjunctivitis*).

Acute serous conjunctivitis

Etiology

It is typically caused by a mild degree of viral infection, which does not give rise to follicular response.

Clinical features

Acute serous conjunctivitis is characterized by a minimal degree of congestion, a watery discharge and a boggy swelling of the conjunctival mucosa.

Treatment

Antiviral drugs: acyclovir, zovirax, ganciclovir, interferon eye drops may be used three times a day for about 7 days.

Acute haemorrhagic conjunctivitis

(Epidemic hemorrhagic conjunctivitis)

It is an acute inflammation of conjunctiva characterized by multiple conjunctival hemorrhages, conjunctival hyperemia and mild follicular hyperplasia.

Etiology

The disease is caused by picornaviruses (enterovirus type 70) which are RNA viruses of small (pico) size.

The disease has occurred in an epidemic form in the Far East, Africa and England and hence the name “epidemic hemorrhagic conjunctivitis (EHC)” has been suggested.

Incubation period of EHC is very short (1–2 days).

Symptoms and signs include pain, redness, watering, mild photophobia, transient blurring of vision, lid swelling, chemosis, multiple hemorrhages in bulbar conjunctiva, mild follicular hyperplasia, lid edema and pre-auricular lymphadenopathy.

Corneal involvement may occur in the form of fine epithelial keratitis.

Treatment

Antiviral drugs: acyclovir, zovirax, ganciclovir, interferon eye drops may be used three times a day for about 7 days.

NSAID: diclofenac, nimesil, ibuprofen or *voltaren* may be given orally for 2–3 days to provide symptomatic relief from mild pain especially in sensitive patients.

Follicular Conjunctivitis

It is the inflammation of conjunctiva, characterized by formation of follicles, conjunctival hyperemia and discharge from the eyes. Follicles are formed due to localized aggregation of lymphocytes in the adenoid layer of conjunctiva. Follicles appear as tiny, greyish white translucent, rounded swellings, 1–2 mm in diameter. Their appearance resembles boiled sago grains.

Types

1. Acute follicular conjunctivitis.
2. Chronic follicular conjunctivitis.
3. Specific type of conjunctivitis with follicle formation, e. g., trachoma.

Acute follicular conjunctivitis

It is an acute catarrhal conjunctivitis associated with follicular hyperplasia especially of the lower fornix and lower palpebral conjunctiva.

Signs and symptoms are similar to acute catarrhal conjunctivitis and include: redness, watering, mild mucoid discharge, mild photophobia and feeling of discomfort and foreign body sensation, conjunctival hyperemia, associated with multiple follicles, more prominent in lower lid than the upper lid.

Etiological types

Etiologically, acute follicular conjunctivitis is of the following types:

– adult inclusion conjunctivitis;
– epidemic keratoconjunctivitis;
– pharyngoconjunctival fever;
– Newcastle conjunctivitis;
– acute herpetic conjunctivitis.

Epidemic keratoconjunctivitis (EKC) is a type of acute follicular conjunctivitis mostly associated with superficial punctate keratitis and usually occurs in epidemics, hence the name EKC.

Etiology

It is caused by adenoviruses of type 8 and 19. The condition is markedly contagious and spreads through the contact with contaminated fingers, solutions and tonometers.

Clinical picture

Incubation period after infection is about 8 days and virus is shed from the inflamed eye for 2–3 weeks.

Clinical stages

The condition mainly affects young adults. Clinical picture can be arbitrarily divided into three stages for the purpose of description only:

– *acute serous conjunctivitis* is characterized by non-specific conjunctival hyperemia, mild chemosis and lacrimation;

– *typical acute follicular conjunctivitis* is characterized by formation of follicles which are more marked in lower lid;

– *acute pseudomembranous conjunctivitis* is recognized due to formation of a pseudomembrane on the conjunctival surface.

Corneal involvement is in the form of *superficial punctate keratitis, preauricular lymphadenopathy.*

Treatment

Antiviral drugs: acyclovir, zovirax, ganciclovir, interferon eye drops may be used three times a day for about 7 days.

NSAID: diclofenac, *nimesil*, *ibuprofen* or *voltaren* may be given orally for 2–3 days to provide symptomatic relief from mild pain especially in sensitive patients.

Pharyngoconjunctival fever (PCF)

Etiology

It is an adenoviral infection commonly associated with subtypes 3 and 7.

Clinical picture

Pharyngoconjunctival fever is characterized by an acute follicular conjunctivitis, associated with pharyngitis, fever and preauricular lymphadenopathy. The disease primarily affects children and appears in epidemic form. Corneal involvement in the form of superficial punctate keratitis is seen only in 30 % of cases.

Treatment

Because EKC and PCF are contagious and self-limiting, the primary treatment is patient education. Instruct patients to stay home from work or school until there is absolutely no discharge. Also instruct them not to share utensils, glasses, linens or wash cloths with others.

Medical management can range from cold compresses and artificial tears to topical vasoconstrictors

(e. g., naphazoline) and steroids (dexamethasone) two to four times daily. If a membrane is present, peel it off with a wet, cotton-tipped applicator or forceps. After removal, prescribe a topical antibiotic–steroid combination such as tobradex or maxitrol qid.

Newcastle conjunctivitis

Etiology

It is a rare type of acute follicular conjunctivitis caused by Newcastle virus. The infection is derived from contact with diseased owls; and thus the condition mainly affects poultry workers.

Clinically the condition is similar to pharyngo-conjunctival fever.

Acute herpetic conjunctivitis

Acute herpetic follicular conjunctivitis is always an accompaniment of the *primary herpetic infection*, which mainly occurs in small children and in adolescents.

Etiology

The disease is commonly caused by herpes simplex virus of type 1 and spreads by kissing or other close personal contacts. HSV type 2 associated with genital infections, may also involve the eyes in adults as well as children, though rarely.

Clinical picture

Incubation period is 3–10 days. It may occur in two clinical forms: typical and atypical.

In *the typical form*, the follicular conjunctivitis is usually associated with other lesions of primary infection such as vesicular lesions of face and lids.

In *the atypical form*, the follicular conjunctivitis occurs without lesions of the face, eyelid and the condition then resembles epidemic keratoconjunctivitis.

Corneal involvement, though rare, is not uncommon in primary herpes. It may be in the form of fine or coarse epithelial keratitis or typical dendritic keratitis.

Preauricular lymphadenopathy occurs almost always.

Treatment

Antiviral drugs: acyclovir, zovirax, ganciclovir, interferon eye drops may be used three times a day for about 7 days.

NSAID: diclofenac, *nimesil*, *ibuprofen* or *voltaren* may be given orally for 2–3 days to provide symptomatic relief from mild pain especially in sensitive patients.

Steroids: dexamethasone.

Chronic Follicular Conjunctivitis

It is a mild type of chronic catarrhal conjunctivitis associated with follicular hyperplasia, predominantly involving the lower lid.

Etiological types

1. *Infective chronic follicular conjunctivitis.*
2. *Toxic type of chronic follicular conjunctivitis.*
3. *Chemical chronic follicular conjunctivitis.*

The common topical preparations associated with chronic follicular conjunctivitis are: idoxuridine (IDU), eserine, pilocarpine, DFP and adrenaline.

4. *Chronic allergic follicular conjunctivitis.*

Ophthalmia Neonatorum

Ophthalmia neonatorum is the name given to bilateral inflammation of the conjunctiva occurring in an infant, less than 30 days old. It is a preventable disease usually occurring as a result of carelessness at the time of birth. As a matter of fact, *any discharge or even watering from the eyes in the first week of life should arouse suspicion of ophthalmia neonatorum*, as tears are not formed till then.

Etiology

Infection may occur in three ways: before birth, during birth or after birth.

Causative agents

1. *Chemical conjunctivitis:* it is caused by silver nitrate or antibiotics used for prophylaxis.

2. *Gonococcal infection.*

3. *Other bacterial infections* are Staphylococcus aureus, Streptococcus hemolytic, and Streptococcus pneumonia.

4. *Neonatal inclusion conjunctivitis* caused by serotypes D to K of *Chlamydia trachomatis* is the commonest cause of ophthalmia neonatorum in developed countries.

5. *Herpes simplex ophthalmia neonatorum* is a rare condition caused by herpes simplex-II virus.

Clinical features

Causative agent incubation period

1. Chemical 4–6 hours.

2. Gonococcal 2–4 days.

3. Other bacterial 4–5 days.

4. Neonatal inclusion conjunctivitis 5–14 days.

5. Herpes simplex 5–7 days.

Symptoms and signs

1. *Pain* and *tenderness* in the eyeball.

2. *Conjunctival discharge*. It is purulent in gonococcal ophthalmia neonatorum and mucoid or mucopurulent in other bacterial cases and neonatal inclusion conjunctivitis.

3. *Lids* are usually swollen.

4. *Conjunctiva* may show hyperemia and chemosis. There might be mild papillary response in neonatal inclusion conjunctivitis and herpes simplex ophthalmia neonatorum.

5. *Corneal involvement*, though rare, may occur in the form of superficial punctate keratitis especially in herpes simplex ophthalmia neonatorum.

Complications

Untreated cases, especially of gonococcal ophthalmia neonatorum, may develop corneal ulceration, which may perforate rapidly resulting in corneal opacification or staphyloma formation.

Treatment

Prophylactic treatment is always better than curative.

A. *Prophylaxis* needs antenatal, natal and postnatal care.

1. *Antenatal measures* include thorough care of mother and treatment of genital infections when suspected.

2. *Natal measures* are of utmost importance, as mostly infection occurs during childbirth.

Deliveries should be conducted under hygienic conditions taking all aseptic measures. The newborn baby's closed lids should be thoroughly cleansed and dried.

3. *Postnatal measures* include:

– use of either Floxal, Tobrex, 1 % tetracycline or 1 % erythromycin ointment or into the eyes of the babies immediately after birth;

– single injection of ceftriaxone 50 mg/kg IM or IV (not to exceed 125 mg) should be given to infants born to mothers with untreated gonococcal infection.

B. *Curative treatment.* As a rule, conjunctival cytology samples and culture sensitivity swabs should be taken before starting the treatment.

1. *Chemical ophthalmia neonatorum* is a self-limiting condition, and does not require any treatment.

2. *Gonococcal ophthalmia neonatorum* needs prompt treatment to prevent complications.

Systemic therapy

Neonates with gonococcal ophthalmia should be treated for 7 days with one of the following regimes: IV or IM, qid – ceftriaxone 75–100 mg/kg/day, cefotaxime 100–150 mg/kg/day

12 hourly, ciprofloxacin 10–20 mg/kg/day or norfloxacin 10 mg/kg/day.

If the gonococcal isolate is proved to be susceptible to penicillin, crystalline benzyl penicillin G 50 000 units to full term, normal weight babies and 20 000 units to premature or low weight babies should be given intramuscularly twice daily for 3 days.

Other bacterial ophthalmia neonatorum should be treated by broad spectrum antibiotic drops and ointments for 2 weeks.

Herpes simplex conjunctivitis is usually a self-limiting disease. However, topical antiviral drugs control the infection more effectively and may prevent the recurrence.

Allergic Conjunctivitis

It is the inflammation of conjunctiva due to allergic or hypersensitivity reactions which may be immediate (humoral) or delayed (cellular). The conjunctiva is ten times more sensitive than the skin to allergens.

Types

1. Simple allergic conjunctivitis:
   - hay fever conjunctivitis;
   - seasonal allergic conjunctivitis (sac);
   - perennial allergic conjunctivitis (pac).
2. Vernal keratoconjunctivitis (VKC).
3. Atopic keratoconjunctivitis (AKC).
4. Giant papillary conjunctivitis (GPC).
5. Phlyctenule keratoconjunctivitis (PKC).
6. Contact dermo conjunctivitis (CDC).

Simple allergic conjunctivitis

It is a mild, non-specific allergic conjunctivitis characterized by itching, edema of lids, hyperemia and mild

papillary response. Basically, it is an acute or subacute urticarial reaction.

Vernal keratoconjunctivitis (VKC), or spring catarrh

It is a recurrent, bilateral, interstitial, self-limiting, allergic inflammation of the conjunctiva having a periodic seasonal incidence (more common in summer and prevalent in tropics).

Symptoms

Spring catarrh is characterized by marked burning and itching sensation, mild photophobia, lacrimation, stringy (ropy) discharge and heaviness of lids.

Differential diagnosis

Palpebral form of VKC needs to be differentiated from trachoma with predominant papillary hypertrophy.

Atopic keratoconjunctivitis (AKC)

It can be thought of as an adult equivalent of vernal keratoconjunctivitis and is often associated with atopic dermatitis. Most of the patients are young atopic adults, with male predominance.

Signs and symptoms include: itching, soreness, dry sensation, mucoid discharge, photophobia or blurred vision, corneal vascularization, thinning and plaques.

Lid margins are chronically inflamed with rounded posterior borders.

Tarsal conjunctiva has a milky appearance. There are very fine papillae, hyperemia and scarring with shrinkage.

Giant papillary conjunctivitis (GPC)

It is the inflammation of conjunctiva with formation of very large sized papillae.

Etiology: it is a localized allergic response to a physically rough or deposited surface (contact lens, prosthesis,

left out nylon sutures). Probably it is a sensitivity reaction to components of the plastic leached out by the action of tears.

Symptoms: itching, stringy discharge and reduced wearing time of contact lens or prosthetic shell.

Signs: papillary hypertrophy (1 mm in diameter) of the upper tarsal conjunctiva, similar to that seen in palpebral form of VKC with hyperemia.

Phlyctenule keratoconjunctivitis (PKC)

It is a nodular affliction characterized by the formation of small circumscribed lesions at the corneal limbus that represents an allergic cell-mediated response within the conjunctiva and/or cornea to some antigen.

It was thought that the main antigen responsible for PKC was tuberculoprotein. However, the other sensitizing antigens were also reported in different sources, namely staphylococcal products, worm infestation, fungi, viruses, and parasites.

Pathology

1. Stage of nodule formation.
2. Stage of ulceration.
3. Stage of granulation.
4. Stage of healing.

Symptoms in simple phlyctenule conjunctivitis are few, like mild discomfort in the eye, irritation and reflex.

Contact dermo conjunctivitis

It is an allergic disorder, involving conjunctiva and skin of lids along with surrounding area of face.

Etiology

It is in fact a delayed hypersensitivity (type IV) response to prolonged contact with chemicals and drugs.

Clinical picture

1. *Cutaneous involvement* is in the form of weeping eczematous reaction, involving all areas with which medication comes in contact.

2. *Conjunctival response* is in the form of hyperemia with a generalized papillary response affecting the lower fornix and lower palpebral conjunctiva more than the upper.

Treatment of allergic conjunctivitis

1. *Local therapy.*

Antibiotic drops: 1 % oxytetracycline, 0.3 % tobramycin, 0.3 % gentamycin, 0.3 % floxal eye ointment and eye drops 2–3 times a day for 9–14 days will eradicate the infection.

Topical steroids in the form of eye drops or ointment (dexamethasone or betamethasone) should be added to take care of the associated secondary infection (mucopurulent conjunctivitis).

Atropine (1 %) eye ointment should be applied once daily when cornea is involved.

Topical antihistaminic: 2 % bromoform, cromoglicic acid (2 % ALERGOKROM, 2 % IFIRAL), 20 or 40 mg eye drops Lecrolyn, Opatanol eye drops three times a day for 2 weeks.

Mast cell stabilizers such as 2 % sodium cromoglycate drops 4–5 times a day are quite effective in controlling VKC, especially atopic cases. Azelastine eye drops are also effective in controlling VKC.

2. *Specific therapy.* Attempts must be made to search and eradicate the following causative conditions:

– *Tuberculous* infection should be excluded by X-rays of the chest, Mantoux test, TLC, DLC and ESR. In case a tubercular focus is discovered, antitubercular treatment should be started to combat the infection.

– *Septic focus* in the form of tonsillitis, adenoiditis, or caries teeth, when present should be adequately treated by systemic antibiotics and necessary surgical measures.

– *Parasitic infestation* should be ruled out by repeated stool examination, and when discovered should be adequately treated for complete eradication.

3. *General measures* aimed to improve the health of a child are equally important. Attempts should be made to provide high protein diet supplemented with vitamins A, C, D.

Granulomatous Conjunctivitis

Granulomatous conjunctivitis is the term used to describe certain specific chronic inflammations of the conjunctiva.

Granulomatous conjunctivitis in association with preauricular lymphadenopathy is known as *Parinaud's oculoglandular syndrome*. Bacteria such as Bartonella henselae (cat-scratch disease) and Francisella tularensis (tularemia), mycobacteria (tuberculosis), and treponemes (syphilis) are possible causes. The diagnosis can be made by serology, culture, polymerase chain reaction (PCR), or a combination of these. If conjunctival biopsy is performed, the granulomas in infectious granulomatous conjunctivitis will typically demonstrate central necrosis (caseation). The bacteria may be demonstrated with Gram, acid-fast, or Warthin–Starry stains, depending on the organism.

Common granulomatous conjunctival inflammations are:

– tuberculosis of conjunctiva;
– sarcoidosis of conjunctiva;
– syphilitic conjunctivitis;
– leprotic conjunctivitis;
– conjunctivitis in tularaemia;
– ophthalmia nodosa.

Parinaud's oculoglandular syndrome

It is the name given to a group of conditions characterized by:

- unilateral granulomatous conjunctivitis;
- preauricular lymphadenopathy;
- fever.

Ophthalmia nodosa (Caterpillar hair conjunctivitis)

It is a granulomatous inflammation of the conjunctiva characterized by formation of a nodule on the bulbar conjunctiva in response to irritation caused by the retained hair of caterpillar. The disease is, therefore, common in summers. The condition may be often mistaken for a tubercular nodule.

Histopathological examination reveals hair surrounded by giant cells and lymphocytes.

Treatment consists of excision biopsy of the nodule.

Degenerative Diseases of the Conjunctiva

Pinguecula

Pinguecula is an extremely common degenerative condition of the conjunctiva. It is characterized by formation of a yellowish white patch on the bulbar conjunctiva near the limbus. This condition is termed pinguecula because of its resemblance to fat, which means pinguis.

Etiology

Pinguecula is not known exactly. It has been considered as *an age-change*, occurring more commonly in persons exposed to strong sunlight, dust and wind. It is also considered a precursor of pterygium.

Clinical features

Pinguecula is a bilateral, usually stationary condition, presenting as yellowish white triangular patch near the limbs. Apex of the triangle is away from the cornea. It affects the

nasal side first and then the temporal side. When conjunctiva is congested, it stands out as an avascular prominence.

Complications

Pinguecula include its inflammation, intraepithelial abscess formation and rarely conversion into pterygium.

Treatment

In routine no treatment is required for pinguecula. However, if so desired, it may be excised.

Pterygium

Pterygium (Lat. *Pterygion* – a wing) is a wing-shaped fold of conjunctiva encroaching upon the cornea from either side within the internal pebral fissure.

Etiology

Etiology of pterygium is not definitely known. But the disease is more common in people living in hot climates. Therefore, the most accepted view is that it is a response to prolonged effect of environmental factors such as exposure to sun (ultraviolet rays), dry heat, high wind and abundance of dust.

Clinical features

Pterygium is more common in elderly males doing outdoor work.

It may be *unilateral* or *bilateral*. It presents as a triangular fold of conjunctiva injureing the cornea in the area of the palpebral aperture, usually on the nasal side, but may also occur on the temporal side. Deposition of iron seen sometimes in corneal epithelium anterior to advancing head of pterygium is called *Stockes line*.

A fully developed pterygium consists of three ***parts***:

– *head* (apical part present on the cornea);

– *neck* (limbal part);

– *body* (scleral part) extending between limbs and the canthus.

Types

Depending on the progression it may be progressive or regressive pterygium.

Progressive pterygium is thick, fleshy and vascular with a few infiltrates in the cornea, in front of the head of the pterygium (called "cap of pterygium").

Regressive pterygium is thin, atrophic, attenuated with very little vascularity. There is no cap. Ultimately it becomes membranous but never disappears.

Complications like cystic degeneration and infection are infrequent. Rarely, neoplastic change to epithelioma, fibrosarcoma or malignant melanoma may occur.

Differential diagnosis

Pterygium must be differentiated from pseudo pterygium.

Pseudo pterygium is a fold of bulbar conjunctiva attached to the cornea. It is formed due to adhesions of chemosed bulbar conjunctiva to the marginal corneal ulcer. It usually occurs following chemical burns of the eye.

Treatment

Surgical excision is the only satisfactory treatment, which may be indicated for:

– cosmetic reasons;

– continued progression threatening to encroach onto the pupillary area (once the pterygium has encroached pupillary area, wait till it crosses on the other side);

– diplopia due to interference in ocular movements.

Recurrence of the pterygium after surgical excision is the main problem (30–50 %).

Concretions

They are commonly seen in elderly people as a degenerative condition and also in patients with scarring stage of trachoma

Etiology

Concretions are formed due to accumulation of thickened mucus and dead epithelial cell debris into the conjunctival depressions called *loops of Henle*.

Clinical features

Concretions are seen on palpebral conjunctiva, more commonly on upper than the lower one. They may also be seen in lower fornix. These are yellowish white, looking hard, raised areas, varying in size from pin point to pin head. Being hard, they may produce foreign body sensations and lacrimation by rubbing the corneal surface. Occasionally they may even cause corneal abrasions.

Treatment

It consists of their removal with the help of a hypodermic needle under topical anesthesia.

Tumours of Conjunctiva

Conjunctival tumours are one of the most frequent tumours of the eye and adnexa. They comprise a large variety of conditions, from benign lesions to malignant lesions. Early diagnosis is essential for preventing ocular and systemic spread and to preserve visual function.

Classification

1. *Non-pigmented tumours*

I. *Congenital*: dermoid and lipodermoid (choristomas).

II. *Benign*: simple granuloma, papilloma, adenoma, fibroma and angiomas.

III. *Premalignant*: intraepithelial epithelioma (Bowen's disease).

IV. *Malignant*: epithelioma or squamous cell carcinoma, basal cell carcinoma.

2. *Pigmented tumours*

I. *Benign*: naevi or congenital moles.

II. *Precancerous melanosis*: superficial spreading melanoma and lentigo maligna (Hutchinson's freckle).

III. *Malignant*: primary melanoma (malignant melanoma).

Non-Pigmented Tumours

I. *Congenital tumours*

1. *Dermoid.* These are common congenital tumours which usually occur at the limbus. They appear as solid white masses, firmly fixed to the cornea. Dermoid consists of collagenous connective tissue, sebaceous glands and hair, lined by epidermoid epithelium.

Treatment is simple excision.

2. *Lipodermoid*. It is a congenital tumour, usually found at the limbs or outer canthus. It appears as soft, yellowish white, movable subconjunctival mass. It consists of fatty tissue and the surrounding dermis-like connective tissue, hence the name lipodermoid. Sometimes the epibulbar dermoids or lipodermoids may be associated with accessory auricles and other congenital defects (*Goldenhar's syndrome).*

II. *Benign tumours*

1. *Simple granuloma.* It consists of an extensive polypoid, cauliflower-like growth of granulation tissue. Simple granulomas are common following squint surgery, as foreign body granuloma and following inadequately scraped chalazion.

Treatment consists of complete surgical removal.

2. *Papilloma.* It is a benign polypoid tumour usually occurring at inner canthus, fornices or limbus. It may resemble the cocks comb type of conjunctival tubercular lesion. It has a tendency to undergo malignant change and hence needs complete excision.

3. *Fibroma.* It is a rare soft or hard polypoid growth usually occurring in lower fornix.

III. *Premalignant tumours*

Bowen's intraepithelial epithelioma (carcinoma in situ) is a rare, precancerous condition, usually occurring at the limbs as a flat, reddish grey, vascularized plaque. Histologically, it is confined within the epithelium. It should be treated by complete local excision.

IV. *Malignant tumours*

1. *Squamous cell carcinoma* (epithelioma). It usually occurs at the transitional zones, i. e. at limbs and the lid margin. The tumour invades the stromal deeply and may be fixed to underlying tissues. Histologically, it is similar to squamous cell carcinomas occurring elsewhere.

Treatment: early cases may be treated by complete local excision combined with extensive diathermy cautery of the area. However, in advanced and recurrent cases radical excision including enucleation or even exenteration may be needed along with postoperative radiotherapy.

2. *Basal cell carcinoma.* It may invade the conjunctiva from the lids or may appear pari-passu from the plica semilunaris or caruncle. Though it responds very favourably to radiotherapy, the complete surgical excision, if possible, should be preferred to avoid complications of radio-therapy.

Pigmented Tumours

1. *Naevi or congenital moles.* These are common pigmented lesions, usually presenting as grey gelatinous, brown or black, flat or slightly raised nodules on the bulbar conjunctiva, mostly near the limbus. They usually appear during early childhood and may increase in size at puberty or during pregnancy. Histologically, they resemble their cutaneous brethren. Malignant change is very rare and when it occurs we see its sudden increase in size or pigmentation, or appearance of signs of inflammation. Therefore, excision is usually indicated for cosmetic reasons and rarely for medical

reasons. Whatever may be the indication, excision should be complete.

2. *Precancerous melanosis*. Precancerous melanosis (intraepithelial melanoma) of conjunctiva occurs in adults as *superficial spreading melanoma*. It never arises from a congenital nevus.

Clinically a small pigmented tumour develops at any site on the bulbar or palpebral conjunctiva, which spreads as a diffuse, flat, asymptomatic pigmented patch. As long as it maintains its superficial spread, it does not metastasize. However, ultimately in about 20 percent cases it involves the sub epithelial tissues and proceeds to frank malignant change.

Treatment: in early stages local excision with postoperative radiotherapy may be sufficient. But in case of recurrence, it should be treated as malignant melanoma.

3. *Malignant melanoma (primary melanoma)*. It mostly appears de-novo, usually near the limbus, or rarely may occur due to malignant change in pre-existing nevus.

The condition usually occurs in elderly patients.

Clinically it may present as pigmented or nonpigmented mass near limbs or on any other part of the conjunctiva. It spreads over the surface of the globe and rarely penetrates it. Distant metastasis occurs elsewhere in the body, commonly in liver.

Histologically, the neoplasm may be alveolar, round celled or spindle-celled.

Treatment

Once suspected, enucleation or exenteration is the treatment of choice, depending upon the extent of growth.

TOPIC 8. DISEASES OF THE CORNEA AND SCLERA

Classification of the corneal diseases:

I. Inflammatory (keratitis).
II. Degenerative diseases.
III. Ectatic corneal conditions.
IV. Miscellaneous subjects.

Inflammations of the Cornnea

Characteristics of inflammations of the cornea are corneal edema, cellular infiltration and ciliary congestion.

Classification

It is difficult to classify and assign a group to each and every case of keratitis; as overlapping or concurrent findings tend to obscure the picture.

However, the following simplified topographical and etiological classifications provide a workable knowledge.

Topographical (morphological) classification

Ulcerative keratitis (corneal ulcer)

Corneal ulcer can be further classified variously.

1. *Depending on location:*
- central corneal ulcer;
- peripheral corneal ulcer.

2. *Depending on purulence:*
- purulent corneal ulcer or suppurative corneal ulcer;
- non-purulent corneal ulcers.

3. *Depending upon association of hypopyon:*
- simple corneal ulcer (without hypopyon);
- hypopyon corneal ulcer.

4. *Depending upon depth of ulcer:*
- superficial corneal ulcer;
- deep corneal ulcer;
- corneal ulcer with impending perforation;
- perforated corneal ulcer.

5. *Depending upon slough formation:*
– non-sloughing corneal ulcer;
– sloughing corneal ulcer.

Non-ulcerative keratitis

1. *Superficial keratitis:*
– diffuse superficial keratitis;
– superficial punctate keratitis (SPK).

2. *Deep keratitis:*
– non-suppurative:
  · interstitial keratitis;
  · disciform keratitis;
  · sclerosing keratitis;
– suppurative deep keratitis:
  · central corneal abscess;
  · posterior corneal abscess.

Etiological classification

1. *Infective keratitis:*
– bacterial;
– viral;
– fungal;
– chlamydial;
– protozoal;
– spirochetal.

2. *Allergic keratitis:*
– phlyctenule keratitis;
– vernal keratitis;
– atopic keratitis.

3. *Trophic keratitis:*
– exposure keratitis;
– neuroparalytic keratitis;
– keratomalacia;
– atheromatous ulcer.

4. ***Keratitis associated with diseases of skin and mucous membrane.***

5. ***Keratitis associated with systemic collagen vascular disorders.***

6. ***Traumatic keratitis***, which may be due to mechanical trauma, chemical trauma, thermal burns, radiations.

7. ***Idiopathic keratitis:***

– Mooren's corneal ulcer;

– superior limbic keratoconjunctivitis;

– Thygeson's superficial punctate keratopathy (TSPK, also Thygeson Superficial Punctate Keratitis).

Ulcerative Keratitis

Corneal ulcer may be defined as discontinuation in normal epithelial surface of cornea associated with necrosis of the surrounding corneal tissue.

Pathologically it is characterized by edema and cellular infiltration. Common types of corneal ulcers are described below.

Infective Keratitis

Bacterial Corneal Ulcer

Being the most anterior part of an eyeball, the cornea is exposed to atmosphere and hence prone to get infected easily. At the same time cornea is protected from the day-to-day minor infections by the normal defense mechanisms present in tears in the form of lysozyme, beta lysin, and other protective proteins.

Therefore, infective corneal ulcer may develop when:

– either the local ocular defense mechanism is jeopardized;

– there is some local ocular predisposing disease, or host's immunity is compromised;

– the causative organism is very virulent.

Etiology

There are two main factors in the production of purulent corneal ulcer:

– damage to corneal epithelium;

– infection of the eroded area.

However, the following three pathogens can invade the intact corneal epithelium and produce ulceration: *Neisseria gonorrhoeae, Corynebacterium diphtheriae* and *Neisseria meningitidis.*

1. *Corneal epithelial damage*. It is a prerequisite for most of the infecting organisms to produce corneal ulceration. It may occur in the following conditions:

– *corneal abrasion* due to small foreign body, misdirected cilia, concretions and trivial trauma in contact lens wearers or otherwise;

– *epithelial drying* as in xerosis and exposure keratitis;

– *necrosis of epithelium* as in keratomalacia;

– *desquamation of epithelial cells* as a result of corneal edema as in bullous keratopathy;

– *epithelial damage due to trophic changes* as in neuroparalytic keratitis.

2. *Source of infection* includes:

Exogenous infection. Most of the times corneal infection arises from exogenous source like conjunctival sac, lacrimal sac (dacryocystitis), infected foreign bodies, infected vegetative material and water-borne or air-borne infections.

From the ocular tissue. Owing to direct anatomical continuity, diseases of the conjunctiva readily spread to corneal epithelium, those of sclera to stroma, and of the uveal tract to the endothelium of cornea.

Endogenous infection. Owing to avascular nature of the cornea, endogenous infections are of rare occurrence.

3. *Causative organisms.* Common bacteria associated with corneal ulceration are: staphylococcus aureus,

pseudomonas pyocyanea, streptococcus pneumoniae, E. coli, proteus, Klebsiella, n. gonorrhea, n. meningitidis and c. diphtheriae.

Pathogenesis and pathology of the corneal ulcer

Depending upon the prevalent circumstances the course of corneal ulcer may take one of the three forms. Ulcer may:

– become localized and heal;
– penetrate deep leading to corneal perforation;
– spread fast in the whole cornea as sloughing corneal ulcer.

The salient pathological features of these are:

Pathology of localized corneal ulcer

1. *Stage of progressive infiltration.*
2. *Stage of active ulceration*.
3. *Stage of regression.*
4. *Stage of cicatrization.*

Pathology of perforated corneal ulcer

Perforation of corneal ulcer occurs when the ulcerative process deepens and reaches up to the Descemet's membrane. This membrane is tough and bulges out as descemetocele. At this stage, any exertion on the part of patient, such as coughing, sneezing, straining for stool, etc. will perforate the corneal ulcer. Immediately after perforation, the aqueous escapes, intraocular pressure falls and the iris-lens diaphragm moves forward.

Pathology of sloughing corneal ulcer and formation of anterior staphyloma

The iris becomes inflamed, and exudates block the pupil and cover the iris surface; thus a *false cornea* is formed. Ultimately these exudates organize and form a thin fibrous layer over which the conjunctival or corneal epithelium rapidly grows and thus a *pseudo cornea* is formed.

This ectatic cicatrix is called *anterior staphyloma* which, depending upon its extent, may be either partial or total.

The bands of scar tissue on the staphyloma vary in breadth and thickness, producing a lobulated surface often blackened with iris tissue which resembles a bunch of black grapes (hence the name staphyloma).

Clinical picture

In bacterial infections the outcome depends upon the virulence of organism, its toxins and enzymes, and the response of host tissue.

Broadly bacterial corneal ulcers may manifest as:

– purulent corneal ulcer without hypopyon;
– hypopyon corneal ulcer.

Symptoms

1. *Pain* and *foreign body sensation* occurs due to mechanical effects of lids and chemical effects of toxins on the exposed nerve endings.

2. *Watering from the eye* occurs due to reflex hyper lacrimation.

3. *Photophobia*, i. e. intolerance to light results from stimulation of nerve endings.

4. *Blurred vision* results from corneal haze.

5. *Redness of eyes* occurs due to congestion of circumcorneal vessels.

Signs

1. *Lids* are swollen.

2. Marked *blepharospasm* may be there.

3. *Conjunctiva* is chemosed and shows conjunctival hyperemia and ciliary congestion.

4. *Corneal ulcer* usually. Bacterial ulcer is characterized by:

– yellowish-white area of ulcer which may be oval or irregular in shape;
– margins of the ulcer are swollen and over hanging;
– floor of the ulcer is covered by necrotic material;
– stromal edema is present surrounding the ulcer area.

Characteristic features produced by some of the causative bacteria are as follows: *Staphylococcal aureus and streptococcus pneumonia; Pseudomonas species; Enterobacteria* (E. coli, Proteus sp., and Klebsiella sp.).

5. *Anterior chamber* may or may not show pus (*hypopyon).* In bacterial corneal ulcers the hypopyon remains sterile so long as the Descemet's membrane is intact.

6. *Iris* may be slightly muddy in colour.

7. Pupil may be small due to associated toxin-induced iritis.

8. Intraocular pressure may sometimes be raised (inflammatory glaucoma).

Hypopyon Corneal Ulcer

Etiology

Many pyogenic organisms (staphylococci, streptococci, gonococci, Moraxella) may produce hypopyon, but by far the most dangerous are *pseudomonas pyocyanea* and *pneumococcus.*

Source of infection for pneumococcal infection is usually the chronic dacryocystitis.

Symptoms are the same as described above for bacterial corneal ulcer. However, it is important to note that during initial stage of ulcus Serpens there is remarkably little pain. As a result, the treatment is often unduly delayed.

Signs

In general, the signs are the same as described above for the bacterial ulcer. *Typical features of ulcus Serpens* are:

– ulcus Serpens is a greyish white or yellowish disc shaped ulcer occurring near the centre of cornea;

– the ulcer has a tendency to creep over the cornea in a serpiginous fashion. One edge of the ulcer, along which the ulcer spreads, shows more infiltration. The other side of the

ulcer may be undergoing simultaneous cicatrization and the edges may be covered with fresh epithelium;

– violent iridocyclitis is commonly associated with a definite hypopyon;

– hypopyon increases in size very rapidly and often results in secondary glaucoma;

– ulcer spreads rapidly and has a great tendency for early perforation.

Treatment

It is the same as for other bacterial corneal ulcer. *Special points* which must be considered are:

– *secondary glaucoma* should be anticipated and treated with 0.5 % timolol maleate, eye drops and oral acetazolamide;

– *source of infection*, i. e. chronic dacryocystitis if detected, should be treated by dacryocystectomy.

Complications of the corneal ulcer:

1. Toxic iridocyclitis.
2. Secondary glaucoma.
3. Descemetocele.
4. Perforation of corneal ulcer.
5. Corneal scarring.

Clinical evaluation

Each case with corneal ulcer should be subjected to:

1. *Thorough history taking* to elicit mode of onset, duration of disease and severity of symptoms.

2. *General physical examination*, especially nourishment, anemia and any immune compromising disease.

3. *Ocular examination* should include:

– *diffuse light examination* for gross lesions of the lids, conjunctiva and cornea including testing for sensations;

– *regurgitation test and syringing* to rule out lacrimal sac infection;

– *biomicroscopic examination* after staining of corneal ulcer with 2 % freshly prepared aqueous solution of fluorescein dye or sterilized fluorescein impregnated filter paper strip to note site, size, shape, depth, margin, floor and vascularization of corneal ulcer. On biomicroscope, also note presence of precipitates at the back of cornea, depth and contents of anterior chamber, colour and pattern of iris and condition of crystalline lens.

Laboratory investigations

Routine laboratory investigations such as hemoglobin, TLC, DLC, ESR, blood sugar, complete urine and stool examination should be carried out in each case.

Microbiological investigations

These studies are essential to identify causative organism, confirm the diagnosis and guide the treatment to be instituted.

Material for such investigations is obtained by scraping the base and margins of the corneal ulcer (under local anesthesia, using 2 % xylocaine) with the help of a modified Kimura spatula or by simply using the bent tip of a 20-gauge hypodermic needle. The material obtained is used for the following investigations:

– Gram and Giemsa stained smears for possible identification of infecting organisms;

– 10 % Koh wet preparation for identification of fungal hyphae;

– culture on blood agar medium for aerobic organisms;

– culture on Sabouraud Dextrose Agar (SDA) medium for fungi.

Treatment

I. *Treatment of uncomplicated corneal ulcer*

Bacterial corneal ulcer is a vision threatening condition that demands urgent treatment by identification and eradication of causative bacteria.

Treatment of corneal ulcer can be discussed under three headings:

1. Specific treatment for the cause.
2. Non-specific supportive therapy.
3. Physical and general measures.

1. The specific treatment

Topical antibiotics: initial therapy (before the results of culture and sensitivity are available) should be with combination therapy to cover both gram-negative and gram-positive organisms.

It is preferable to start fortified 14 mg/ml gentamycin or fortified 14mg/ml tobramycin eye drops along with fortified 50 mg/ml cephazolin, every ½ to one hour for the first few days and then reduced to twice hourly. Once the favourable response is obtained, the fortified drops can be substituted by more diluted commercially available eye-drops, e. g.:

- 0.3 % ciprofloxacin eye drops;
- 0.3 % ofloxacin eye drops;
- 0.3 % gemifloxacin eye drops.

Systemic antibiotics are usually not required. However, a cephalosporin and an aminoglycoside or oral 750 mg ciprofloxacin twice daily may be given in fulminating cases with perforation and when sclera is also involved.

2. Non-specific treatment

Cycloplegic drugs: preferably 1 % atropine eye drops or ointment should be used to reduce pain from ciliary spasm and to prevent the formation of posterior synechiae from secondary iridocyclitis.

Systemic analgesics and anti-inflammatory drugs such as paracetamol and ibuprofen relieve the pain and decrease edema.

Vitamins (A, B-complex and C) help in early healing of ulcer.

3. Physical and general measures

Hot fomentation: local application of heat (preferably dry) gives comfort, reduces pain and causes vasodilatation.

Dark goggles may be used to prevent photophobia.

Rest, good diet and *fresh air* may have a soothing effect.

II. *Treatment of non-healing corneal ulcer*

If the ulcer progresses despite the above therapy the following additional measures should be taken:

1. *Removal of any known cause of non-healing ulcer.* A thorough search for any already missed causes not allowing healing should be made and when found, such factors should be eliminated.

Common causes of non-healing ulcers are as follows:

Local causes: associated raised intraocular pressure, concretions, misdirected cilia, impacted foreign body, dacryocystitis, inadequate therapy, wrong diagnosis, lagophthalmos and excessive vascularization of ulcer.

Systemic causes: diabetes mellitus, severe anemia, malnutrition, chronic debilitating diseases and patients on systemic steroids.

2. *Mechanical debridement of ulcer* or removing necrosed material by scraping floor of the ulcer with a spatula under local anesthesia may hasten the healing.

3. *Cauterization of the ulcer* may also be considered in non-responding cases. Cauterization may be performed with pure carbolic acid or 10–20 % trichloroacetic acid.

4. *Bandage soft contact lens* may also help in healing.

5. *Peritomy*, i. e. severing of perilimbal conjunctival vessels may be performed when excessive corneal vascularization is hindering healing.

III. *Treatment of impending perforation*

When ulcer progresses and perforation seems imminent, the following additional measures may help to prevent perforation and its complications:

1. *No strain.* The patient should be advised to avoid sneezing, coughing and straining during stool, etc. He should be advised strict bed rest.

2. *Pressure bandage* should be applied to give some external support.

3. *Lowering of intraocular pressure* by simultaneous use of 250 mg acetazolamide qid orally, intravenous 20 % mannitol drip stat, oral glycerol twice a day, 0.5 % timolol eye drops twice a day, and even paracentesis with slow evacuation of aqueous from the anterior chamber may be performed if required.

4. *Tissue adhesive glue* such as cyanoacrylate is helpful in preventing perforation.

5. *Conjunctival flap.* The cornea may be covered completely or partly by a conjunctival flap to give support to the weak tissue.

6. *Bandage soft contact lens* may also be used.

7. *Penetrating therapeutic keratoplasty* (tectonic graft) may be undertaken in suitable cases, when available.

IV. *Treatment of perforated corneal ulcer*

Best is to prevent perforation. However, if perforation has occurred, immediate measures should be taken to restore the integrity of perforated cornea.

Depending upon the size of perforation and availability, measures like use of tissue adhesive glues, covering with conjunctival flap, use of bandage soft contact lens or therapeutic keratoplasty should be undertaken. Best is an urgent therapeutic keratoplasty.

Marginal Catarrhal Ulcer

These superficial ulcers situated near the limbs are frequently seen especially in old people.

Etiology: marginal catarrhal ulcer is thought to be caused by a hypersensitivity reaction to staphylococcal toxins.

It occurs in association with chronic staphylococcal blepharoconjunctivitis. Moraxella and Hemophilus are also known to cause such ulcers.

Clinical features

1. Patient usually presents with mild ocular irritation, pain, photophobia and watering.

2. The ulcer is shallow, slightly infiltrated and often multiple, usually associated with staphylococcal conjunctivitis.

3. Soon vascularization occurs followed by resolution. Recurrences are very common.

Treatment

1. A short course of topical corticosteroid drops along with adequate antibiotic therapy often heals the condition.

2. Adequate treatment of associated blepharitis and chronic conjunctivitis is important to prevent recurrences.

Mycotic Corneal Ulcer

The incidence of suppurative corneal ulcers caused by fungi has increased in the recent years due to injudicious use of antibiotics and steroids.

Etiology

1. *Causative fungi*. The fungi which may cause corneal infections are:

– *Filamentous fungi,* e. g., Aspergillus, Fusarium, Alternaria, Cephalosporium, Curvularia and Penicillium.

– *Yeasts*, e. g., Candida and Cryptococcus. (The fungi more commonly responsible for mycotic corneal ulcers are Aspergillus (most common), Candida and Fusarium).

2. Modes of infection*: injury by vegetative material* such as crop leaf, branch of a tree, straw, hay or decaying vegetable matter. Common sufferers are field workers, especially during harvesting season.

Injury by animal tail is another mode of infection.

Secondary fungal ulcers are common in patients who are immunosuppressed systemically or locally such as patients suffering from dry eye, herpetic keratitis, bullous keratopathy or postoperative cases of keratoplasty.

3. *Role of antibiotics and steroids*. Antibiotics disturb the symbiosis between bacteria and fungi; and the steroids make the fungi facultative pathogens which are otherwise symbiotic saprophytes. Therefore, excessive use of these drugs predisposes the patients to fungal infections.

Symptoms are similar to the central bacterial corneal ulcer, but in general they are less marked than the equal-sized bacterial ulcer and the overall course is slow and torpid.

Signs

A typical fungal corneal ulcer has the following salient features:

– corneal ulcer is *dry-looking*, *greyish white*, with *elevated rolled out margins*;

– delicate *feathery finger-like extensions* are present into the surrounding stroma under the intact epithelium;

– a *sterile immune ring* (yellow line of demarcation) may be present where fungal antigen and host antibodies meet;

– multiple, small *satellite lesions* may be present around the ulcer;

– usually a *big hypopyon* is present even if the ulcer is very small;

– unlike bacterial ulcer, the hypopyon may not be sterile as the fungi can penetrate into the anterior chamber without perforation;

– *perforation* in mycotic ulcer is rare but can occur;

– *corneal vascularization* is conspicuously absent.

Diagnosis

1. *Typical clinical manifestations* associated with history of injury by vegetative material are diagnostic of a mycotic corneal ulcer.

2. *Chronic ulcer worsening* in spite of the most efficient treatment should arouse suspicion of mycotic involvement.

3. *Laboratory investigations* required for confirmation, include examination of wet KOH, Calcofluor white, Gram's and Giemsa-stained films for fungal hyphae and culture on Sabouraud's agar medium.

Treatment

I. *Specific treatment* includes antifungal drugs

1. *Topical antifungal eye drops* should be used for a long period (6 to 8 weeks). These include:

- 5 % *natamycin* eye drops;
- 0.2 % *fluconazole* eye drops;
- 3.5 % *nystatin* eye ointment.

2. *Systemic antifungal drugs* may be required for severe cases of fungal keratitis. Tablet of *fluconazole* or *ketoconazole* may be given for 2–3 weeks.

II. *Nonspecific treatment.* Non-specific treatment and general measures are similar to those of bacterial corneal ulcer.

III. *Therapeutic penetrating keratoplasty* may be required for unresponsive cases.

Viral Corneal Ulcers

Most of the viruses tend to affect the epithelium of both the conjunctiva and cornea; hence the typical viral lesions constitute the viral keratoconjunctivitis.

Common viral infections include herpes simplex keratitis, herpes zoster ophthalmicus and adenovirus keratitis.

Herpes Simplex Keratitis

Ocular infections with herpes simplex virus (HSV) are extremely common and constitute herpetic keratoconjunctivitis and iritis.

Etiology: it is a DNA virus. Its only natural host is human. Basically HSV is epitheliotropic but may become neurotropic.

According to different clinical and immunological properties, HSV is of two types, namely:

– HSV type I typically causes infection above the waist;

– HSV type II causes infection below the waist (herpes genitals). HSV-II has also been reported to cause ocular lesions.

Mode of Infection

HSV-1 infection: it is acquired by kissing or coming in close contact with a patient suffering from herpes labialis.

HSV-II infection: it is transmitted to eyes of neonates through infected genitalia of the mother.

Ocular lesions of herpes simplex occur in two forms, *primary* and *recurrent*, with the following lesions:

Primary herpes

1. *Skin lesions*.
2. *Conjunctiva-acute follicular conjunctivitis*.
3. *Cornea*:

– fine epithelial punctate keratitis;
– coarse epithelial punctate keratitis;
– dendritic ulcer.

Recurrent herpes

1. *Active epithelial keratitis*:

– punctate epithelial keratitis;
– dendritic ulcer;
– geographical ulcer.

2. *Stromal keratitis*:

– disciform keratitis;
– diffuse stromal necrotic keratitis.

3. *Trophic keratitis (meta-herpetic).*
4. *Herpetic iridocyclitis*.

Primary ocular herpes

Primary infection (first attack) involves a non-immune person. It typically occurs in children between 6 months and 5 years of age and in teenagers.

Clinical features

1. *Skin lesions.* Vesicular lesions may occur involving skin of lids, periorbital region and the lid margin (vesicular blepharitis).

2. *Acute follicular conjunctivitis* with regional lymphadenitis is the usual and sometimes the only manifestation of the primary infection.

3. *Keratitis.* Cornea is involved in about 50 percent of the cases. Primary infection is usually self-limiting but the virus travels up to the trigeminal ganglion and establishes the latent infection.

Recurrent ocular herpes

The virus which lies dormant in the trigeminal ganglion periodically reactivates and causes recurrent infection.

Predisposing stress stimuli, which trigger an attack of herpetic keratitis, include: fever such as malaria, flu, exposure to ultraviolet rays, general ill-health, emotional or physical exhaustion, mild trauma, menstrual stress, following administration of topical or systemic steroids and immunosuppressive agents.

Epithelial keratitis

Punctate epithelial keratitis. The initial epithelial lesions of recurrent herpes resemble those seen in primary herpes and may be either in the form of fine or coarse superficial punctate lesions.

Dendritic ulcer. Dendritic ulcer is a typical lesion of recurrent epithelial keratitis. The ulcer is of an irregular, zigzag linear branching shape.

Geographical ulcer. Sometimes, the branches of dendritic ulcer enlarge and coalesce to form a large epithelial

ulcer with a "geographical" or "amoeboid" configuration, hence the name. The use of steroids in dendritic ulcer hastens the formation of geographical ulcer.

Symptoms of epithelial keratitis are: photophobia, lacrimation, pain.

Disciform stromal keratitis

Pathogenesis

It is due to delayed hypersensitivity reaction to the HSV antigen. There occurs low grade stromal inflammation and damage to the underlying endothelium. Endothelial damage results in corneal edema due to imbibition of aqueous humor.

Signs

Disciform keratitis is characterized by:

– focal disc-shaped patch of stromal edema without necrosis;

– folds in descemet's membrane;

– keratic precipitates;

– ring of stromal infiltrate (Wesley immune ring) may be present surrounding the stromal edema;

– corneal sensations are diminished;

– intraocular pressure (IOP) may be raised despite only mild anterior uveitis; in severe cases, anterior uveitis may be marked;

– sometimes epithelial lesions may be associated with disciform keratitis.

Diffuse stromal necrotic keratitis

It is a type of interstitial keratitis caused by active viral invasion and tissue destruction.

Symptoms

Pain, photophobia and redness are common symptoms.

Signs

It presents as necrotic, blotchy, cheesy white infiltrates that may lie under the epithelial ulcer or may present independently under the intact epithelium. It may be associated

with mild iritis and keratic precipitates. After several weeks of smoldering inflammation, stromal vascularization may occur.

Metaherpetic keratitis

Metaherpetic keratitis (epithelial sterile trophic ulceration) is not an active viral disease, but is a mechanical healing problem (similar to recurrent traumatic erosions) which occurs at the site of a previous herpetic ulcer.

Clinically it presents as an indolent linear or ovoid epithelial defect.

Treatment of herpes simplex keratitis

I. *Specific treatment*

Antiviral drugs are the first choice presently: 15 % ganciclovir gel, zovirax, 3 % acyclovir ointment 5 times a day until ulcer heals and then 3 times a day for 5 days. It is least toxic and most commonly used antiviral drug.

NSAID: diclofenac, nimesil, ibuprofen or *voltaren* may be given orally for 2–3 days to provide symptomatic relief from mild pain especially in sensitive patients.

Topical antihistaminic: 2 % kromofarm, 2 % alerhokrom, 2 % ifiral, 20 or 40 mg lekrolin eye drops, opatanol eye drops three times a day for 2 weeks.

Corneal protectors: corneregel and ophtagel are gels for nighttime protection.

2. *Mechanical debridement* of the involved area along with a rim of surrounding healthy epithelium with the help of sterile cotton applicator.

II. *Non-specific supportive therapy* as well as physical and general measures is the same as for the bacterial corneal ulcer.

Herpes Zoster Ophthalmicus

Herpes zoster ophthalmicus is an acute infection of Gasserian ganglion of the fifth cranial nerve by the varicella-

zoster virus (VZV). It constitutes approximately 10 percent of all cases of herpes zoster.

Etiology

Varicella-zoster virus is a DNA virus and produces acidophilic intranuclear inclusion bodies. It is neurotropic in nature.

Mode of infection

The infection is contracted in childhood, which manifests as chickenpox and the child develops immunity. The virus then remains dormant in the sensory ganglion of trigeminal nerve.

It is thought that, usually in elderly people (can occur at any age) with depressed cellular immunity, the virus reactivates, replicates and travels down along one or more of the branches of the ophthalmic division of the fifth nerve.

Clinical features:

– in herpes zoster ophthalmicus, the frontal nerve is more frequently affected than the lacrimal and nasociliary nerves;

– about 50 percent cases of herpes zoster ophthalmicus get ocular complications;

– the Hutchinson's rule, which implies that ocular involvement is frequent if the side or tip of nose presents vesicles (cutaneous involvement of nasociliary nerve), is useful but not infallible;

– lesions of herpes zoster are strictly limited to one side of the midline of head.

Clinical phases of herpes zoster ophthalmicus are:

– *acute*, which may totally resolve;

– *chronic*, which may persist for years;

– *relapsing*, where the acute or chronic lesions reappear sometimes years later.

Clinical features of herpes zoster ophthalmic include general features, cutaneous lesions and ocular lesions. In

addition, there may be associated other neurological complications.

The onset of illness is sudden with fever, malaise and severe neuralgic pain along the course of the affected nerve. The distribution of pain is so characteristic of zoster.

Cutaneous lesions in the area of distribution of the involved nerve appear usually after 3–4 days of onset of the disease. To begin with, the skin of lids and other affected areas become red and edematous (mimicking erysipelas), followed by vesicle formation. In due course of time vesicles are converted into pustules, which subsequently burst to become crusting ulcers. When crusts are shed, permanent pitted scars are left. The active eruptive phase lasts for about 3 weeks. Main symptom is severe neuralgic pain which usually diminishes with the subsidence of eruptive phase; but sometimes it may persist for years with little diminution of intensity. There occurs some anesthesia of the affected skin which when associated with continued post-herpetic neuralgia is called *anesthesia dolorosa*.

Ocular complications usually appear at the subsidence of skin eruptions and may present as a combination of two or more of the following lesions:

1. *Conjunctivitis* is one of the most common complications of herpes zoster. It may occur as mucopurulent conjunctivitis with petechial hemorrhages or acute follicular conjunctivitis with regional lymphadenopathy. Sometimes, severe necrotizing membranous inflammation may be seen.

2. *Zoster keratitis* occurs in 40 percent of all patients and sometimes may precede the neuralgia or skin lesions. It may occur in several forms, which in order of chronological clinical occurrence are: *punctate epithelial keratitis, micro dendritic epithelial ulcers, nummular disciform keratitis, neuroparalytic ulceration, mucous plaque keratitis.*

Associated neurological complications

Herpes zoster ophthalmic may also be associated with other neurological complications such as: *motor nerve palsies, optic neuritis, encephalitis, episcleritis and scleritis, iridocyclitis, acute retinal necrosis, anterior segment necrosis and phthisis bulbi, secondary glaucoma.*

Treatment

I. *Systemic therapy for herpes zoster*

1. *Oral antiviral drugs:* 800 mg *acyclovir,* 500 mg *valaciclovir* 5 times a day for 10 days.

2. *Analgesics.* Pain during the first 2 weeks of an attack is very severe and should be treated by analgesics such as combination of mephenamic acid and paracetamol or pentazocine or even pethidine (when very severe).

3. *Systemic steroids.* They appear to inhibit development of post-herpetic neuralgia when given in high doses. Steroids are commonly recommended in cases developing neurological complications such as the third nerve palsy and optic neuritis.

4. 300 mg *cimetidine* qid for 2–3 weeks starting within 48–72 hours of onset has also been shown to reduce pain and pruritus in acute zoster – presumably by histamine blockade.

5. *Amitriptyline* should be used to relieve the accompanying depression in acute phase.

II. *Local therapy for skin lesions*

1. *Antibiotic–corticosteroid skin ointment* or *lotions.*

These should be used three times a day till skin lesions heal.

2. *No calamine lotion.* Cool zinc calamine application, as advocated earlier, is better avoided, as it promotes crust formation.

III. *Local therapy for ocular lesions*

1. *For zoster keratitis, iridocyclitis* and *scleritis*:

– *topical steroids*, in the form of eye drops or ointment (dexamethasone or betamethasone) eye drops 4 times a day;

– *cycloplegic* such as cyclopentolate eye drops or atropine eye ointment;

– *topical antiviral drugs* – acyclovir, zovirax, ganciclovir, interferon eye drops may be used three times a day for about 2 weeks.

2. *To prevent secondary infections* topical antibiotics are used.

3. *For secondary glaucoma* 0.5 % timolol or 0.5 % betaxolol drops, 250 mg acetazolamide qid.

4. *For neuroparalytic corneal ulcer* caused by herpes zoster, lateral tarsorrhaphy should be performed.

5. *For persistent epithelial defects* use lubricating artificial tear drops (corneregel, ophtagel) and bandage soft contact lens.

6. *Keratoplasty* may be required for visual rehabilitation of zoster-patients with dense scarring. However, these are poor risk patients.

Protozoal Keratitis

Acanthamoeba keratitis

Acanthamoeba keratitis has recently gained importance because of its increasing incidence, difficulty in diagnosis and unsatisfactory treatment.

Etiology

Acanthamoeba is a free lying amoeba found in soil, fresh water, well water, sea water, sewage and air. It exists in trophozoite and encysted forms.

Mode of infection

Corneal infection with acanthamoeba results from direct corneal contact with any material or water contaminated with the organism.

Following situations of contamination have been described:

1. *Contact lens wearers* using home-made saline (from contaminated tap water and saline tablets) is the commonest situation recognized for acanthamoeba infection in western countries.

2. Other situations include *mild trauma* associated with contaminated vegetable matter, salt water diving, wind-blown contaminant and hot tub use. Trauma with organic matter and exposure to muddy water are the major predisposing factors in developing countries.

3. *Opportunistic infection.* Acanthamoeba keratitis can also occur as opportunistic infection in patients with herpetic keratitis, bacterial keratitis, bullous keratopathy and neuroparalytic keratitis.

Clinical features

Symptoms include very severe pain (out of proportion to the degree of inflammation), watering, photophobia, blepharospasm and blurred vision.

Signs

Acanthamoeba keratitis evolves over several months as a gradual worsening keratitis with periods of temporary remission. Presentation is markedly variable, making diagnosis difficult. Characteristic features are described below:

1. *Initial lesions* of acanthamoeba keratitis are in the form of limbitis, coarse, opaque streaks, fine epithelial and subepithelial opacities, and radial kerato-neuritis, in the form of infiltrates along corneal nerves.

2. *Advanced cases* show a central or paracentral ring-shaped lesion with stromal infiltrates and an overlying epithelial defect, ultimately presenting as ring abscess. Hypopyon may also be present.

Diagnosis

1. *Clinical diagnosis* is difficult and usually made by exclusion with strong clinical suspicion out of the non-responsive patients being treated for herpetic, bacterial or fungal keratitis.

2. *Laboratory diagnosis.* Corneal scrapings may be helpful in some cases as under:

– *potassium hydroxide (koh) mount* is reliable in experienced hands for recognition of acanthamoeba cysts;

– *calcofluor white stain* is a fluorescent brightener which stains the cysts of acanthamoeba bright apple green under fluorescence microscope;

– *lactophenol cotton blue stained* film is also useful for demonstration of acanthamoeba cysts in the corneal scrapings;

– *culture on non-nutrient agar* (*E. coli* enriched) may show trophozoites within 48 hours, which gradually turn into cysts.

Treatment is usually unsatisfactory.

1. *Non-specific treatment* is on the general lines for corneal ulcer.

2. *Specific medical treatment* includes: Brolene eye drops (0.1 % propamidine isethionate drops), neomycin drops, 0.01–0.02 % polyhexamethylene biguanide solution, and chlorhexidine. Other drugs that may be useful are paromomycin and various topical and oral imidazoles such as fluconazole, itraconazole and miconazole. Duration of medical treatment is very large (6 months to 1 year).

3. *Penetrating keratoplasty* is frequently required in non-responsive cases.

Allergic keratitis

1. Phlyctenule keratitis.
2. Vernal keratitis.
3. Atopic keratitis.

Trophic corneal ulcers

Trophic corneal ulcers develop due to disturbance in metabolic activity of epithelial cells. This group includes:

– neuroparalytic keratitis;
– exposure keratitis.

Neuroparalytic keratitis

Neuroparalytic keratitis occurs due to paralysis of the sensory nerve supply of the cornea.

Causes

I. *Congenital:* familial dysautonomia (Riley–Day syndrome), congenital insensitivity to pain, anhidrotic ectodermal dysplasia.

II. *Acquired:*

1. Following alcohol-block or electrocoagulation of Gasserian ganglion or section of the sensory root of trigeminal nerve for trigeminal neuralgia.

2. A neoplasm pressing on Gasserian ganglion.

3. Gasserian ganglion destruction due to acute infection in herpes zoster ophthalmicus.

4. Acute infection of Gasserian ganglion by herpes simplex virus.

5. Syphilitic (luetic) neuropathy.

6. Involvement of corneal nerves in leprosy.

7. Injury to Gasserian ganglion.

Pathogenesis

Exact pathogenesis is not clear; presumably, the disturbances in the antidromic corneal reflex occur due to the fifth nerve paralysis.

Clinical features

1. Characteristic features are no pain, no lacrimation, and complete loss of corneal sensations.

2. Ciliary congestion is marked.

3. Corneal sheen is dull.

4. Initial corneal changes are in the form of punctate epithelial erosions in the inter-palpebral area followed by ulceration due to exfoliation of corneal epithelium.

5. Relapses are very common, even the healed scar quickly breaks down again.

Treatment

1. Initial treatment with antibiotic and atropine eye ointment with patching is tried. Healing is usually very slow. Recently described treatment modality includes topical nerve growth factor drops and amniotic membrane transplantation.

2. If, however, relapses occur, it is best to perform lateral tarsorrhaphy which should be kept for at least one year. Along with it the prolonged use of artificial tears is also recommended.

Exposure keratitis

Normally cornea is covered by eyelids during sleep and is constantly kept moist by blinking movements during awaking. When eyes are covered insufficiently by the lids and there is loss of protective mechanism of blinking the condition of *exposure keratopathy* (*keratitis lagophthalmos*) develops.

Causes

The following factors which produce lagophthalmos may lead to exposure keratitis:

1. *Extreme proptosis* due to any cause will allow inadequate closure of lids.

2. *Bell's palsy* or any other cause of facial palsy.

3. *Ectropion* of severe degree.

4. *Symblepharon* causing lagophthalmos.

5. *Deep coma* associated with inadequate closure of lids.

6. *Physiological lagophthalmos.* Occasionally, lagophthalmos during sleep may occur in healthy individuals.

Pathogenesis

Due to exposure the corneal epithelium dries up followed by desiccation. After the epithelium is cast off, invasion by infective organisms may occur.

Clinical features

Initial desiccation occurs in the interpalpebral area leading to fine punctate epithelial keratitis which is followed by necrosis, frank ulceration and vascularization. Bacterial superinfection may cause deep suppurative ulceration which may even perforate.

Treatment

1. *Prophylaxis.* Once lagophthalmos is diagnosed, the following measures should be taken to prevent exposure keratitis:

– *frequent instillation of artificial tear eyedrops*;

– *instillation of ointment* and *closure of lids by a tape* or *bandage during sleep*;

– *soft bandage contact lens* with frequent instillation of *artificial tears* is required in cases of moderate exposure;

– *treatment of cause of exposure* – if the possible cause of exposure (proptosis, ectropion, etc.) should be treated.

2. *Treatment of corneal ulcer* is on the general lines.

3. *Tarsorrhaphy* is invariably required when it is not possible to treat the cause or when recovery of the cause (e. g., facial palsy) is not anticipated.

Keratitis Associated With Skin Diseases and Mucous Membrane

Rosacea keratitis

Corneal ulceration is seen in about 10 percent cases of acne rosacea, which is primarily a disease of the sebaceous glands of the skin.

Clinical features

1. The condition typically occurs in elderly women in the form of *facial eruptions* presenting as butterfly configuration, predominantly involving the malar and nasal area of face.

2. *Ocular lesions* include chronic blepharoconjunctivitis and keratitis. Rosacea keratitis occurs as yellowish white marginal infiltrates, and small ulcers that progressively advance across the cornea and almost always become heavily vascularized.

Treatment

1. *Local treatment.* Rosacea keratitis responds to topical steroids, but recurrences are very common.

2. *Systemic treatment.* The essential and most effective treatment of rosacea keratitis is a long course of systemic 250 mg tetracycline qid for 3 weeks.

Metronidazole exhibits antimicrobial (antibacterial and antiparasitic), anti-inflammatory, and immunosuppressive properties and has been found to be effective against rosacea. In fact, oral metronidazole has been advocated as first-line therapy.

Corneal Ulcer Associated With Systemic Collagen Vascular Diseases

Peripheral corneal ulceration and/or melting of corneal tissue are not infrequent occurrence in patients suffering from systemic diseases such as rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa and Wegener's granulomatosis.

Such corneal ulcers are usually indolent and difficult to treat. Systemic treatment of the primary disease may be beneficial.

Idiopathic Corneal Ulcers

Mooren's ulcer

The *Moorhen's ulcer* (*chronic serpiginous or rodent ulcer*) is a severe inflammatory peripheral ulcerative keratitis.

Etiology

Exact etiology is not known. Different views are:

1. It is an idiopathic degenerative condition.

2. It may be due to an ischemic necrosis resulting from vasculitis of limbal vessels.

3. It may be due to the effects of enzyme collagenase and proteoglycanase produced from conjunctiva.

4. Most probably it is *an autoimmune disease* (antibodies against corneal epithelium have been demonstrated in serum).

Clinical picture

Two clinical varieties of Moorhen's ulcer have been recognized.

1. *Benign form*, which is usually unilateral, affects the elderly people and is characterised by a relative slow progress.

2. *Virulent type* also called the *progressive form* is bilateral, more often occurs in younger patients.

The ulcer is rapidly progressive with a high incidence of scleral involvement.

Symptoms

These include severe pain, photophobia, lacrimation and defective vision.

Treatment

Since exact etiology is still unknown, its treatment is highly unsatisfactory. Following measures may be tried:

1. *Topical corticosteroids* instilled every 2–3 hours are tried as initial therapy with limited success.

2. *Immunosuppressive therapy* with systemic steroids may be of help. Immunosuppression with *cyclosporin* or other cytotoxic agents may be quite useful in virulent type of disease.

3. *Soft contact lenses* have also been used with some relief in pain.

4. *Lamellar or full thickness corneal grafts* often melt or vascularize.

Non-Ulcerative Keratitis

Non-ulcerative keratitis can be divided into two groups:

– non-ulcerative superficial keratitis;
– non-ulcerative deep keratitis.

Non-ulcerative superficial keratitis

This group includes a number of conditions of varied etiology. Here the inflammatory reaction is confined to epithelium, Bowman's membrane and superficial stromal lamellae. Non-ulcerative superficial keratitis may present in two forms:

– diffuse superficial keratitis;
– superficial punctate keratitis.

I. *Diffuse superficial keratitis*

Diffuse inflammation of superficial layers of cornea occurs in two forms, acute and chronic.

1. *Acute diffuse superficial keratitis*

Etiology

Mostly of infective origin, it may be associated with staphylococcal or gonococcal infections.

Clinical features

It is characterised by faint diffuse epithelial edema associated with grey farinaceous appearance being interspersed with relatively clear area. Epithelial erosions may be formed at places. If uncontrolled, it usually converts into ulcerative keratitis.

Treatment

It consists of frequent instillation of antibiotic eyedrops such as tobramycin or gentamycin every 2–4 hours.

2. Chronic diffuse superficial keratitis

It may be seen in rosacea, phlyctenules and is typically associated with pannus formation.

II. *Superficial punctate keratitis (spk)*

Superficial punctate keratitis is characterised by occurrence of multiple, spotty lesions in the superficial layers of cornea. It may result from a number of conditions, identification of which (causative condition) might not be possible most of the times.

Some important ***causes*** of superficial punctate keratitis are listed here:

1. *Viral infections* are the chief cause. More common are: herpes zoster, adenovirus infections, epidemic keratoconjunctivitis, pharyngo-conjunctival fever and herpes simplex.

2. *Chlamydial infections* include trachoma and inclusion conjunctivitis.

3. *Toxic lesions*, e. g., due to staphylococcal toxin in association with blepharoconjunctivitis.

4. *Trophic lesions*, e. g., exposure keratitis and neuroparalytic keratitis.

5. *Allergic lesions*, e. g., vernal keratoconjunctivitis.

6. *Irritative lesions*, e. g., effect of some drugs such as idoxuridine.

7. *Disorders of skin and mucous membrane,* such as acne rosacea and pemphigoid.

8. *Dry eye syndrome*, i. e., keratoconjunctivitis sicca.

9. *Specific type of idiopathic SPK*, e. g., Thygeson's superficial punctate keratitis and Theodore's superior limbic keratoconjunctivitis.

10. *Photo-ophthalmia*.

Morphological types

– punctate epithelial erosions (multiple superficial erosions);

– punctate epithelial keratitis;

– punctate subepithelial keratitis;

– punctate combined epithelial and subepithelial keratitis;

– filamentary keratitis.

Clinical features

Superficial punctate keratitis may present as different morphological types as enumerated above. Punctate epithelial lesions usually stain with fluorescein, rose Bengal and other vital dyes. The condition mostly presents acutely with pain, photophobia and lacrimation; and is usually associated with conjunctivitis.

Treatment

Treatment of most of these conditions is symptomatic.

1. *Topical steroids* have a marked suppressive effect.

2. *Artificial tears* have soothing effect.

3. *Specific treatment* of cause should be instituted whenever possible, e. g., antiviral drugs in cases of herpes simplex.

Photo-ophthalmia

Photo-ophthalmia refers to occurrence of multiple epithelial erosions due to the effect of ultraviolet rays especially from 311 to 290μ.

Causes

1. Exposure to bright light of a short circuit.

2. Exposure to a naked arc light as in industrial welding and cinema operators.

3. *Snow blindness* due to reflected ultraviolet rays from snow surface.

Pathogenesis

After an interval of 4–5 hours (latent period) of exposure to ultraviolet rays there occurs desquamation of corneal epithelium leading to formation of multiple epithelial erosions.

Clinical features:

– typically, patient presents with severe burning pain, lacrimation, photophobia, blepharospasm, swelling of palpebral conjunctiva and retrotarsal folds;

– there is history of exposure to ultraviolet rays 4–5 hours earlier;

– on fluorescein staining multiple spots are demonstrated on both corneas.

Prophylaxis

Croker's glass which cuts off all infrared and ultraviolet rays should be used by those who are prone to exposure, e. g., welding workers, cinema operators, etc.

Treatment

1. Cold compresses.
2. Pad and bandage with antibiotic ointment for 24 hours, heals most of the cases.
3. Oral analgesics may be given if pain is intolerable.
4. Single dose of tranquillizer may be given to apprehensive patients.

Superior Limbic Keratoconjunctivitis

Superior limbic keratoconjunctivitis of Theodore is the name given to inflammation of superior limbic, bulbar and tarsal conjunctiva associated with punctate keratitis of the superior part of cornea.

Etiology

Exact etiology is not known. It occurs with greater frequency in patients with hyperthyroidism and is more common in females.

Clinical course

It has a chronic course with remissions and exacerbations.

Symptoms include:

– bilateral ocular irritation;

– mild photophobia and redness in superior bulbar conjunctiva.

Signs include:

– *congestion* of superior limbic, bulbar and tarsal conjunctiva;

– *punctate keratitis* which stains with fluorescein and rose Bengal stain is seen in superior part of cornea;

– *corneal filaments* are also frequently seen in the involved area.

Treatment

1. Topical artificial tears.

2. Low doses of topical corticosteroids may reduce the symptoms temporarily.

3. Faint diathermy of superior bulbar conjunctiva in a checker board pattern gives acceptable results.

4. Recession or resection of a 3–4 mm wide perilimbal strip of conjunctiva from the superior limbus (from 10.30 to 1.30 o'clock position) may be helpful if other measures fail.

5. Therapeutic soft contact lenses for a longer period may be helpful in healing the keratitis.

Thygeson's Superficial Punctate Keratitis

It is a type of chronic, recurrent bilateral superficial punctate keratitis, which has got a specific clinical identity.

Etiology

Exact etiology is not known:

– a *viral origin* has been suggested without any conclusion;

– an *allergic* or *dyskeratotic nature* also has been suggested owing to its response to steroids.

Clinical features:

– *age and sex* – it may involve all ages with no sex predilection;

– *laterality* – usually bilateral;

– *course* – it is a chronic disease characterized by remissions and exacerbations.

Symptoms

It may be asymptomatic, but is usually associated with foreign body sensation, photophobia and lacrimation.

Signs

1. *Conjunctiva* is uninflamed (no conjunctivitis).

2. *Corneal lesions.* There are coarse punctate epithelial lesions (snow flake) circular, oval or stellate in shape, slightly elevated and situated in the central part (pupillary area) of cornea.

Treatment

1. The disease is self-limiting with remissions and may permanently disappear in the period of 5–6 years.

2. During exacerbations the lesions and associated symptoms usually respond quickly to topical steroids (so, should be tapered rapidly).

3. Therapeutic soft contact lenses may be required in steroid-resistant cases.

Deep Keratitis

An inflammation of corneal stroma with or without involvement of posterior corneal layers constitutes deep keratitis, which may be non-suppurative or suppurative.

Non-suppurative deep keratitis includes interstitial keratitis, disciform keratitis, keratitis profunda and sclerosing keratitis.

Suppurative deep keratitis includes central corneal abscess and posterior corneal abscess, which are usually metastatic in nature.

Interstitial keratitis

Interstitial keratitis is an inflammation of the corneal stroma without primary involvement of the epithelium or endothelium.

Its common ***causes*** are: congenital syphilis, tuberculosis, Cogan's syndrome, acquired syphilis, trypanosomiasis, malaria, leprosy, and sarcoidosis.

Syphilitic (luetic) interstitial keratitis

Syphilitic interstitial keratitis is associated more frequently (90 percent) with congenital syphilis than the acquired syphilis. The disease is generally bilateral in inherited syphilis and unilateral in acquired syphilis. In congenital syphilis, manifestations develop between 5–15 years of age.

Clinical features

Interstitial keratitis characteristically forms one of the late manifestations of congenital syphilis. Sometimes it may be a part of *Hutchinson's triad* which includes: interstitial keratitis, Hutchinson's teeth and vestibular deafness.

The clinical picture of interstitial keratitis can be divided into three stages: *initial progressive stage*, *florid stage* and *stage of regression.*

1. *Initial progressive stage.* The disease begins with edema of the endothelium and deeper stroma, secondary to anterior uveitis, as evidenced by the presence of keratic precipitates (KPs). There is associated pain, lacrimation, photophobia, blepharospasm and circumcorneal injection followed by a diffuse corneal haze giving it a *ground glass appearance.* This stage lasts for about 2 weeks.

2. *Florid stage.* In this stage eye remains acutely inflamed. Deep vascularization of cornea, consisting of radial bundle of brush-like vessels develops. Since these vessels are covered by hazy cornea, they look dull reddish pink which is called "*salmon patch appearance*". There is often a moderate degree of superficial vascularization. These vessels arising from the terminal arches of conjunctival vessels run a short distance over the cornea. These vessels and conjunctiva heap at the limbus in the form of *epulit.* This stage lasts for about 2 months.

3. *Stage of regression.* The acute inflammation resolves with the progressive appearance of vascular invasion. Clearing of cornea is slow and begins from periphery and advances centrally. Resolution of the lesion leaves behind some opacities and *ghost vessels.* This stage may last for about 1 to 2 years.

Diagnosis is usually evident from the clinical profile. A positive VDRL or *Treponema pallidum* immobilization test confirms the diagnosis.

Treatment should include *topical treatment for keratitis* and *systemic treatment for syphilis*.

1. *Local treatment:*

– *topical corticosteroid drops*, e. g., dexamethasone 0.1 % drops every 2–3 hours. As the condition is allergic in origin, corneal clearing occurs with steroids if started well in time and a useful vision is obtained;

– *atropine eye ointment:* 1 % of ointment 2–3 times a day;

– *dark goggles* are to be used for photophobia;

– *keratoplasty* is required in cases where dense corneal opacities are left.

2. *Systemic treatment*

Penicillin in high doses should be started to prevent development of further syphilitic lesions. However, an early treatment of congenital syphilis usually does not prevent the onset of keratitis at a later stage.

Systemic steroids may be added in refractory cases of keratitis.

Tuberculous interstitial keratitis

The features of tubercular interstitial keratitis are similar to syphilitic interstitial keratitis except that it is more frequently unilateral and sectorial (usually involving a lower sector of cornea).

Treatment consists of systemic antitubercular drugs, topical steroids and cycloplegics.

Cogan's syndrome

This syndrome comprises of the interstitial keratitis of unknown etiology, acute tinnitis, vertigo, and deafness. It typically occurs in middle-aged adults and is often bilateral.

Treatment consists of topical and systemic *corticosteroids.* An early treatment usually prevents permanent deafness and blindness.

Corneal Degenerations

Corneal degenerations refer to the conditions in which the normal cells undergo some degenerative changes under the influence of age or some pathological condition.

Classification

Depending upon location:

I. *Axial corneal degenerations:*

1. Fatty degeneration.
2. Hyaline degeneration.
3. Amyloidosis.
4. Calcific degeneration (Band keratopathy).
5. Salzmann's nodular degeneration.

II. *Peripheral degenerations:*

1. Arcus senilis.
2. Vogt's white limbal girdle.
3. Hassall–Henle bodies.
4. Terrine's marginal degeneration.
5. Mooren's ulcer.
6. Pellucid marginal degeneration.
7. Furrow degeneration (senile marginal degeneration).

Depending upon etiology:

I. *Age related degenerations* (Arcus senilis, Vogt's white limbal girdle, Hassall–Henle bodies, Mosaic degeneration).

II. *Pathological degenerations* (fatty degeneration, amyloidosis, calcific degeneration, Salzmann's nodular

degeneration, Furrow degeneration, spheroidal degeneration, pellucid marginal degeneration, Terrine's marginal degeneration, Mooren's ulcer).

Congenital Anomalies

Megalocornea

Horizontal diameter of cornea at birth is about 10 mm and the adult size of about 11.7 mm is attained by the age of 2 years. Megalocornea is labelled when the horizontal diameter of cornea is of adult size at birth or 13 mm or greater after the age of 2 years. The cornea is usually clear with normal thickness and vision. The condition is not progressive. Systemic association includes Marfan's, Apert, Ehlers Danlos and Down syndromes.

Differential diagnosis

1. *Buphthalmos.* In this condition IOP is raised and the eyeball is enlarged as a whole. The enlarged cornea is usually associated with central or peripheral clouding and Descemet's tears (Haab's striae).

2. *Keratoglobus.* In this condition, there is thinning and excessive protrusion of cornea, which seems enlarged; but its diameter is usually normal.

Microcornea

In microcornea, the horizontal diameter is less than 10 mm since birth. The condition may occur as an isolated anomaly (rarely) or in association with *non ophthalmic* (normal small eyeball) or *microphthalmos* (abnormal small eyeball).

Cornea plana

This is a rare anomaly in which bilaterally cornea is comparatively flat since birth. It may be associated with microcornea. Cornea plana usually results in marked astigmatic refractive error.

Congenital cloudy cornea

The acronym STUMPED helps to remember the common conditions to be included in differential diagnosis of neonatal cloudy cornea. The conditions are as follows: **s**clerocornea, **t**ears in Descemet's membrane, **u**lcer, **m**etabolic conditions, **p**osterior corneal defect, **e**ndothelial dystrophy, **d**ermoid.

Abnormalities of Corneal Transparency

Normal cornea is a transparent structure. Any condition which upsets its anatomy or physiology causes loss of its transparency to some degree.

Common causes of loss of corneal transparency are:

- corneal edema;
- drying of cornea;
- depositions on cornea;
- inflammations of cornea;
- corneal degenerations;
- dystrophies of cornea;
- vascularization of cornea;
- scarring of cornea (corneal opacities).

TOPIC 9. DISEASES OF THE UVEAL TRACT

Congenital Anomalies of the Uveal Tract

Heterochromia of iris

It refers to variations in the iris colour and is a common congenital anomaly. In *heterochromia iridum*, the colour of one iris differs from the other. Sometimes, one sector of the iris may differ from the remainder of iris; such a condition is called segmental *heterochromia iridis.* Congenital heterochromia must be differentiated from the acquired heterochromia seen in heterochromic cyclitis, siderosis and malignant melanoma of iris.

Corectopia

It refers to abnormally eccentric placed pupil. Normally the pupil is placed slightly nasal to the centre.

Polycoria

In this condition, there is more than one pupil in one or both eyes.

Congenital aniridia (iridremia)

It refers to the congenital absence of the iris. *True aniridia*, i. e. complete absence of the iris, is extremely rare. Usually, a peripheral rim of iris is present and this condition is called "*clinical aniridia"*. Zonules of the lens and ciliary processes are often visible. The condition is usually genetical and may be associated with glaucoma due to angle anomalies.

Persistent pupillary membrane

It represents the remnants of the vascular sheath of the lens. It is characterised by stellate-shaped shreds of the pigmented tissue coming from anterior surface of the iris (attached at collarette). These float freely in the anterior chamber or may be attached to the anterior surface of the lens.

Congenital Coloboma of the Uveal Tract

Congenital coloboma (absence of tissue) of iris, ciliary body and choroid may be seen in association or independently. Coloboma may be typical or atypical.

Typical coloboma is seen in the inferionasal quadrant and occurs due to defective closure of the embryonic fissure.

Atypical coloboma is occasionally found in other positions.

Complete coloboma extends from the pupil to the optic nerve, with a sector-shaped gap occupying about one-eighth of the circumference of the retina, choroid, ciliary body, iris thus causing a corresponding indentation of the lens where the zonular fibers are missing.

Uveitis

The term *uveitis* strictly means inflammation of the uveal tissue only. However, practically there is always some associated inflammation of the adjacent structures such as retina, vitreous, sclera and cornea.

Due to close relationship of the anatomically distinct parts of the uveal tract, the inflammatory process usually tends to involve the uvea as a whole.

Classification

I. Anatomical classification:

1. *Anterior uveitis.* It is the inflammation of the uveal tissue from iris up to pars plicata (folded portion) of ciliary body. It may be subdivided into:

– *iritis* in which inflammation predominantly affects the iris;

– *iridocyctitis* in which iris and pars plicata of the ciliary body are equally involved;

– *cyclitis* in which pars plicata of the ciliary body is predominantly affected.

2. *Intermediate uveitis.* It includes inflammation of the pars plana and peripheral part of the retina and underlying choroid. It is also called "*pars planitis*".

3. *Posterior uveitis.* It refers to the inflammation of the choroid (choroiditis). There is usually an associated inflammation of the retina hence the term "*chorioretinitis*" is used.

4. *Panuveitis.* It is the inflammation of the whole uvea.

II. Clinical classification:

1. *Acute uveitis.* It has a sudden symptomatic onset and the disease lasts for about six weeks to 3 months.

2. *Chronic uveitis.* It frequently has an insidious and asymptomatic onset. It persists longer than 3 months to even years and is usually diagnosed when it causes defective vision.

III. Pathological classification

1. *Suppurative or purulent uveitis*.

2. *Non-suppurative uveitis*. It has been further subdivided in two groups (Wood's classification):

– non-granulomatous uveitis;
– granulomatous uveitis.

IV. Etiological (Duke Elder's) classification:

1. *Infective uveitis*.
2. *Allergic uveitis*.
3. *Toxic uveitis*.
4. *Traumatic uveitis*.
5. *Uveitis associated with non-infective systemic diseases*.
6. *Idiopathic uveitis*.

Etiology of uveitis

Despite a great deal of experimental research and many sophisticated methods of investigations, etiology and immunology of uveitis is still largely not understood. Even today, the cause of many clinical conditions is disputed (remains presumptive) and in many others etiology is

unknown. The etiological concepts of uveitis as proposed by *Duke Elder*, in general, are discussed here.

1. *Infective uveitis*

Here, inflammation of the uveal tissue is induced by invasion of microorganisms. Uveal infections may be exogenous, secondary or endogenous.

Exogenous infection occurs wherein the infecting organisms directly gain entrance into the eye from outside. It can occur following penetrating injuries, perforation of corneal ulcer and postoperatively (after intraocular operations).

Secondary infection of the uvea occurs by the spread of infection from neighbouring structures, e. g., acute purulent conjunctivitis, (pneumococcal and gonococcal), keratitis, scleritis, retinitis, orbital cellulitis and orbital thrombophlebitis.

Endogenous infections are caused by the entrance of organisms from some source situated elsewhere in the body, by way of the bloodstream.

Endogenous infections play important roles in the inflammations of uvea.

Types of infective uveitis

Depending upon the causative organisms, infective uveitis may be classified as follows:

– *bacterial infections.* These may be granulomatous, e. g., tubercular, leprotic, syphilitic, brucellosis or pyogenic such as streptococci, staphylococci, pneumococci and gonococcus;

– *viral infections* associated with uveitis are herpes simplex, herpes zoster and cytomegalo inclusion virus (CMV);

– *fungal uveitis* is rare and may accompany systemic aspergillosis, candidiasis and blastomycosis. It also includes presumed ocular histoplasmosis syndrome;

– *parasitic uveitis* is seen in toxoplasmosis, toxocariasis, onchocerciasis and amoebiasis;

– *rickettsial uveitis* may occur in scrub typhus and epidemic typhus.

2. *Allergic (hypersensitivity linked) uveitis*

Allergic uveitis is one of the commonest occurrences in clinical practice. The complex subject of hypersensitivity linked inflammation of uveal tissue is still not clearly understood. It may be caused by the following ways:

Microbial allergy

In this type, primary source of infection is somewhere else in the body and the escape of the organisms or their products into the bloodstream causes sensitization of the uveal tissue with formation of antibodies. Later, a renewal of infection in the original focus may again cause dissemination of the organisms or their products (antigens), which on meeting the sensitized uveal tissue excite an allergic inflammatory response.

Primary focus of infection can be a minute tubercular lesion in the lymph nodes or lungs. Once, it used to be the most common cause of uveitis worldwide, but now it is rare. However, in developing countries like India or Ukraine tubercular infections still play an important role. Other sources of primary focus are streptococcus and other infections in the teeth, paranasal sinuses, tonsils, prostate, genitals and urinary tract.

Anaphylactic uveitis

It is said to accompany systemic anaphylactic reactions like serum sickness and angioneurotic edema.

Atopic uveitis

It occurs due to airborne allergens and inhalants, e. g., seasonal iritis due to pollens. A similar reaction to such materials as danders of cats, chicken feathers, house dust, egg albumin and beef proteins has also been noted.

Autoimmune uveitis is found in association with autoimmune disorders such as Still's disease, rheumatoid

arthritis, Wegener's granulomatosis, systemic lupus erythematosus, Reiter's disease and so on.

In phacoanaphylactic endophthalmitis, lens proteins play the role of autoantigens. Similarly, sympathetic ophthalmitis has been attributed to be an autoimmune reaction to uveal pigments, by some workers.

HLA-associated uveitis

Human leucocytic antigens (HLA) is the old name for the histocompatibility antigens. There are about 70 such antigens in human beings, on the basis of which an individual can be assigned to different HLA phenotypes. Recently, lot of emphasis is being laid on the role of HLA in uveitis, since a number of diseases associated with uveitis occur much more frequently in persons with certain specific HLA phenotypes.

A few examples of HLA-associated diseases with uveitis are as follows:

– HLA–B27: acute anterior uveitis associated with ankylosing spondylitis and also in Reiter's syndrome;

– HLA–B5: uveitis in Behcet's disease;

– HLA–DR4 and DW15: Vogt Koyanagi Harada's disease.

3. *Toxic uveitis*

Toxins responsible for uveitis can be endotoxins, endocular toxins or exogenous toxins.

Endotoxins, produced inside the body play a major role. These may be autotoxins or microbial toxins (produced by organisms involving the body tissues). Toxic uveitis seen in patients with acute pneumococcal or gonococcal conjunctivitis and in patients with fungal corneal ulcer is thought to be due to microbial toxins.

Endocular toxins are produced from the ocular tissues. Uveitis seen in patients with blind eyes, long-standing retinal detachment and intraocular haemorrhages is said to be due to endocular toxins.

Other examples are uveitis associated with intraocular tumours and phacotoxic uveitis.

Exogenous toxins causing uveitis are irritant chemical substances of inorganic, animal or vegetative origin. Certain drugs producing uveitis (such as miotics and cytotoxic drugs) are other examples of exogenous toxins.

4. *Traumatic uveitis*

It is often seen in accidental or operative injuries to the uveal tissue. Different mechanisms which may produce uveitis following trauma include:

– direct mechanical effects of trauma;

– irritative effects of blood products after intraocular haemorrhage (haemophthalmitis);

– microbial invasion;

– chemical effects of retained intraocular foreign bodies;

– sympathetic ophthalmia in the other eye.

5. *Uveitis associated with non-infective systemic diseases*

Certain systemic diseases frequently complicated by uveitis include: sarocoidosis, collagen related diseases (polyarteritis nodosa (PAN), disseminated lupus erythematosus (DLE), rheumatic and rheumatoid arthritis), metabolic diseases (diabetes mellitus and gout), disease of the central nervous system (e. g., disseminated sclerosis) and diseases of skin (psoriasis, lichen planus, erythema nodosum, pemphigus and so on).

6. *Idiopathic uveitis*

It may be specific or nonspecific:

– *specific idiopathic uveitis entities* include the conditions which have certain special characteristics of their own, e. g., pars planitis, sympathetic ophthalmitis and Fuchs' hetero-chromic iridocyclitis;

– *nonspecific idiopathic uveitis entities* include the conditions which do not belong to any of the known etiological groups. About more than 25 percent cases of uveitis fall in this group.

Pathology of Uveitis

Inflammation of the uvea fundamentally has the same characteristics as any other tissue of the body, i. e, a vascular and a cellular response. However, due to extreme vascularity and looseness of the uveal tissue, the inflammatory responses are exaggerated and thus produce special results.

Pathologically, inflammations of the uveal tract may be divided into suppurative (purulent) and nonsuppurative (non-purulent) varieties. Wood has further classified non-suppurative uveitis into non-granulomatous and granulomatous types. Although morphologic description is still of some value, the rigid division of uveitis by Wood into these two categories has been questioned on both clinical and pathological grounds. Certain transitional forms of uveitis have also been recognized. Some of these (e. g., phacoanaphylactic endophthalmitis and sympathetic ophthalmia) showing pathological features of granulomatous uveitis are caused by hypersensitivity reactions. While uveitis due to tissue invasion by leptospira presents the manifestation of non-granulomatous uveitis.

Nonetheless, the classification is often useful in getting oriented towards the subject of uveitis, its workup and therapy. Therefore, it is worthwhile to describe the pathological features of these overlapping (both clinically and pathologically) conditions as distinct varieties.

1. *Pathology of suppurative uveitis*

Purulent inflammation of the uvea is usually a part of endophthalmitis or panophthalmitis occurring as a result of exogenous infection by pyogenic organisms which include staphylococcus, streptococcus, psuedomonas, pneumococcus

and gonococcus. The pathological reaction is characterised by an outpouring of purulent exudate and infiltration by polymorphonuclear cells of uveal tissue, anterior chamber, posterior chamber and vitreous cavity. As a result, the whole uveal tissue is thickened and necrotic, and the cavities of eye become filled with pus.

2. *Pathology of non-granulomatous uveitis*

The pathological alterations of the non-granulomatous reaction consist of marked dilation and increased permeability of vessels, breakdown of blood aqueous barrier with an outpouring of fibrinous exudate and infiltration by lymphocytes, plasma cells and large macrophages of the uveal tissue, anterior chamber, posterior chamber and vitreous cavity. The inflammation is usually diffuse.

As a result of these pathological reactions iris becomes waterlogged, edematous, muddy with blurring of crypts and furrows. As a consequence its mobility is reduced, pupil becomes small in size due to sphincter irritation and engorgement of radial vessels of iris. Exudates and lymphocytes poured into the anterior chamber result in aqueous flare and deposition of fine KPs at the back of cornea. Due to exudates in the posterior chamber, the posterior surface of iris adheres to the anterior capsule of lens leading to posterior synechiae formation. In severe inflammation, due to pouring of exudate from ciliary processes behind the lens, an exudative membrane called *cyclitic membrane* may be formed.

3. *Pathology of granulomatous uveitis*

Granulomatous uveitis is a chronic inflammation of proliferative nature. The common organisms which excite this type of inflammation are those responsible for tuberculosis, leprosy, syphilis, brucellosis, leptospirosis, sarcoidosis, sympathetic ophthalmitis and Vogt–Koyanagi–Harada disease.

The pathological reaction in granulomatous uveitis is characterized by infiltration with lymphocytes and plasma

cells, with the mobilization and proliferation of large mononuclear cells which eventually become epithelioid giant cells and aggregate into nodules.

Iris nodules are usually formed near pupillary border (*Koeppe's nodules*). Similar nodular collection of the cells is deposited at the back of cornea in the form of mutton fat keratic precipitates and aqueous flare is minimal. Necrosis in the adjacent structures leads to a reparative process resulting in fibrosis and gliosis of the involved area.

Anterior Uveitis (Iridocyclitis)

Clinical features

Although anterior uveitis almost always presents as a combined inflammation of iris and ciliary body (iridocyclitis), the reaction may be more marked in iris (iritis) or the ciliary body (cyclitis). Clinically it may present as acute or chronic anterior uveitis. Main symptoms of *acute anterior uveitis* are pain, photophobia, redness, lacrimation and decreased vision. In *chronic uveitis*, however, the eye may be white with minimal symptoms even in the presence of signs of severe inflammation.

Symptoms

1. Pain.
2. Redness.
3. Photophobia and blepharospasm.
4. Lacrimation.
5. Defective vision.

Signs

Slitlamp biomicroscopic examination is essential to elicit most of the signs of uveitis.

I. *Lid edema*, usually mild, may accompany a severe attack of acute anterior uveitis.

II. *Circumcorneal congestion* is marked in acute iridocyclitis and minimal in chronic iridocyclitis. It must be

differentiated from superficial congestion occurring in acute conjunctivitis.

III. *Corneal signs* include: corneal edema, KPs and posterior corneal opacities.

1. *Corneal edema* is due to toxic endothelitis and raised intraocular pressure when present.

2. *Keratic precipitates* (*KPs*) are proteinous cellular deposits occurring at the back of cornea.

Mostly, these are arranged in a triangular fashion occupying the centre and inferior part of cornea due to convection currents in the aqueous humor. The composition and morphology of KPs varies with the severity, duration and type of uveitis.

The following *types of KPs* may be seen:

Mutton fat KPs typically occur in granulomatous iridocyclitis and are composed of epithelioid cells and macrophages. They are large, thick, fluffy, lardaceous KPs, having a greasy or waxy appearance. Mutton fat KPs are usually a few (10 to 15) in number.

Small and medium KPs (*granular KPs*) are pathognomic of non-granulomatous uveitis and are composed of lymphocytes. These small, discrete, dirty white KPs are arranged irregularly at the back of cornea. Small KPs may be hundreds in number and form the so called *endothelial dusting*.

Red KPs are formed when in addition to inflammatory cells; RBCs also take part in composition. They may be seen in hemorrhagic uveitis.

Old KPs are signs of healed uveitis. Either of the above described KPs, as a result of the healing process shrink, fade, become pigmented and irregular in shape (crenated margins). Old mutton fat KPs usually have a ground glass appearance due to hyalinization.

3. *Posterior corneal opacity* may be formed in longstanding cases of iridocyclitis.

IV. *Anterior chamber signs*

1. *Aqueous cells.* It is an early feature of iridocyclitis. The cells should be counted in an oblique slit-lamp beam, 3 mm long and 1mm wide, with maximal light intensity and magnification, and graded as:

= 0 cells;
± = 1–5 cells;
+1 = 6–10 cells;
+2 = 11–20 cells;
+3 = 21–50 cells;
+4 = over 50 cells.

2. *Aqueous flare.* It is due to leakage of protein particles into the aqueous humor from damaged blood vessels. It is demonstrated on the slitlamp examination by a point beam of light passed obliquely to the plane of the iris. In the beam of light, protein particles are seen as suspended and moving dust particles. This is based on the "Brownian movements" or "Tyndal phenomenon". Aqueous flare is usually marked in non-granulomatous and minimal in granulomatous uveitis. The flare is graded from 0 to +4. The grade is as follows:

0 = no aqueous flare;
+1 = just detectable;
+2 = moderate flare with clear iris details;
+3 = marked flare (iris details not clear);
+4 = intense flare (fixed coagulated aqueous with considerable fibrin).

3. *Hypopyon.* When exudates are heavy and thick, they settle down in lower part of the anterior chamber as hypopyon (sterile pus in the anterior chamber).

4. *Hyphaema* (blood in the *anterior chamber*): it may be seen in hemorrhagic type of uveitis.

Koeppe's nodules are situated at the pupillary border and may initiate posterior synechia.

Busacca's nodules situated near the collarette are large but less common than the Koeppe's nodules.

5. *Posterior synechiae.* These are adhesions between the posterior surface of iris and anterior capsule of crystalline lens or any other structure which may be artificial lens, cataract, posterior capsule (left after extracapsular cataract extraction) or anterior hyaloid face. These are formed due to organization of the fibrin-rich exudates. Morphologically, posterior synechiae may be *segmental*, *annular* or *total*.

Segmental posterior synechiae refers to adhesions of iris to the lens at some points.

Annular posterior synechiae (ring synechiae are 360º adhesions of pupillary margin to anterior capsule of lens. These prevent the circulation of aqueous humor from posterior chamber to anterior chamber (*seclusio pupillae*). Thus, the aqueous humor collects behind the iris and pushes it anteriorly (leading to "*iris bombe*" formation). This is usually followed by a rise in intraocular pressure.

Total posterior synechiae occurring due to plastering of total posterior surface of iris with the anterior capsule of lens are rarely formed in acute plastic type of uveitis. They result in deepening of anterior chamber.

6. *Neovascularsation of iris* (*rubeosis iridis*) develops in some eyes with chronic iridocyclitis.

7. *Changes in depth and shape of anterior chamber* may occur due to synechiae formation.

8. *Changes in the angle of anterior chamber* are observed with gonioscopic examination. In active stage, cellular deposits are seen, and in chronic stage peripheral anterior synechiae may be seen.

Iris signs

1. *Loss of normal pattern.* It occurs due to edema and waterlogging of iris in active phase and due to atrophic changes

in chronic phase. Iris atrophy is typically observed in Fuchs' heterochromic iridocyclitis.

2. *Changes in iris colour.* Iris usually becomes muddy in colour during active phase and may show hyper pigmented and depigmented areas in healed stage.

3. *Iris nodules*. These occur typically in granulomatous uveitis.

4. *Pupillary reaction* becomes sluggish or may even be absent due to edema and hyperemia of iris which hamper its movements.

5. *Occlusion pupillae* results when the pupil is completely occluded due to organization of the exudates across the entire pupillary area.

V. *Changes in the lens*

1. *Pigment dispersal* on the anterior capsule of lens is almost of universal occurrence in a case of anterior uveitis.

2. *Exudates* may be deposited on the lens in cases with acute plastic iridocyclitis.

3. *Complicated cataract* may develop as a complication of persistent iridocyclitis. Typical features of a complicated cataract in early stage are *polychromatic luster* and *bread-crumb* appearance of the early posterior subcapsular opacities. In the presence of posterior synechiae, the complicated cataract progresses rapidly to maturity.

VI. *Change in the vitreous*

Anterior vitreous may show exudates and inflammatory cells after an attack of acute iridocyclitis.

Complications and sequelae:

1. *Complicated cataract.* It is a common complication of iridocyclitis as described above.

2. *Secondary glaucoma.* It may occur as an early or late complication of iridocyclitis.

Early glaucoma: in active phase of the disease, presence of exudates and inflammatory cells in the anterior

chamber may cause clogging of trabecular meshwork resulting in the decreased aqueous drainage and thus a rise in intraocular pressure (*hypertensive uveitis*).

Late glaucoma in iridocyclitis (post inflammatory glaucoma) is the result of pupil block (seclusio pupillae due to ring synechiae formation, or occlusio pupillae due to organized exudates) not allowing the aqueous humor to flow from posterior to anterior chamber. There may or may not be associated peripheral anterior synechiae formation.

3. *Cyclitic membrane.* It results due to fibrosis of exudates present behind the lens. It is a late complication of acute plastic type of iridocyclitis.

4. *Choroiditis.* It may develop in prolonged cases of iridocyclitis owing to their continuity.

VII. *Pupillary signs*

1. *Narrow pupil.* It occurs in acute attack of iridocyclitis due to irritation of sphincter pupillae by toxins. Iris edema and engorged radial vessels of iris also contribute in making the pupil narrow.

2. *Irregular pupil shape.* It results from segmental posterior synechiae formation. Dilation of pupil with atropine at this stage results in *festooned pupil*.

3. *Ectropion pupillae* (evertion of pupillary margin). It may develop due to contraction of fibrinous exudate on the anterior surface of the iris.

4. *Retinal complications.* These include cystoid macular edema, macular degeneration, exudative retinal detachment and secondary periphlebitis retinae.

5. *Papillitis* (inflammation of the optic disc). It may be associated with severe cases of iridocyclitis.

6. *Band-shaped keratopathy.* It occurs as a complication of long-standing chronic uveitis, especially in children having Still's disease.

7. *Phthisis bulbi.* It is the final stage end result of any form of chronic uveitis. In this condition, ciliary body is disorganised and so aqueous production is hampered. As a result of it the eye becomes soft, shrinks and eventually becomes a small atrophic globe (phthisis bulbi).

Differential diagnosis

1. *Acute red eye.* Acute iridocyclitis must be differentiated from other causes of acute red eye, especially acute congestive glaucoma and acute conjunctivitis.

2. *Granulomatous versus non-granulomatous uveitis.* Once diagnosis of iridocyclitis is established, an attempt should be made to know whether the condition is of granulomatous or non-granulomatous type.

3. *Etiological differential diagnosis.* Efforts should also be made to distinguish between the different etiological varieties of iridocyclitis. This may be possible in some cases after thorough investigations and with knowledge of special features of different clinical entities, which are described under the subject of special types of iridocyclitis.

Investigations

1. *Haematological investigations TLC* and *DLC* (total leukocyte/white blood cells count, differential leukocyte count) to have general information about inflammatory response of body:

– *ESR* – to ascertain existence of any chronic inflammatory condition in the body.

– *Blood sugar levels* – to rule out diabetes mellitus.

– *Blood uric acid* in patients suspected of having gout.

– *Serological tests* for syphilis, toxoplasmosis, and histoplasmosis.

– *Tests* for antinuclear antibodies, Rh factor, LE (lupus erythematosus) cells, C-reactive proteins and antistreptolysin 0.

2. *Urine examination* for WBCs, pus cells, RBC and culture to rule out urinary tract infections.

3. *Stool examination* for cyst and ova to rule out parasitic infestations.

4. *Radiological investigations* include X-rays of chest, paranasal sinuses, sacroiliac joints and lumbar spine.

5. *Skin tests* include tuberculin test, Kveim test and toxoplasmin test.

Treatment of iridocyclitis

I. *Non-specific treatment*

Local therapy

1. *Mydriatic-cycloplegic drugs.* These are very useful and most effective during acute phase of iridocyclitis. Commonly used drug is 1 percent *atropine sulfate* eye ointment or drops 2–3 times a day. In case of atropine allergy, other cycloplegics like 2 percent *homatropine* or 1 percent *cyclopentolate* eyedrops may be instilled 3–4 times a day. Alternatively for more powerful cycloplegic effect a subconjunctival injection of 0.25 ml *mydricain* (a mixture of atropine, adrenaline and procaine) should be given. The cycloplegics should be continued for at least 2–3 weeks after the eye becomes better, otherwise relapse may occur.

Mode of action

In iridocyclitis, atropine gives comfort and rest to the eye by relieving spasm of iris sphincter and ciliary muscle, prevents the formation of synechiae and may break the already formed synechiae, reduces exudation by decreasing hyperaemia and vascular permeability and increases the blood supply to anterior uvea by relieving pressure on the anterior ciliary arteries. As a result more antibodies reach the target tissues and more toxins are absorbed.

2. *Corticosteroids* administered locally, are very effective in cases of iridocyclitis. They reduce inflammation by their anti-inflammatory effect; being anti-allergic, they are of special use in allergic type of uveitis; and due to their antifibrotic activity, they reduce fibrosis and thus prevent

disorganisation and destruction of the tissues. Commonly used steroidal preparations contain dexamethasone, betamethasone, hydrocortisone or prednisolone.

Route of administration: locally, steroids are used as eye drops 4–6 times a day, eye ointment at bed time, and anterior sub-Tenon injection is given in severe cases.

3. *Broad spectrum antibiotic drops* although are of no use in iridocyclitis, they are usually prescribed with topical steroid preparations to provide an umbrella cover for them.

Systemic therapy

1. *Corticosteroids.*

Dosage schedules: usually, treatment is started with high doses of prednisolone (60–100 mg) or equivalent quantities of other steroids (dexamethasone or betamethasone).

Daily therapy regime is preferred for marked inflammatory activity for at least 2 weeks. In the absence of acute disease, *alternate day therapy regime* should be chosen. The dose of steroids is decreased by a week's interval and tapered completely in about 6–8 weeks in both the regimes.

Note. Steroids (both topical and systemic) may cause many ocular (e. g., steroid-induced glaucoma and cataract) and systemic side-effects. Hence, eagle eye watchfulness is required for it

2. *Non-steroidal anti-inflammatory drugs* (NSAIDS).

3. *Immunosuppressive drugs.* These should be used only in desperate and extremely serious cases of uveitis, in which vigorous use of steroids have failed to resolve the inflammation and there is an imminent danger of blindness. These drugs are dangerous and should be used with great caution in the supervision of a haematologist and an oncologist. These drugs are especially useful in severe cases of Behcet's syndrome, sympathetic ophthalmia, pars planitis and VKH syndrome. A few available cytotoxic immunosuppressive drugs include cyclophosphamide, chlorambucil, azathioprine and methotrexate. Cyclosporin is a powerful anti-T-cell

immunosuppressive drug which is effective in cases resistant to cytotoxic immunosuppressive agents, but has high renal toxicity.

II. *Specific treatment of the cause*

Unfortunately, in spite of the advanced diagnostic tests, still it is not possible to ascertain the cause in a large number of cases.

So, a full course of anti-tubercular drugs for underlying Koch's disease, adequate treatment for syphilis, toxoplasmosis etc, when detected should be carried out. When no cause is ascertained, a full course of broad spectrum antibiotics may be helpful by eradicating some masked focus of infection in patients with non-granulomatous uveitis.

III. *Treatment of complications*

1. *Inflammatory glaucoma* (hypertensive uveitis). In such cases, drugs to lower intraocular pressure such as 0.5 % timolol maleate eyedrops twice a day and tablet acetazolamide (250 mg thrice a day) should be added, over and above the usual treatment of iridocyclitis. Pilocarpine and latanoprost eye drops are contraindicated in inflammatory glaucoma.

2. *Post-inflammatory glaucoma* due to ring synechiae is treated by laser iridotomy. Surgical iridectomy may be done when laser is not available.

However, surgery should be performed in a healthy eye only under high doses of corticosteroids.

3. *Complicated cataract* requires lens extraction with guarded prognosis in spite of all precautions. The presence of fresh KPs is considered a contraindication for intraocular surgery.

4. *Retinal detachment* of exudative type usually settles itself if uveitis is treated aggressively. A tractional detachment requires vitrectomy and management of complicated retinal detachment, with poor visual prognosis.

5. *Phthisis bulbi* especially when painful requires removal by enucleation operation.

Posterior Uveitis

Posterior uveitis refers to inflammation of the choroid (*choroiditis).* Since the outer layers of retina are in close contact with the choroid and also depend on it for the nourishment, the choroidal inflammation almost always involves the adjoining retina, and the resultant lesion is called *chorioretinitis.*

Etiology and pathology

These are the same as described for uveitis in general considerations.

Clinical types

I. *Suppurative choroiditis* (purulent inflammation of the choroid). It usually does not occur alone and almost always forms part of endophthalmitis.

II. *Non-suppurative choroiditis*. It may be non-granulomatous or granulomatous (more common).

Non-suppurative choroidal inflammation is characterized by exudation and cellular infiltration, resulting in a greyish white lesion hiding the normal reddish hue of choroidal vessels.

Non-suppurative choroiditis is usually bilateral and can be morphologically (depending upon the number and location of lesions) classified into *diffuse, disseminated* and *circumscribed* (*localized*) *choroiditis.*

1. *Diffuse choroiditis*. It refers to large spreading lesions involving most of the choroidal tissue. It is usually tubercular or syphilitic in origin.

2. *Disseminated choroiditis*. It is characterized by multiple but small areas of inflammation scattered over the greater part of choroid. Such a condition may be due to syphilis or tuberculosis, but in many cases the cause is obscure.

3. *Circumscribed/localized/focal choroiditis*. It is characterized by a single patch or a few small patches of inflammation localized in a particular area. Such patches of choroiditis are described by a name depending upon the location of the lesion:

Central choroiditis: as the name indicates it involves the macular area and may occur either alone or in combination with disseminated choroiditis. A typical patch of central choroiditis may occur in toxoplasmosis, histoplasmosis, tuberculosis, syphilis and rarely due to visceral larva migrans.

Juxtacaecal or *juxtapapillary choroiditis:* it is the name given to a patch of choroiditis involving an area adjoining the optic disc. One example is *Jensen's choroiditis* which typically occurs in young persons.

Anterior peripheral choroiditis: it implies occurrence of multiple small patches of choroiditis (similar to disseminated choroiditis) only in the peripheral part of choroid (anterior to equator). Such lesions are often syphilitic in origin.

Equatorial choroiditis: it involves the choroid in the equatorial region only.

Clinical picture

Symptoms

1. *Defective vision.* It is usually mild due to vitreous haze, but may be severe as in central choroiditis.

2. *Photopsia.* It is a subjective sensation of flashes of light resulting due to irritation of rods and cones.

3. *Black spots floating in front of the eyes.* It is a very common complaint of such patients. They occur due to large exudative clumps in the vitreous.

4. *Metamorphopsia.* Herein, patients perceive distorted images of the object. This results due to alteration in the retinal contour caused by a raised patch of choroiditis.

5. *Micropsia* which results due to separation of visual cells is a common complaint. Here, the objects appear smaller than they are.

6. *Macropsia*, i. e. perception of the objects larger than they are, may occur due to crowding together of rods and cones.

7. *Positive scotoma,* i. e. perception of a fixed large spot in the field of vision, corresponding to the lesion may be noted by many patients.

Complications

These include extension of the inflammation to anterior uvea, complicated cataract, vitreous degeneration, macular edema, secondary periphlebitis retinae and retinal detachment.

Treatment

It is broadly on the lines of anterior uveitis.

1. *Non-specific therapy* consists of topical and systemic corticosteroids. Posterior sub-tenon injections of corticosteroids are effective in checking the acute phase of posterior uveitis. Rarely, immunosuppressive agents may be required to check the inflammation.

2. *Specific treatment* is required for the causative disease such as toxoplasmosis, toxocariasis, tuberculosis, syphilis, etc.

Purulent Uveitis

Purulent uveitis is suppurative inflammation of the uveal tract occurring as a result of direct invasion by the pyogenic organisms. It may start as purulent anterior uveitis (iridocyclitis) or purulent posterior uveitis (choroiditis) which soon progresses to involve the retina and vitreous cavity, resulting in purulent endophthalmitis.

Endophthalmitis

Endophthalmitis is defined as an inflammation of the inner structures of the eyeball, i. e. uveal tissue and retina associated with pouring of exudates into the vitreous cavity, anterior and posterior chambers.

Etiology

Etiologically, endophthalmitis may be infectious or non-infectious (sterile).

A. *Infective endophthalmitis*

Modes of infection

1. *Exogenous infections.* Purulent inflammations are generally caused by exogenous infections following perforating injuries, perforation of infected corneal ulcers or as postoperative infections following intraocular operations.

2. *Endogenous or metastatic endophthalmitis.* It may occur rarely through the blood stream from some infected focus in the body such as dental caries, generalized septicemia and puerperal sepsis.

3. *Secondary infections from surrounding structures.* It is very rare. However, cases of purulent intraocular inflammation have been reported following extension of infection from orbital cellulitis, thrombophlebitis and infected corneal ulcers.

Causative organisms

1. *Bacterial endophthalmitis.* The most frequent pathogens causing acute bacterial endophthalmitis are gram positive cocci, i. e. staphylococcus epidermidis and staphylococcus aureus. Other causative bacteria include streptococci, pseudomonas, pneumococci and corynebacterium.

Propionibacterium acnes and actinomyces are gram-positive organisms capable of producing slow grade endophthalmitis.

2. *Fungal endophthalmitis* is comparatively rare. It is caused by aspergillus, fusarium, candida, etc.

B. *Non-infective (sterile) endophthalmitis*

Sterile endophthalmitis refers to inflammation of inner structures of eyeball caused by certain toxins/toxic substances. It occurs in the following situations:

1. *Postoperative sterile endophthalmitis* may occur as toxic reaction to chemicals adherent to intraocular lens (IOL) or chemicals adherent to instruments.

2. *Post-traumatic sterile endophthalmitis* may occur as toxic reaction to retained intraocular foreign body, e. g., pure copper.

3. *Intraocular tumour* necrosis may present as sterile endophthalmitis (masquerade syndrome).

4. *Phacoanaphylactic endophthalmitis* may be induced by lens proteins in patients with Morgagnian cataract.

Note. Since postoperative acute bacterial endophthalmitis is most important, clinical features and treatment described below pertain to this condition

Clinical picture

Acute postoperative endophthalmitis is a catastrophic complication of intraocular surgery with an incidence of about 0.1 %. Source of infection in most of the cases is thought to be patient's own periocular bacterial flora of the eyelids, conjunctiva, and lacrimal sac. Other potential sources of infection include contaminated solutions and instruments, and environmental flora including that of surgeon and operating room personnel.

Symptoms

Acute bacterial endophthalmitis usually occurs within 7 days of operation and is characterized by severe ocular pain, redness, lacrimation, photophobia and marked loss of vision.

Signs

1. *Lids* become red and swollen.

2. *Conjunctiva* shows chemosis and marked circumcorneal congestion.

Note. Conjunctival congestion, corneal edema, hypopyon and yellowish white exudates in the vitreous cavity seen in the pupillary area behind the IOL

3. *Cornea* is edematous, cloudy, and ring infiltration may be formed.

4. *Edges of wound* become yellow and necrotic and the wound may rupture in exogenous form.

5. *Anterior chamber* shows hypopyon; soon it becomes full of pus.

6. *Iris*, when visible, is edematous and muddy.

7. *Pupil* shows "yellow reflex" due to purulent exudation in vitreous cavity. When anterior chamber becomes full of pus, iris and pupil details are not seen.

8. *Vitreous exudation.* In metastatic forms and in cases with deep infections, vitreous cavity is filled with exudation and pus. Soon a yellowish white mass is seen through the fixed dilated pupil.

This sign is called *amaurotic cat's-eye reflex* or *leukocoria.*

9. *Intraocular pressure* is raised in early stages, but in severe cases, the ciliary processes are destroyed, and a fall in intraocular pressure may ultimately result in shrinkage of the eyeball.

Treatment

A. Antibiotic therapy

1. *Intravitreal antibiotics and diagnostic tap* should be made as early as possible. It is performed transconjunctivally under topical anesthesia from the area of pars plana (4–5 mm from the limbus).

Usually a combination of two antibiotics – one effective against gram-positive coagulase negative staphylococci and the other against gram-negative bacilli is used:

– *first choice:* vancomycin 1 mg in 0.1 ml plus ceftazidime 2.25 mg in 0.1 ml;

– *second choice:* vancomycin 1 mg in 0.1 ml plus amikacin 0.4 mg in 0.1 ml;

– *third choice:* vancomycin 1 mg in 0.1 ml plus gentamycin 0.2 mg in 0.1 ml.

Note:

1. Some surgeons prefer to add dexamethasone 0.4 mg in 0.1 ml to limit post-inflammatory consequences.

2. Gentamycin is 4 times more retinotoxic (causes macular infarction) than amikacin. Preferably the aminoglycosides should be avoided.

3. The aspirated fluid sample should be used for bacterial culture and smear examination. If vitreous aspirate is collected in an emergency when immediate facilities for culture are not available, it should be stored promptly in refrigerator at 4 °C.

4. If there is no improvement, a repeat intravitreal injection should be given after 48 hours taking into consideration the reports of bacteriological examination

2. *Subconjunctival injections* of antibiotics should be given daily for 5–7 days to maintain therapeutic intraocular concentration:

– *first choice:* vancomycin 25 mg in 0.5 ml plus ceftazidime 100 mg in 0.5 ml;

– *second choice:* vancomycin 25 mg in 0.5 ml plus cefuroxime 125 mg in 0.5 ml.

3. *Topical concentrated antibiotics* should be started immediately and used frequently (every 30 minutes to 1 hour). To begin with a combination of two drugs should be preferred, one having a predominant effect on the gram-positive organisms and the other against gram-negative organisms as

below: vancomycin (50 mg/ml) or cefazoline (50 mg/ml) or amikacin (20 mg/ml) or tobramycin (15 mg/ml).

4. *Systemic antibiotics* have a limited role in the management of endophthalmitis, but most of the surgeons use them. *Ciprofloxacin* intravenous infusion 200 mg Bd for 3–4 days followed by 500 mg Bd orally for 6–7 days, or *Vancomycin* 1 mg IV Bd and *ceftazidime* 2 g IV 8 hourly, or *Cefazoline* 1.5 gm IV 6 hourly and *amikacin* 1 mg IV three times a day.

B. Steroid therapy

Steroids limit the tissue damage caused by inflammatory process. Most surgeons recommend their use after 24 to 48 hours of control of infection by intensive antibiotic therapy. However, some surgeons recommend their immediate use (controversial). The routes of administration and doses are:

– *intravitreal injection* of dexamethasone 0.4 mg in 0.1 ml;

– *subconjunctival injection* of dexamethasone 4 mg (1 ml) OD for 5–7 days;

– *topical* dexamethasone (0.1 %) or predacetate (1 %) used frequently;

– *systemic steroids:* oral corticosteroids should preferably be started after 24 hours of intensive antibiotic therapy. A daily therapy regime with 60 mg prednisolone to be followed by 50, 40, 30, 20 and 10 mg for 2 days each may be adopted.

C. Supportive therapy

1. *Cycloplegics.* Preferably 1 % atropine or alternatively 2 % homatropine eyedrops should be instilled TDS or qid (3–4 times daily).

2. *Anti-glaucoma drugs.* In patients with raised intraocular pressure drugs such as oral acetazolamide (250 mg tds) and timolol (0.5 % Bd) may be prescribed.

D. Vitrectomy operation should be performed if the patient does not improve with the above intensive therapy for 48 to 72 hours or when the patient presents with severe infection with visual acuity reduced to light perception. Vitrectomy helps in removal of infecting organisms, toxins and enzymes present in the infected vitreous mass.

Panophthalmitis

It is an intense purulent inflammation of the whole eyeball including the Tenon's capsule. The disease usually begins either as purulent anterior or purulent posterior uveitis; and soon a full-fledged picture of panophthalmitis develops, following a very short stage of endophthalmitis.

Etiology: panophthalmitis is an acute bacterial infection.

Symptoms:

– severe ocular pain and headache;
– complete loss of vision;
– profuse watering;
– purulent discharge;
– marked redness and swelling of the eyes;
– associated constitutional symptoms are malaise and fever.

Signs:

– *lids* show a marked edema and hyperaemia;
– *eyeball* is slightly proptosed; ocular movements are limited and painful;
– *conjunctiva* shows marked chemosis with ciliary and conjunctival congestion;
– *cornea* is cloudy and edematous;
– *anterior chamber* is full of pus;
– *vision* is completely lost and perception of light is absent;
– *intraocular pressure* is markedly raised;

– *eyeball perforation* may occur at limbus, pus comes out and intraocular pressure falls.

Complications include:

– orbital cellulitis;
– cavernous sinus thrombosis;
– meningitis or encephalitis.

Treatment

There is little hope of saving such an eye and the pain and toxemia indicate urgency to its removal.

1. *Anti-inflammatory drugs and analgesics* should be started immediately to relieve pain.

2. *Broad spectrum antibiotics* should be administered to prevent further spread of infection in the surrounding structures.

3. *Evisceration* operation should be performed to avoid the risk of intracranial dissemination of infection.

Evisceration: it is the removal of the contents of the eyeball leaving behind the sclera. Frill evisceration is preferred over simple evisceration. In it, only about 3-mm frill of the sclera is left around the optic nerve.

Indications: panophthalmitis, expulsive choroidal haemorrhage and bleeding anterior staphyloma.

Specific Clinico-Etiological Types of Non-Suppurative Uveitis

Classification

I. Uveitis associated with chronic systemic bacterial infections:

1. Tubercular uveitis.
2. Syphilitic uveitis.
3. Leprotic uveitis.

II. Uveitis associated with noninfectious systemic diseases:

1. Uveitis in sarcoidosis.

2. Behcet's disease.

III. Uveitis associated with arthritis:

1. Uveitis with ankylosing spondylitis.
2. Reiter's syndrome.
3. Still's disease.

IV. Parasitic uveitis:

1. Toxoplasmosis.
2. Toxocariasis.
3. Onchocerciasis.
4. Amoebiasis.

V. Fungal uveitis:

1. Presumed ocular histoplasmosis syndrome.
2. Candidiasis.

VI. Viral uveitis:

1. Herpes simplex uveitis.
2. Herpes zoster uveitis.
3. Acquired cytomegalovirus uveitis.
4. Uveitis in AIDS.

VII. Lens induced uveitis:

1. Phacotoxic uveitis.
2. Phacoanaphylactic endophthalmitis.

VIII. Traumatic uveitis.

IX. Uveitis associated with malignant intraocular tumours.

X. Idiopathic specific uveitis syndromes:

1. Fuchs' uveitis syndrome.
2. Intermediate uveitis (pars planitis).
3. Sympathetic ophthalmitis.
4. Glaucomatocyclitic crisis.
5. Vogt–Koyanagi–Harada's syndrome.
6. Birdshot retinochoroidopathy.
7. AMPPE.
8. Serpiginous choroidopathy.

I. Uveitis in Chronic Systemic Bacterial Infections

Tubercular Uveitis

Tuberculosis is a chronic granulomatous infection caused by bovine or human tubercle bacilli. It may cause both anterior and posterior uveitis. At one time very common, it is now becoming a rare cause. It accounts for 1 % of uveitis in developed countries. However, in developing countries it still continues to be a common cause of uveitis.

1. *Tubercular anterior uveitis.* It may occur as acute non-granulomatous iridocyclitis or granulomatous anterior uveitis which in turn may be in the form of miliary tubercular iritis or conglomerate granuloma (solitary tuberculoma).

2. *Tubercular posterior uveitis.* It may occur as *multiple miliary tubercles* in the choroid which appear as round yellow white nodules one-sixth to two and half disc diameter in size. These are:

– usually associated with tubercular meningitis;

– diffuse or disseminated choroiditis in chronic tuberculosis;

– rarely a large solitary choroidal granuloma.

3. *Vasculitis* (Eales' disease).

Diagnosis

There is no specific clinical finding in tubercular uveitis. Diagnosis is made from positive skin test, associated findings of systemic tuberculosis, intractable uveitis unresponsive to steroid therapy, a positive response to isoniazid test (a dramatic response of iritis to isoniazid 300 mg once daily for 3 weeks).

Treatment

In addition to usual treatment of uveitis, chemotherapy with rifampicin and isoniazid should be given for 12 months. Systemic corticosteroids should be deferred.

Acquired Syphilitic Uveitis

Acquired syphilis is a chronic venereal infection caused by *Treponema Pallidum* (spirochaete). It affects both the anterior and posterior uvea.

1. *Syphilitic anterior uveitis.* It may occur as acute plastic iritis or granulomatous iritis. *Acute plastic iritis* typically occurs in the secondary stage of syphilis and also as a Herxheimer's reaction 24–48 hours after therapeutic dose of the penicillin.

Gummatous anterior uveitis occurs late in the secondary or rarely during the tertiary stage of syphilis. It is characterised by formation of yellowish red highly vascularised multiple nodules arranged near the pupillary border or ciliary border of iris.

2. *Syphilitic posterior uveitis.* It may occur as disseminated, peripheral or diffuse choroiditis.

Diagnosis

Once suspected clinically, diagnosis is confirmed by FTA-ABS (fluorescent treponemal antibody absorption) blood test, which is specific and more sensitive than TPI (treponema pallidum immobilisation) test and VDRL tests.

Treatment

In addition to local therapy of the uveitis, patient should be treated by systemic penicillin or other antisyphilitic drugs.

Leprotic Uveitis

Leprosy (Hansen's disease) is caused by *mycobacterium leprae* which is an acid-fast bacillus.

The disease occurs in two principal forms: *lepromatous* and *tuberculoid*.

Treatment

Besides usual local therapy of iridocyclitis, antileprotic treatment with Dapsone 50–100 mg daily or other drugs should also be instituted.

II. Uveitis in Non-Infectious Systemic Diseases

Uveitis in Sarcoidosis

Sarcoidosis is a multi-system disease of unknown etiology, characterised by formation of noncaseating epithelioid cell granuloma in the affected tissue. The disease typically affects young adults, frequently presenting with bilateral hilar lymphadenopathy, pulmonary infiltration, skin and ocular lesions.

Ocular lesions occur in 20–50 percent of patients and include: uveitis, vitritis with snowball opacities in inferior vitreous, choroidal and retinal granulomas, periphlebitis retinae with "candle wax droppings", conjunctival sarcoid nodule and keratoconjunctivitis sicca.

Clinical types

Sarcoid uveitis accounts for 2 percent of cases of uveitis. It may present as one of the following:

1. *Acute iridocyclitis* (non-granulomatous). It is frequently unilateral, associated with acute sarcoidosis characterised by hilar lymphadenopathy and erythema nodosum.

2. *Chronic iridocyclitis.* It is more common than acute and presents with typical features of bilateral granulomatous iridocyclitis. The disease is often seen in association with chronic sarcoidosis characterised by pulmonary fibrosis.

3. *Uveoparotid fever (Heerfordt syndrome).* It is characterised by bilateral granulomatous panuveitis, painful enlargement of parotid glands, cranial nerve palsies, skin rashes, fever and malaise.

Diagnosis

Once suspected clinically, it is supported by positive Kveim test, abnormal X-ray chest (in 90 percent cases) and raised levels of serum angiotensin converting enzyme (ACE). *Confirmation* of the disease is made by histological proof from

biopsy of the conjunctival nodule, skin lesions or enlarged lymph node.

Treatment

Topical, periocular and systemic steroids constitute the treatment of sarcoid uveitis, depending upon the severity.

Behcet's Disease

Behcet's (beh-CHETS) *disease*, also called *Behcet's syndrome*, is an idiopathic multisystem disease characterized by recurrent, non-granulomatous uveitis, aphthous ulceration, genital ulcerations and erythema multiform.

Etiology

It is still unknown; the basic lesion is an obliterative vasculitis probably caused by circulating immune complexes. The disease typically affects young men who are positive for HLA-B51.

Clinical features

Uveitis seen in Behcet's disease is typically bilateral, acute recurrent iridocyclitis associated with hypopyon. It may also be associated with posterior uveitis, vitritis, periphlebitis retinae and retinitis in the form of white necrotic infiltrates.

Treatment

No satisfactory treatment is available, and thus the disease has got comparatively poor visual prognosis. *Corticosteroids* may be helpful initially but ultimate response is poor. In some cases the disease may be controlled by *chlorambucil.*

Vogt–Koyanagi–Harada (VKH) Syndrome

It is an idiopathic multisystem disorder which includes cutaneous, neurological and ocular lesions. The disease is comparatively more common in Japanese who are usually positive for HLA–DR4 and DW15.

Clinical features

1. *Cutaneous lesions* include: alopecia, poliosis and vitiligo.

2. *Neurological lesions* are in the form of meningism, encephalopathy, tinnitus, vertigo and deafness.

3. *Ocular features* are bilateral chronic granulomatous anterior uveitis, posterior uveitis and exudative retinal detachment.

Treatment

It comprises steroids administered topically, periocularly and systemically.

III. Uveitis in Arthritis

Uveitis with Ankylosing Spondylitis

Ankylosing spondylitis is an idiopathic chronic inflammatory arthritis, usually involving the sacroiliac and posterior inter-vertebral joints. The disease affects young males (20–40 years) who are positive for HLA–B27. About 30 to 35 percent patients with ankylosing spondylitis develop uveitis.

Uveitis associated with ankylosing spondylitis is characteristically an acute, recurrent, non-granulomatous type of iridocyclitis. The disease usually affects one eye at a time.

Treatment

It is on the lines of usual treatment of anterior uveitis. Long-term aspirin or indomethacin may decrease the recurrences.

Reiter's Syndrome

It is characterized by a triad of urethritis, arthritis and conjunctivitis with or without iridocyclitis.

Etiology

It is not known exactly. The syndrome typically involves young males who are positive for HLA–B27. The

disease occurs in three forms: post venereal due to non-gonococcal arthritis, post dysenteric and articular form.

Treatment

Iridocyclitis responds well to usual treatment. A course of systemic tetracycline 250 mg qid (4 times daily) for 10 days may be useful in post-venereal form suspected of being caused by Chlamydia infection.

Juvenile Chronic Arthritis

Juvenile chronic arthritis (JCA) is an idiopathic chronic inflammatory arthritis involving multiple joints (knee, elbow, ankle and interphalangeal joints) in children below the age of 16 years.

Anterior uveitis associated with JCA is a bilateral (70 %), chronic non-granulomatous disease, affecting female children more than male (4:1). It usually develops before the age of 6 years. Nearly half of the patients are positive for HLA–DW5 and 75 percent are positive for antinuclear antibodies (ANA). The onset of uveitis is asymptomatic and the eye is white even in the presence of severe uveitis. Therefore, slit-lamp examination is mandatory in children suffering from JCA.

Complications like posterior synechiae, complicated cataract and band-shaped keratopathy are fairly common.

Treatment is on the usual lines.

IV. Parasitic Uveitis

Toxoplasmosis

It is a protozoan infestation caused by *toxoplasma gondii*, derived from cats (definitive host). Humans and other animals (cattle, sheep and pigs) are intermediate hosts. The disease primarily affects central nervous system (brain) and retina. Systemic toxoplasmosis occurs in humans in two forms: congenital and acquired.

1. *Congenital toxoplasmosis.* It is much more common than the acquired form, and the infestation is acquired by the fetus through transplacental route after the mother contracts acute infestation during pregnancy. When pregnant females catch the disease, about 49 percent of infants are born with the disease which may be active or inactive at birth.

The characteristic *triad of congenital toxoplasmosis* includes: convulsions, chorio-retinitis and intracranial calcification. In active stage the typical lesion is *necrotic granulomatous retinochoroiditis* involving the macular region. Most of the infants are born with inactive disease, characterised by bilateral healed punched out heavily pigmented chorioretinal scars in the macular area, which is usually discovered when the child is brought for defective vision or squint check up.

2. *Acquired toxoplasmosis.* It is very rare (of doubtful existence). The infestation is acquired by eating under-cooked meat of an intermediate host containing cyst-form of the parasite. Most of the patients are subclinical (asymptomatic); and the typical chorioretinal lesion similar to congenital toxoplasmosis is discovered by chance.

Diagnosis

The clinically suspected lesion is confirmed by indirect-fluorescent antibody test, haemagglutination test or ELISA test. The old methylene blue dye test is obsolete.

Treatment

The active lesion of toxoplasmosis is treated by topical and systemic steroids along with a course of an antitoxoplasmic drug: spiramycin, clindamycin, sulfadiazine or pyremethamine.

Toxocariasis

It is an infestation caused by an intestinal round worm of dogs (*toxocara canis*) and cats (*toxocara catis*).

Young children who play with dogs and cats or eat dirt are infested by ova of these worms. These ova develop into larvas in the human gut, and then produce the condition visceral larva migrans (VLM).

Ocular toxocariasis

It is ocular infestation by these larvas and is almost always unilateral. Clinically it can present as follows:

1. *Toxocara chronic endophthalmitis.*
2. *Posterior pole granuloma.*
3. *Peripheral granuloma.*

Diagnosis is made on the basis of clinical picture and ELISA blood test.

Treatment

It consists of periocular (posterior sub-tenon) injection of steroid and systemic steroids. Pars plana vitrectomy may be required in unresponsive patients with endophthalmitis and in patients with vitreous band formation.

V. Fungal Uveitis

Presumed Ocular Histoplasmosis Syndrome (POHS)

Etiology

It is thought to be caused by the fungus *Histoplasma capsulatum.*

Clinical features

POHS is characterised by the following features:

1. *Histospots.* These are atrophic spots scattered in the mid-retinal periphery. They are roundish, yellowish-white lesions measuring 0.2 to 0.7 disc diameter in size. These begin to appear in early childhood and represent the scars of disseminated histoplasma choroiditis.

2. *Macular lesion.* It starts as atrophic macular scar (macular histospot); followed by a hole in the Bruch's membrane, which then allows ingrowth of capillaries leading to sub-retinal choroidal neovascularisation. Leakage of fluid

from the neovascular membrane causes serous detachment, which when complicated by repeated haemorrhages constitutes haemorrhagic detachment. Ultimately, fibrous disciform scar develops, which is associated with a marked permanent visual loss.

Diagnosis

The clinical diagnosis is supported but not confirmed by positive histoplasmin test, and complement fixation tests (negative in two thirds of cases). Fluorescein angiography helps in early diagnosis of subretinal neovascular membrane.

Treatment

Early argon laser photocoagulation of subretinal neovascular membrane may prevent marked permanent visual loss which occurs due to fibrous disciform scars.

Candidiasis

It is an opportunistic infection caused by *Candida albicans*. It occurs in immuno-compromised patients including: patients suffering from AIDS, malignancies, those receiving long-term antibiotics, steroids or cytotoxic drugs. Patients with long-term indwelling intravenous catheter used for haemodialysis, and drug addicts are also prone to such infection.

Ocular candidiasis is not a common condition. It may occur as anterior uveitis, multifocal chorioretinitis, or endophthalmitis.

Treatment

It consists of topical cycloplegics, and antifungal drugs.

Systemic antifungal drugs like ketoconazole, flucytosine or amphotericin-B are also needed.

Pars plana vitrectomy is required for candida endophthalmitis.

VI. Viral Uveitis

Uveitis in Herpes Zoster Ophthalmicus

Herpes zoster ophthalmicus (HZO) is the involvement of ophthalmic division of the fifth cranial nerve by varicella zoster.

Anterior uveitis develops in 40–50 percent cases with HZO within 2 weeks of onset of the skin rashes. A typical HZO keratitis may be associated with mild iritis especially in patients with a vesicular eruption on the tip of nose. The iridocyclitis is *non-granulomatous* characterized by presence of small KPs, mild aqueous flare and occasional hemorrhagic hypopyon.

Complications like iris atrophy and secondary glaucoma are not uncommon. Complicated cataract may also develop in late stages.

Treatment

Topical steroids and cycloplegics are to be continued for several months. *Systemic acyclovir* helps in early control of lesions of HZO.

Herpes Simplex Uveitis

It is associated with keratitis in most of the cases. It may be seen in association with dendritic or geographical corneal ulceration or with disciform keratitis. Rarely, anterior uveitis may occur even without keratitis. It is a mild grade non-granulomatous iridocyclitis excited by hypersensitivity reaction.

Treatment

Keratitis is treated with antiviral drugs and cycloplegics. *Steroids for iritis are contraindicated* in the presence of active viral ulcers. Nonsteroidal anti-inflammatory drugs may be added in such cases.

Cytomegalic Inclusion Disease

It is a multisystem disease caused by cytomegalovirus (CMV). It occurs in two forms: congenital and acquired.

1. *Congenital cytomegalic inclusion disease*. It affects neonates. The infection is acquired either transplacentally in utero or during birth from the infected cervix of mother. Its common systemic features are sensory deafness, mental retardation and convulsions.

Ocular involvement occurs in the form of peripheral, central or total necrotizing chorioretinitis with associated vitreous haze. Posterior pole is involved more commonly and the lesions may be similar to those found in congenital toxoplasmosis. Secondary involvement of anterior uvea may occur rarely.

2. *Acquired cytomegalic inclusion disease*. It occurs only in immunosuppressed patients (due to any cause). The infection may be acquired by droplet infection or by transfusion of fresh blood containing infected white cells.

Ocular involvement is in the form of *CMV retinitis* characterized by presence of yellow-white exudates (areas of retinal necrosis) associated with areas of vasculitis and retinal hemorrhages. Some eyes may develop exudative retinal detachment. Ultimately, total retinal atrophy occurs.

Treatment

There is no specific treatment of CID.

Recently treatment with intravenous dihydroxypropylmethyl guanine has been shown to cause regression in some cases.

VII. Lens-Induced Uveitis

Phacoanaphylactic Uveitis

It is an immunological response to lens proteins in the sensitized eyes presenting as severe granulomatous anterior uveitis. The disease may occur following extracapsular cataract extraction, trauma to lens or leak of proteins in hypermature cataract.

Clinical features

These include severe pain, loss of vision, marked congestion and signs of granulomatous iridocyclitis associated with presence of lens matter in the anterior chamber.

Treatment

It consists of removal of causative lens matter, topical steroids and cycloplegics. Visual prognosis is usually poor.

Phacotoxic Uveitis

It is an ill-understood entity. This term is used to describe mild iridocyclitis associated with the presence of lens matter in the anterior chamber either following trauma or extracapsular cataract extraction or leak from hypermature cataracts. The uveal response due to direct toxic effect of lens matter or a mild form of allergic reaction is yet to be ascertained.

Treatment

It consists of removal of lens matter, topical steroids and cycloplegics.

VIII. Traumatic Uveitis

It is often seen in accidental or operative injuries to the uveal tissue. Different mechanisms which may produce uveitis following trauma include:

- direct mechanical effects of trauma;
- haemophthalmitis;
- microbial invasion;
- chemical effects of retained intraocular foreign bodies;
- sympathetic ophthalmia in the other eye.

IX. Uveitis Associated With Intraocular Tumours

X. Idiopathic Specific Uveitis Syndromes

Fuchs' Uveitis Syndrome (FUS)

Fuchs' heterochromic iridocyclitis is a chronic non-granulomatous type of low grade anterior uveitis. It typically occurs unilaterally in middle-aged persons.

The disease is characterised by:

– heterochromia of iris;
– diffuse stromal iris atrophy;
– fine KPs at back of cornea;
– faint aqueous flare;
– absence of posterior synechiae;
– fairly common rubeosis iridis, sometimes associated with neovascularization of the angle of anterior chamber;
– comparatively early development of complicated cataract and secondary glaucoma (usually open angle type).

Treatment

Topical corticosteroids are all that is required. Cycloplegics are not required as usually there are no posterior synechiae.

Glaucomatocyclitic Crisis

Posner–Schlossman syndrome is characterized by: recurrent attacks of acute rise of intraocular pressure (40–50 mm of Hg) without shallowing of anterior chamber associated with fine KPs at the back of cornea, without any posterior synechiae, epithelial edema of cornea, a dilated pupil, and a white eye (no congestion).

The disease typically affects young adults, 40 percent of whom are positive for HLA–BW54.

Treatment

It includes medical treatment to lower intraocular pressure along with a short course of topical steroids.

Sympathetic Ophthalmitis

It is a rare bilateral granulomatous panuveitis which is known to occur following penetrating ocular trauma usually associated with incarceration of uveal tissue in the wound. The injured eye is called "exciting eye" and the other eye which also develops uveitis is called "sympathising eye".

Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE)

It is a rare idiopathic self-limiting disorder characterized by bilateral, deep, placoid, cream coloured or grey white chorioretinal lesions involving the posterior pole and post-equatorial part of the fundus. Visual loss seen in early stage due to macular lesions, usually recovers within 2 weeks.

Complications though rare include mild anterior uveitis, vascular sheathing, and exudative retinal detachment. After healing, multifocal areas of depigmentation and pigment clumping involving the retinal pigment epithelium is left. *No treatment* is effective.

Serpiginous Geographical Choroidopathy

It is a rare, idiopathic, recurrent, bilaterally asymmetrical inflammation involving the choriocapillaries and pigment epithelium of the retina. The disease typically affects patients between 40 and 60 years of age and is characterized by cream coloured patches with hazy borders present around the optic disc which spread in a tongue fashion. After few weeks the lesions heal leaving behind punched out areas of retinal pigment epithelium and choroidal atrophy. *No treatment* is effective.

Intermediate Uveitis (Pars Planitis)

It denotes inflammation of pars plana part of ciliary body and most peripheral part of the retina.

Etiology

It is an idiopathic disease usually affecting both eyes (80 percent) of children and young adults.

Pars planitis is a rather common entity, constituting 8 percent of uveitis patients.

Symptoms

Most of the patients present with history of floaters. Some patients may come with defective vision due to associated cystoid macular edema.

Signs

The eye is usually without changes. Slit-lamp examination may show: mild aqueous flare, and fine KPs at the back of cornea. Anterior vitreous may show cells. Fundus examination with indirect ophthalmoscope reveals whitish exudates near the ora serrata in the inferior quadrant. These typical exudates are referred as *snow ball* opacities. These may coalesce to form a grey white plaque called *snow banking.*

Complications of long-standing pars planitis include: cystoid macular edema, complicated cataract and traction retinal detachment.

Treatment

1. *Corticosteroids* administered systemically and periocular injections may be effective in some cases.

2. *Immunosuppressive drugs* may be helpful in steroid resistant cases.

3. *Peripheral cryotherapy* is also reported to be effective.

Degenerative Conditions of the Uveal Tract

Degenerations of the Iris

1. *Simple iris atrophy.* It is characterized by depigmentation and thinning of iris stroma.

2. *Essential iris atrophy.* It is a rare idiopathic condition characterized by unilateral progressive atrophy of the iris

3. *Iridoschisis.* It is characterized by formation of a cleft between the anterior and posterior stroma of the iris.

Degenerations and Dystrophies of the Choroid

Primary choroidal degenerations

1. *Senile central choroidal atrophy.* It is characterized by formation of multiple drusens (colloid bodies) which look like yellowish spots. These are scattered throughout the fundus, but more marked in the macular area.

2. *Central areolar choroidal atrophy.* It comprises bilateral punched out, circular atrophic lesion in the macular region. The lesion is characterized by white shining sclera, traversed by large ribbon-shaped choroidal vessels. Thus, there occurs atrophy of the choriocapillaries, retinal pigment epithelium and photoreceptors.

3. *Essential gyrate atrophy.* It is an inborn error of amino acid (ornithine) metabolism characterized by progressive patches of atrophy of choroid and retinal pigment epithelium (RPE). The disease begins in the first decade of life with symptoms of night blindness and progresses slowly to involve the whole fundus by the age of 40–50 years with preservation of only the macula.

4. *Choroidremia.* It is a hereditary choroidal dystrophy involving the males. The fundus picture is characterized by whitish sclera with overlying almost normal retinal vessels.

5. *Myopic chorioretinal degeneration.*

Secondary choroidal degeneration

It occurs following inflammatory lesions of the fundus. It is characterized by a scattered area of chorioretinal atrophy and pigment clumping. Ophthalmoscopic picture resembles

retinitis pigmentosa and hence also labelled sometimes as *pseudoretinitis pigmentosa.*

Tumours of the Uveal Tract

Classification:

I. Tumours of choroid

Benign tumours: naevus, haemangioma, melanocytoma, choroidal osteoma.

Malignant tumour: melanoma.

Treatment

1. *Conservative treatment:* to salvage the eyeball should be tried unless the tumour is very large. Methods used and their indications are:

- brachytherapy;
- external beam radiotherapy;
- transpupillary thermotherapy (TTT);
- trans-scleral local resection;
- stereostatic radiosurgery.

2. *Enucleation.* It is indicated for very large tumours in which conservative methods to salvage the eyeball are not effective.

3. *Exenteration* or *debulking* with chemotherapy and radiotherapy is required in the stage of extraocular spread.

4. *Palliative treatment* with chemotherapy and immunotherapy may be of some use in prolonging life of the patients with distant metastasis.

II. Tumours of ciliary body

Benign tumours: hyperplasia, meduloepithelioma, benign cyst.

Malignant tumour: melanoma.

Treatment

1. Enucleation.

2. Local resection (cyclectomy or irido-cyclectomy).

III. Tumours of iris

Benign tumours: naevus, naevoxanthoendothelioma, benign cyst.

Malignant tumour: melanoma.

Treatment

1. *Wide iridectomy.* It is performed for a tumour limited to the iris.

2. *Iridocyclectomy.* It is required for a tumour involving iris and ciliary body.

3. *Enucleation.*

Investigations:

– indirect ophthalmoscopic examination;
– transillumination test;
– ultrasonograph;
– fluorescein;
– radioactive tracer;
– MRI.

TOPIC 10. GLAUCOMA

Glaucoma is not a single disease process but a group of disorders characterized by a progressive optic neuropathy resulting in a characteristic appearance of the optic disc and a specific pattern of irreversible visual field defects that are associated frequently but not invariably with raised intraocular pressure (IOP). Thus, IOP is the most common risk factor but not the only risk factor for development of glaucoma.

Consequently the term "ocular hypertension" is used for cases having constantly raised IOP without any associated glaucomatous damage. Conversely, the term "normal or low tension glaucoma" (NTG/LTG) is suggested for the typical cupping of the disc and/or visual field defects associated with a normal or low IOP.

The range of normal IOP measurements is 16 to 24 mm Hg. An IOP measurement of 24 mm Hg or greater is considered abnormal.

Classification

I. Congenital and developmental glaucoma:

1. Primary congenital glaucoma (without associated anomalies).

2. Developmental glaucoma (with associated anomalies).

II. Primary adult glaucoma:

1. Primary open angle glaucoma (POAG).

2. Primary angle closure glaucoma (PACG).

3. Primary mixed mechanism glaucoma.

III. Secondary glaucoma:

1. Depending on the mechanism of rise in IOP:

– secondary open angle glaucoma in which aqueous outflow may be blocked by a pretrabecular membrane, trabecular clogging, edema and scarring or elevated episcleral venous pressure.

2. Secondary angle closure glaucoma which may or may not be associated with pupil block.

3. Depending on the causative primary disease, secondary glaucoma is named as follows:

– lens-induced (phacogenic) glaucoma;

– inflammatory glaucoma (glaucoma due to intraocular inflammation);

– pigmentary glaucoma;

– neovascular glaucoma;

– glaucoma associated with irido-corneal endothelial syndromes;

– pseudo-exfoliative glaucoma;

– glaucoma associated with intraocular haemorrhage;

– steroid-induced glaucoma;

– traumatic glaucoma;

– glaucoma-in-aphakia;

– glaucoma associated with intraocular tumours.

Risk Factors for Open-Angle Glaucoma

Strong risk factors for open-angle glaucoma include:

– high intraocular pressure;

– family history of glaucoma;

– age 40 and older for African Americans;

– age 60 and older for the general population, especially Mexican Americans;

– thin cornea;

– suspicious optic nerve appearance with increased cupping (size of cup, the space at the centre of optic nerve, is larger than normal).

Potential risk factors for open-angle glaucoma include:

– high myopia (very severe near sightedness);

– diabetes;

– eye surgery or injury;

– high blood pressure;

– use of corticosteroids (for example, eye drops, pills, inhalers, and creams).

Risk Factors for Angle-Closure Glaucoma:

– age 40 and older;
– family history of glaucoma;
– poor short-distance vision (farsightedness);
– eye injury or eye surgery;
– East Asian and Inuit ethnicity.

Risk Factors for Normal-Tension Glaucoma:

– cardiovascular disease;
– family history of glaucoma;
– low intraocular pressure;
– Japanese ethnicity.

Pathogenesis of Ocular Damage in Glaucoma

As mentioned in definition, all kinds of glaucoma (classified above and described later) are characterized by a progressive optic neuropathy. It has now been recognized that progressive optic neuropathy results from the death of retinal ganglion cells (RGCs) in a typical pattern which results in characteristic optic disc appearance and specific visual field defects.

Congenital Glaucoma

The *congenital glaucoma* is a group of diverse disorders in which abnormally high intraocular pressure results due to developmental abnormalities of the angle of anterior chamber that obstructs the drainage of aqueous humor. Sometimes glaucoma may not occur until several years after birth; therefore, the term *developmental glaucoma* is preferred to describe such disorders.

Congenital glaucoma is a heterogeneous group of diseases with the following classifications based on age:

– congenital glaucoma (~ 40 % of cases) is existent or becomes evident at birth;

– infantile glaucoma (~ 50 % of cases) becomes evident during early childhood (< 3 years old);

– juvenile glaucoma (~ 10 % cases) becomes apparent in later childhood (> 3 years old).

Signs:

– enlarged optic cupping (cup to disc ration > 0.2) and/or asymmetric cupping;

– newborn IOP > 10–12 mm Hg;

– anterior insertion of the iris and Barkan membrane;

– Haab's striae (cracks in Descemet membrane);

– increased horizontal corneal diameter (above 11mm in first year of life, above 13 mm thereafter).

Symptoms:

– triad:

· *epiphora*;

· *photophobia*;

· *blepharospasm*;

– corneal opacities;

– buphthalmos;

– aniridia;

– iris heterochromia;

– ectopia lens;

– nystagmus;

– microcystic edema;

– lens dislocation and eyeball perforation (in severe, untreated cases).

Treatment of congenital glaucoma is primarily surgical. However, IOP must be lowered by use of hyperosmotic agents, acetazolamide and beta blockers till surgery is taken up. Miotics are of no use in such cases.

Primary Open Angle Glaucoma

Primary open-angle glaucoma (POAG), also referred to as *chronic simple glaucoma*, is a generally bilateral disease of adult onset characterized by:

– An IOP > 21 mm Hg at some stage;
– glaucomatous optic nerve damage;
– an open anterior chamber angle;
– characteristic visual field loss as damage progresses;
– absence of signs of secondary glaucoma or a non-glaucomatous cause for the optic neuropathy.

POAG is the most prevalent type of glaucoma in individuals of European and African ethnic origin. It affects both sexes equally. The eye has more optic nerve damage.

Signs

The majority of patients with POAG are asymptomatic. Typically, patients are only symptomatic in late disease, when they may become aware of constricted visual field or blurred vision. Occasionally, patients become aware of earlier visual field defects when performing monocular tasks (such as using the viewfinder of a camera). More frequently, patients are usually diagnosed as part of a routine eye test or as an incidental finding when presenting with another ophthalmic condition (e. g., diabetic retinopathy). A detailed general history would reveal any causes of secondary glaucoma (and therefore refuting POAG), such as steroid usage.

Medical therapy

Medical therapy may be topical or systemic. However, it is important to note that topical medication may cause significant systemic side effects, especially beta-blockers.

Topical therapy

– alpha-agonists (apraclonidine, brimonidine);
– beta-blockers (timolol, betaxolol, carteolol, levobunolol, etc.);

– carbonic anhydrase inhibitors (brinzolamide, dorzolamide);
– miotics (pilocarpine, etc.);
– prostaglandins (latanoprost, bimatoprost, travoprost).

Surgery

Non-medical interventions for POAG include:

– laser trabeculoplasty;
– trabeculectomy +/– augmentation;
– non-penetrating drainage surgery;
– shunt procedures;
– cyclodestructive procedures (cyclodiode, cyclocryotherapy).

Primary Angle-Closure Glaucoma

It is a type of primary glaucoma (wherein there is no obvious systemic or ocular cause) in which the rise in intraocular pressure occurs due to blockage of the aqueous humor outflow by closure of a narrower angle of the anterior chamber.

Classification:

– latent primary angle-closure glaucoma (primary angle-closure glaucoma suspect);
– subacute (intermittent) primary angle-closure glaucoma;
– acute primary angle-closure glaucoma;
– post-congestive angle-closure glaucoma;
– chronic primary angle-closure glaucoma;
– absolute glaucoma.

Symptoms

Most patients with angle-closure are asymptomatic, including a majority of those with intermittently or chronically elevated IOP.

Some patients present acutely (congestive glaucoma) with haloes around lights due to corneal edema, ocular pain and headache.

Other patients may have intermittent milder symptoms of blurring (‘smoke-filled room’) unassociated with pain.

Precipitating factors include watching television in a darkened room, reading, pharmacological mydriasis or miosis, acute emotional stress and rarely systemic medication: parasympathetic antagonists or sympathetic agonists (e. g., inhalers, motion sickness patches and cold remedies) and topiramate.

Sings

In a PAC suspect the eye may appear normal (with the exception of a narrow angle, as judged by the van Herick technique or by gonioscopy). In cases with a narrow van Herick angle (≤ 25 % [Grade 1 or 2]) with a normal anterior chamber depth, plateau iris – a type of narrow angle, should be suspected:

– limbal and conjunctival vessels dilated, producing ciliary flush;

– pupil fixed, semi-dilated, vertically elliptical, iris whirling;

– corneal edema;

– shallow ac with peripheral irido-corneal contact;

– high intraocular pressure (40–80mmhg);

– ac flare and cells;

– optic disc edematous and hyperemic;

– grey/white anterior sub-capsular lenticular opacities.

Acute angle-closure glaucoma

Treatment must be initiated immediately because vision can be lost quickly and permanently.

The patient should receive several drugs at once. A suggested regimen is:

– timolol ophthalmic: (0.25 % or 0.5 %) 1 drop into the affected eye(s) twice daily; (0.5 % gel) 1 drop into the affected eye(s) once daily;

– pilocarpine 2 to 4 % one drop q 15 min for 2 doses;

– brimonidine ophthalmic: (0.1 to 0.2 %) 1 drop into the affected eye(s) 3 times daily;

– acetazolamide: 125–250 mg orally (immediate-release) up to four times daily, maximum 1000 mg/day; 250–500 mg intravenously every 2–4 hours, maximum 1000 mg/day;

– osmotic agent, such as oral glycerol 1 mL/kg diluted with an equal amount of cold water, mannitol, 1.5 to 2 g/kg/dose intravenously over 30 minutes.

Response is evaluated by measuring IOP. Miotics (e. g., pilocarpine) are generally not effective when IOP is > 40 or 50 mm Hg because of an anoxic pupillary sphincter.

Definitive treatment is with laser peripheral iridotomy (LPI), which opens another pathway for fluid to pass from the posterior to the anterior chamber, breaking the pupillary block. It is done as soon as the cornea is clear and inflammation has subsided. In some cases the cornea clears within hours of lowering the IOP; in other cases, it can take 1 to 2 days.

Because the chance of having an acute attack in the other eye is 80 %, LPI is done on both eyes.

The risk of complications with LPI is extremely low compared to its benefits. Glare, which can be bothersome, may occur.

Normal-Pressure Glaucoma

Normal-pressure glaucoma (NPG), also referred to as normal- or low-tension glaucoma, is a variant of POAG.

Signs

Characteristic focal or diffuse thinning of the optic nerve head rim, as discussed above, is the physical examination hallmark of all glaucomatous disease. Characteristically, the

following features may be more frequently seen in NTG compared to POAG:

– flame shaped hemorrhages of the optic nerve rim (Drance hemorrhage);

– deep, focal notching of the rim;

– peripapillary atrophy.

Physical examination

Complete ophthalmologic examination is essential as a means of excluding other forms of glaucoma or confounding visually significant conditions (such as other forms of optic neuropathy) that may shape interpretation of subsequent imaging or perimetry testing. Features of the clinical exam should include:

– visual acuity;

– colour vision testing (to help differentiate from non-glaucomatous optic neuropathies);

– intraocular pressure measurement;

– diurnal or supine intraocular pressure measurement (if possible);

– pachymetry;

– afferent pupillary response testing;

– gonioscopy;

– complete slit lamp examination of the anterior segment;

– dilated fundus examination with optic nerve head and retinal nerve fiber layer (RNFL) assessment.

Medical therapy

Topical IOP lowering medications including prostaglandin analogues, alpha-2 agonists, beta-blockers, and carbonic anhydrase inhibitors are the mainstays of medical therapy.

Secondary Open Angle Glaucoma

Secondary glaucoma happens because of something else. It can be something in the eye or in the body that affects the eye. If we total up all those with glaucoma in the world, the secondary ones are still a fair number, maybe 10 % of all glaucoma. They much more often affect one eye and not both eyes, unlike primary open angle or angle closure that affect both eyes. All secondary glaucoma share the feature that the eye pressure is above normal due to something that causes abnormal outflow of aqueous.

Inflammatory Glaucoma

This type of glaucoma is caused by eye inflammation or uveitis. In uveitic open angle glaucoma, there is inflammation of the trabecular meshwork (trabeculitis) or blockage of the trabecular meshwork by inflammatory cells and byproducts. In uveitic closed angle glaucoma, adhesion of the iris to the angle structures (synechia) prevents drainage of the aqueous. Treatment involves the use of anti-inflammatory drugs and appropriate measures to lower eye pressure.

Neovascular Glaucoma

In patients with diabetes mellitus, vascular abnormalities result in poor blood supply to the retina. Ischemia triggers the growth of abnormal blood vessels (neovascularization) in the retina as well as in the anterior chamber angle. These abnormal vessels effectively block the drainage channels in the eye. Panretinal photocoagulation for diabetic retinopathy as well as glaucoma surgery is essential for treatment. Aside from diabetes, central retinal blood vessel occlusion can also cause neovascular glaucoma.

Traumatic Glaucoma

Any injury to the eye can lead to traumatic glaucoma. In the acute phase, red blood cells in the anterior chamber (hyphema) can clog up the angle and raise the intraocular pressure. In other cases, blunt trauma can rip the trabecular meshwork as seen in angle-recession glaucoma. This type of glaucoma can occur from weeks to even months after the injury due to scarring of the meshwork.

Lens-Induced Glaucoma

A complication of untreated cataract is lens-induced glaucoma. A large cataract (intumescent lens) can cause pupillary block and angle closure (phacomorphic glaucoma).

Treatment should be immediate and consists of medical treatment to control IOP by I.V. mannitol, systemic acetazolamide and topical betablockers.

Cataract extraction with implantation of PCIOL (which is the main treatment of phacomorphic glaucoma) should be performed.

A leaky cataract can cause inflammation in the anterior chamber and increased eye pressure (phacolytic glaucoma). In addition to adequate pressure control, cataract surgery has to be performed.

Steroid-Induced Glaucoma

Long-term use of steroids whether as eye drops, eye ointments, injections or oral medications can trigger a rise in intraocular pressure. It is believed that a cascade of reactions leads to increased resistance to aqueous outflow in the trabecular meshwork. Some individuals are more susceptible to pressure spikes brought about by steroids than others.

Pigmentary Glaucoma

Pigmentary glaucoma is a type of secondary glaucoma more commonly found in young adult males. Pigment granules from the iris are dispersed into the anterior chamber eventually blocking the trabecular meshwork.

Treatment is like that of primary open angle glaucoma.

Pseudoexfoliation Syndrome (PES)

Pseudoexfoliation syndrome (PES) is characterized by deposition of an amorphous grey dandruff-like material on the pupillary border, anterior lens surface, and posterior surface of iris, zonules and ciliary processes. The exact source of the exfoliate material is still not known.

The condition is associated with secondary open-angle glaucoma in about 50 % of the cases. Exact mechanism of rise of IOP is also not clear. Trabecular blockage by the exfoliate material is considered as the probable cause. Clinically the glaucoma behaves like POAG and is thus managed on the same lines.

Glaucoma Associated with Intraocular Tumours

Secondary glaucoma due to intraocular tumours such as malignant melanoma (of iris, choroid, ciliary body) and retinoblastoma may occur by one or more of the following mechanisms:

– trabecular block due to clogging by tumour cells or direct invasion by tumour seedlings;

– neovascularization of the angle;

– venous stasis following obstruction of the vortex veins;

– angle closure due to forward displacement of iris-lens diaphragm by increasing tumour mass.

Treatment: enucleation of the eyeball should be carried out as early as possible.

Glaucoma Examination

It includes:

– visual acuity is likely to be normal except in advanced glaucoma;

– pupils: to exclude a relative afferent pupillary defect (RAPD); if absent then subsequently develops, it indicates substantial progression;

– colour vision assessment such as Ishihara chart testing to check if there is any suggestion of an optic neuropathy other than glaucoma;

– slit-lamp examination: to exclude features of secondary glaucoma such as pigmentary and pseudoexfoliative;

– tonometry, prior to pachymetry, noting the time of day;

– pachymetry for CCT;

– gonioscopy;

– optic disc examination should always be performed with the pupils dilated, provided gonioscopy does not show critically narrow angles; red-free light can be used to detect RNFL defects;

– perimetry should usually be performed prior to clinical examination;

– optic disc or peripapillary RNFL imaging as described above.

Glaucoma Tratment

Glaucoma surgery

The types of glaucoma surgery include:

– trabeculotomy – similar to a trabeculectomy, but an electric current is used to remove a small part of the eye-drainage tubes;

– viscocanalostomy – part of the white outer covering of the eyeball (the sclera) is removed so fluid can drain from the eye more easily;

– deep sclerectomy – the drainage tubes in the eye are widened, sometimes by implanting a tiny device inside them;

– trabecular stent bypass – a tiny tube is placed into the eye to increase the drainage of fluid.

After surgery, the eye might water and be red, and vision may be slightly blurred for up to 6 weeks but should return to normal.

The hospital will advice about which activities you can do while you recover. Most people are advised to keep their eye dry, and avoid driving, reading and heavy lifting for at least a week.

Laser treatment

Laser treatment may be recommended if eye drops do not improve symptoms.

This is where a high-energy beam of light is carefully aimed at part of the eye to stop fluid from building up inside it.

Types of laser treatment include:

– laser trabeculoplasty – a laser is used to open up the drainage tubes within the eye, which allows more fluid to drain out and reduces the pressure inside;

– cyclodiode laser treatment – a laser is used to destroy some of the eye tissue that produces the liquid, which can reduce pressure in the eye;

– laser iridotomy – a laser is used to create holes in your iris to allow fluid to drain from the eye.

Laser treatment is usually carried out while a patient is awake. Local anaesthetic drops are used to numb the eyes – a patient may just feel a brief twinge of pain or heat during the procedure.

The patient may still need to use eye drops after having laser treatment.

Eye Drops

Glaucoma treatment often starts with prescription eye drops. These can help decrease eye pressure by improving how

fluid drains from the eye or by decreasing the amount of fluid the eye makes.

Prescription of eye drop medications includes:

1. *Prostaglandins.* These increase the outflow of the fluid in the eye (aqueous humor) and reduce pressure in the eye. Examples include latanoprost (Xalatan, Lanotan), travaprost (Travatan) and bimatoprost (Lumigan). Possible side effects include mild reddening and stinging of the eyes, darkening of the iris, changes in the pigment of the eyelashes or eyelid skin, and blurred vision.

2. *Beta blockers.* These reduce the production of fluid in the eye, thereby lowering the pressure in the eye (intraocular pressure). Examples include timolol (Betimol, Timoptic) and betaxolol (Betoptic). Possible side effects include difficulty breathing, slowed heart rate, lower blood pressure, impotence and fatigue.

3. *Alpha-adrenergic agonists.* These reduce the production of aqueous humor and increase outflow of the fluid in the eye. Examples include apraclonidine (Iopidine) and brimonidin. Possible side effects include an irregular heart rate; high blood pressure; fatigue; red, itchy or swollen eyes; dry mouth.

4. *Carbonic anhydrase inhibitors.* Rarely used for glaucoma, these drugs may reduce the production of fluid in the eye. Examples include dorzolamide (Trusopt) and brinzolamide (Azopt). Possible side effects include metallic taste, frequent urination, and tingling in the fingers and toes.

5. *Miotic or cholinergic agents.* These increase the outflow of fluid from the eye. An example is pilocarpine. Side effects include smaller pupils, possible blurred or dim vision, and nearsightedness.

TOPIC 11. DISEASES OF THE LENS

Cataract

The crystalline lens is a transparent structure. Its transparency may be disturbed due to degenerative process leading to opacification of lens fibres. Development of opacity in the lens is known as *cataract*.

Causes

There are several underlying causes of cataracts. These include:

- ultraviolet radiation from sunlight and other sources;
- diabetes;
- hypertension;
- obesity;
- smoking;
- prolonged use of corticosteroid medications;
- statin medicines used to reduce cholesterol;
- previous eye injury or inflammation;
- previous eye surgery;
- hormone replacement therapy;
- significant alcohol consumption;
- high myopia;
- family history.

Classification

Etiological classification:

I. *Congenital and developmental cataract.*

II. *Acquired cataract:*

1. Senile cataract.
2. Traumatic cataract.
3. Complicated cataract.
4. Metabolic cataract.
5. Electric cataract.
6. Radiational cataract.
7. Toxic cataract:

– corticosteroid-induced cataract;
– miotics-induced cataract;
– copper (in chalcosis) and iron (in siderosis) induced cataract.

8. Cataract associated with skin diseases (dermatogenic cataract).

9. Cataract associated with osseous diseases.

10. Cataract with miscellaneous syndromes:
– Steinert's disease (dystrophia myotonica);
– Down's syndrome;
– Lowe's syndrome;
– Treacher Collins syndrome.

Morphological classification:

1. Capsular cataract. It involves the capsule and may be:
– anterior capsular cataract;
– posterior capsular cataract.

2. Subcapsular cataract. It involves the superficial part of the cortex (just below the capsule) and includes:
– anterior subcapsular cataract;
– posterior subcapsular cataract.

3. Cortical cataract. It involves the major part of the cortex.

4. Supranuclear cataract. It involves only the deeper parts of cortex (just outside the nucleus).

5. Nuclear cataract. It involves the nucleus of the crystalline lens.

6. Polar cataract. It involves the capsule and superficial part of the cortex in the polar region only and may be:
– anterior polar cataract;
– posterior polar cataract.

Signs and symptoms of cataracts include:
– clouded, blurred or dim vision;
– increasing difficulty with vision at night;
– sensitivity to light and glare;

– need for brighter light for reading and other activities
– seeing “halos” around lights;
– frequent changes in eyeglass or contact lens prescription;
– fading or yellowing of colours;
– double vision in a single eye.

Congenital Cataract

These occur due to some disturbance in the normal growth of the lens. When the disturbance occurs before birth, the child is born with a congenital cataract. Therefore, in congenital cataract the opacity is limited to either embryonic or foetal nucleus. Developmental cataract may occur from infancy to adolescence. Therefore, such opacities may involve infantile or adult nucleus, deeper parts of cortex or capsule. Developmental cataract typically affects the particular zone which is being formed when this process is disturbed. The fibres laid down previously and subsequently are often normally formed and remain clear. Congenital and developmental opacities assume most variegated appearance and minute opacities (without visual disturbance) are very common in normal population. These are detected with the beam of slit lamp under full mydriasis.

Differential diagnosis

Congenital cataracts presenting with leukocoria need to be differentiated from various other conditions presenting with leukocoria such as retinoblastoma, retinopathy of prematurity, persistent hyperplastic primary vitreous (PHPV).

Acquired Cataract

We have studied that congenital and developmental cataracts occur due to disturbance in the formation of the lens fibres, i. e. instead of clear, opaque lens fibres are produced. While, in acquired cataract, opacification occurs due to

degeneration of the already formed normal fibres. The exact mechanism and reasons for the degeneration of lens fibres are not yet clear. However, in general, any factor (physical, chemical or biological) which disturbs the critical intra and extracellular equilibrium of water and electrolytes or deranges the colloid system within the lens fibres, tends to bring about opacification. The factors responsible for disturbing such equilibrium of the lens fibres vary in different types of acquired cataracts and shall be discussed with the individual type. A few common varieties of acquired cataract are described here.

Senile Cataract

Also called as "age-related cataract", this is the commonest type of acquired cataract affecting equally persons of either sex usually above the age of 50 years. By the age of 70 years, over 90 % of the individuals develop senile cataract. The condition is usually bilateral, but almost always one eye is affected earlier than the other.

Etiology

Senile cataract is essentially an ageing process. Though its precise etiopathogenesis is not clear, the various implicated factors affecting age of onset, type and maturation of senile cataract are as follows:

1. *Heredity*. It plays a considerable role in the incidence, age of onset and maturation of senile cataract in different families.

2. *Ultraviolet irradiations*.

3. *Dietary factors*. Diet deficient in certain proteins, amino acids, vitamins (riboflavin, vitamin E, vitamin C), and essential elements have also been blamed for early onset and maturation of senile cataract.

4. *Dehydrational crisis*.

5. *Smoking* has also been reported to have some effect on the age of onset of senile cataract

Causes of presenile cataract

The term *presenile cataract* is used when the cataractous changes similar to senile cataract occur before 50 years of age. Its common causes are:

1. *Heredity*.

2. *Diabetes mellitus*. Age-related cataract occurs earlier in diabetics. Nuclear cataract is more common and tends to progress rapidly.

3. *Myotonic dystrophy* is associated with posterior subcapsular type of presenile cataract.

4. *Atopic dermatitis* may be associated with presenile cataract (atopic cataract) in 10 % of the cases.

Stages of Cataracts

Maturation of the cortical type of senile cataract

1. Stage of lamellar separation. The earliest senile change is demarcation of cortical fibres owing to their separation by fluid. This phenomenon of lamellar separation can be demonstrated by slit-lamp examination only. These changes are reversible.

2. Stage of incipient cataract. In this stage early detectable opacities with clear areas between them are seen. Two distinct types of senile cortical cataracts can be recognised at this stage:

– *cuneiform senile cortical cataract:* it is characterised by wedge-shaped opacities with clear areas in between. These extend from equator towards centre and in early stages can only be demonstrated after dilatation of the pupil;

– *cupuliform senile cortical cataract:* here a saucershaped opacity develops just below the capsule usually in the central part of posterior cortex (posterior subcapsular cataract), which gradually extends outwards.

3. Immature senile cataract (ISC). In this stage, opacification progresses further. The cuneiform or cupuliform

patterns can be recognised till the advanced stage of ISC when opacification becomes more diffuse and irregular. The lens appears greyish white but clear cortex is still present and so iris shadow is visible.

4. Mature senile cataract (MSC). In this stage, opacification becomes complete, i. e. whole of the cortex is involved. Lens becomes pearly white in colour. Such a cataract is also labelled as "ripe cataract".

5. Hypermature senile cataract (HMSC). When the mature cataract is left in situ, the stage of hypermaturity sets in. The hypermature cataract may occur in any of the two forms:

– *Morgagnian hyper mature cataract:* in some patients, after maturity the whole cortex liquefies and the lens is converted into a bag of milky fluid. The small brownish nucleus settles at the bottom, altering its position with change in the position of the head. Such a cataract is called *Morgagnian cataract*.

– *Sclerotic type hyper mature cataract:* sometimes after the stage of maturity, the cortex becomes disintegrated and the lens becomes shrunken due to leakage of water. The anterior capsule is wrinkled and thickened due to proliferation of anterior cells and a dense white capsular cataract may be formed in the pupillary area. Due to shrinkage of lens, anterior chamber becomes deep and iris becomes tremulous (iridodonesis).

Maturation of nuclear senile cataract

In it, the sclerotic process renders the lens inelastic and hard, decreases its ability to accommodate and obstructs the light rays. These changes begin centrally and slowly spread peripherally almost up to the capsule when it becomes mature; however, a very thin layer of clear cortex may remain unaffected.

The nucleus may become diffusely cloudy (greyish) or tinted (yellow to black) due to deposition of pigments. In

practice, the commonly observed pigmented nuclear cataracts are either amber, brown (cataracta brunescens) or black (cataracta nigra) and rarely reddish (cataracta rubra) in colour.

Metabolic cataracts

These cataracts occur due to endocrine disorders and biochemical abnormalities. A few common varieties of metabolic cataracts are described here.

Diabetic cataract

Diabetes is associated with two types of cataracts:

1. Senile cataract in diabetics appears at an early age and progresses rapidly.

2. True diabetic cataract. It is also called "snow flake cataract" or "snow-storm cataract". It is a rare condition, usually occurring in young adults due to osmotic over-hydration of the lens. Initially a large number of fluid vacuoles appear underneath the anterior and posterior capsules, which is soon followed by appearance of bilateral snowflake-like white opacities in the cortex.

Galactosaemic cataract

It is associated with inborn error of galactose metabolism. Galactosaemia occurs in two forms:

1. Classical galactosaemia occurs due to deficiency of galactose-1 phosphate uridyl-transferase (GPUT).

2. A related disorder occurs due to deficiency of galactokinase (GK).

Characterstic features

Galactosaemia is frequently associated with the development of bilateral cataract (oil droplet central lens opacities). The lens changes may be reversible and occurrence of cataract may be prevented, if milk and milk products are eliminated from the diet when diagnosed at an early stage.

Hypocalcaemic cataract

Cataractous changes may be associated with parathyroid tetany, which may occur due to atrophy or

inadvertent removal of parathyroid glands (during thyroidectomy). Multicoloured crystals or small discrete white flecks of opacities are formed in the cortex which is seldom mature.

Cataract in Lowe's syndrome

Lowe's (Oculo-cerebral-renal) syndrome is a rare inborn error of amino acid metabolism.

Ocular features include congenital cataract and glaucoma.

Systemic features of this syndrome are mental retardation, dwarfism, osteomalacia, muscular hypotonia and frontal prominence.

Complicated cataract

It refers to opacification of the lens secondary to some other intraocular disease. Some authors use the term secondary cataract for the complicated cataract. Many authors use the term secondary cataract to denote aftercataract. Therefore, to avoid confusion and controversy, preferably, the term secondary cataract should be discarded.

Etiology

The lens depends for its nutrition on intraocular fluids. Therefore, any condition in which the ocular circulation is disturbed or in which inflammatory toxins are formed, will disturb nutrition of the crystalline lens, resulting in development of complicated cataract. Some important ocular conditions giving rise to complicated cataract are listed here.

1. Inflammatory conditions. These include uveal inflammations (like iridocyclitis, parsplanitis, choroiditis), hypopyon corneal ulcer and endophthalmitis.

2. Degenerative conditions such as retinitis pigmentosa and other pigmentary retinal dystrophies and myopic chorioretinal degeneration.

3. Retinal detachment. Complicated cataract may occur in long-standing cases.

4. Glaucoma (primary or secondary) may sometimes result in complicated cataract. The underlying cause here is probably the embarrassment to the intraocular circulation, consequent to the raised pressure.

5. Intraocular tumours such as retinoblastoma or melanoma may give rise to complicated cataract in late stages.

Complications

1. Phacoanaphylactic uveitis. A hypermature cataract may leak lens proteins into anterior chamber. These proteins may act as antigens and induce antigen antibody reaction leading to uveitis.

2. Lens-induced glaucoma. It may occur by different mechanisms, e. g., due to intumescent lens (phacomorphic glaucoma) and leakage of proteins into the anterior chamber from a hypermature cataract (phacolytic glaucoma).

3. Subluxation or dislocation of lens. It may occur due to degeneration of zonules in hyper mature stage.

Cataract examination

1. Visual acuity.
2. Colour vision.
3. Slit-lamp examination.
4. Tonometry.
5. Ophtalmoscopy.
6. Perimetry should usually be performed prior to clinical examination.

Treatment

Medications should be assessed to see which ones might be photosensitizers, and steroid eye drops should be used for a period as brief as possible.

At the age of 50 or older, it is important to have a comprehensive eye exam at least once every two years. In addition to cataracts, the eye doctor can evaluate for age-related macular degeneration, glaucoma, and other vision problems. Early detection and treatment is the key.

Types and choice of surgical techniques: *cataract extraction and lens implant procedures*

Cataract extraction is usually done using a topical or local anesthetic and IV sedation. There are 3 extraction techniques:

1. In intracapsular cataract extraction, the cataract and lens capsule are removed in one piece; this technique is rarely used.

2. In extracapsular cataract extraction, the hard central nucleus is removed in one piece and then the soft cortex is removed in multiple small pieces.

3. In phacoemulsification, the hard central nucleus is dissolved by ultrasound and then the soft cortex is removed in multiple small pieces.

TOPIC 12. DISEASES OF THE OPTIC NERVE

1. Optic neuritis.
2. Anterior ischaemic optic neuropathy.
3. Papilloedema.
4. Optic atrophy.
5. Congenital anomalies.
6. Tumours.

Optic Neuritis

Optic neuritis includes inflammatory and demyelinating disorders of the optic nerve.

Etiology

1. Idiopathic. In a large proportion of cases the underlying cause is unidentifiable.

2. Hereditary optic neuritis (Leber's disease).

3. Demyelinating disorders are by far the most common cause of optic neuritis. These include multiple sclerosis, neuromyelitis optica (Devic's disease) and diffuse periaxial encephalitis of Schilder. About 70 % cases of established multiple sclerosis may develop optic neuritis.

4. Parainfectious optic neuritis is associated with various viral infections such as measles, mumps, chickenpox, whooping cough and glandular fever. It may also occur following immunization.

5. Infectious optic neuritis may be sinus related (with acute ethmoiditis) or associated with cat scratch fever, syphilis (during primary or secondary stage), Lyme disease and cryptococcal meningitis in patients with AIDS.

6. Toxic optic neuritis (see toxic amblyopias).

Anatomical types

Optic neuritis can be classified into three anatomical types:

1. *Papillitis*. It refers to involvement of the optic disc in inflammatory and demyelinating disorders. This condition is usually unilateral but sometimes may be bilateral.

2. *Neuroretinitis refers* to combined involvement of optic disc and surrounding retina in the macular area.

3. *Retrobulbar neuritis* is characterized by involvement of optic nerve behind the eyeball. Clinical features of acute retrobulbar neuritis are essentially similar to that of acute papillitis except for the fundus changes and ocular changes described below.

Symptoms

Optic neuritis may be asymptomatic or may be associated with the following symptoms:

1. *Visual loss.* Sudden, progressive and profound visual loss is the hallmark of acute optic neuritis.

2. *Dark adaptation* may be lowered.

3. *Visual obscuration* in bright light is a typical symptom of acute optic neuritis.

4. *Impairment of colour vision* is always present in optic neuritis. Typically the patients observe reduced vividness of saturated colours.

5. *Movement phosphenes* and sound induced phosphenes may be percieved by patients with optic neuritis. Phosphenes refer to glowing sensations produced by no photic or the so called inadequate stimuli.

6. *Episodic transient obscuration of vision* on exertion and on exposure to heat, which recovers on resting or moving away from the heat (Uhthoff's symptom), occurs in patient with isolated optic neuritis.

7. *Depth perception*, particularly for the moving object may be impaired (Pulfrich's phenomenon).

8. *Pain.* Patient may complains of mild dull eyeache. It is more marked in patients with retrobulbar neuritis than with papillitis. Pain is usually aggravated by ocular movements,

especially in upward or downward directions due to attachment of some fibres of superior rectus to the dura mater.

Signs

1. Visual acuity is usually reduced markedly.

2. Colour vision is often severely impaired.

3. Pupil shows ill-sustained constriction to light. Marcus Gunn pupil which indicates relative afferent pupillary defect (RAPD) is a diagnostic sign. It is detected by the swinging flash light test.

4. Ophthalmoscopic features. Papillitis is characterised by hyperaemia of the disc and blurring of the margins. Disc becomes edematous and physiological cup is obliterated. Retinal veins are congested and tortuous. Splinter haemorrhages and fine exudates may be seen on the disc. Slit-lamp examination may reveal inflammatory cells in the vitreous. Inflammatory signs may also be present in the surrounding retina when papillitis is associated with macular star formation and the condition is labelled as "neuroretinitis".

In majority of the cases with retrobulbar neuritis fundus appears normal and the condition is typically defined as a disease where neither the ophthalmologist nor the patient sees anything. Occasionally temporal pallor of the disc may be seen.

5. Visual field changes. The most common field defect in optic neuritis is a relative central or centrocecal scotomata. Other field defects noted rarely include: paracentral nerve fibre bundle defect, a nerve fibre bundle defect extending up to periphery and a nerve fibre bundle defect involving fixation point and periphery. The field defects are more marked to red colour than the white.

6. Contrast sensitivity is impaired.

7. Visually evoked response (VER) shows reduced amplitude and delay in the transmission time.

Treatment

Corticosteroid therapy may shorten the period of visual loss, but will not influence the ultimate level of visual recovery in patients with optic neuritis.

Optic Disc Swelling (Papilledema)

The terms *papilledema* and *disc edema* look alike and per se mean swelling of the optic disc. However, arbitrarily the term "papilledema" has been reserved for the passive disc swelling associated with increased intracranial pressure which is almost always bilateral although it may be asymmetrical. The term "disc edema or disc swelling" includes all causes of active or passive edematous swelling of the optic disc.

Causes of disc edema

Congenital anomalous elevation (pseudopapilledema):

1. *Inflammations:*
– papillitis;
– neuroretinitis.
2. *Ocular diseases:*
– uveitis;
– hypotony;
– vein occlusion.
3. *Orbital causes:*
– tumours;
– Graves' orbitopathy;
– orbital cellulitis.
4. *Vascular causes:*
– anaemia;
– uremia;
– anterior ischaemic optic neuropathy.
5. *Increased intracranial pressure:*
– see causes of papilloedema.

As discussed above, papilledema is secondary to raised intracranial pressure which may be associated with the following conditions:

1. Congenital conditions include aqueductal stenosis and craniosynostosis.

2. Intracranial space-occupying lesions (ICSOLs). These include brain tumours, abscess, tuberculoma, gumma, subdural haematoma and aneurysms. The ICSOLs in any position excepting medulla oblongata may induce papilledema. Papilledema is most frequently associated with tumours arising in posterior fossa, which obstruct aqueduct of Sylvius and least with pituitary tumours. Thus, the ICSOLs of cerebellum, midbrain and parieto-occipital region produce papilledema more rapidly than the mass lesions of other areas. Further, the fast progressing lesions produce papilledema more frequently and acutely than the slow growing lesions.

3. Intracranial infections such as meningitis and encephalitis may be associated with papilledema.

4. Intracranial hemorrhages. Cerebral as well as subarachnoid hemorrhage can give rise to papilledema which is frequent and considerable in extent.

5. Obstruction of CSF absorption via arachnoid villi which have been damaged previously.

6. Tumours of spinal cord occasionally give rise to papilledema.

7. Idiopathic intracranial hypertension (IIH) also known as pseudo tumour cerebri, is an important cause of raised intracranial pressure. It is a poorly understood condition, usually found in young obese women. It is characterized by chronic headache and bilateral papilledema without any ICSOLs or enlargement of the ventricles due to hydrocephalus.

8. Systemic conditions include malignant hypertension, pregnancy induced hypertension (PIH) cardiopulmonary insufficiency, blood dyscrasias and nephritis.

9. Diffuse cerebral edema from blunt head trauma may causes papilledema.

Clinical features

General features. Patients usually present to general physicians with general features of raised intracranial pressure. These include headache, nausea, projectile vomiting and diplopia. Focal neurological deficit may be associated.

Ocular features. Patients may give history of recurrent attacks of transient blackout of vision (amaurosis fugax). Visual acuity and pupillary reactions usually remain fairly normal until the late stages of diseases when optic atrophy sets in. Clinical features of papilledema can be described under four stages: *early*, *fully developed*, *chronic* and *atrophic*.

1. Early (incipient) papilledema

Symptoms are usually absent and visual acuity is normal; pupillary reactions are normal. Ophthalmoscopy features of early papilledema are:

– obscuration of the disc margins (nasal margins are involved first followed by the superior, inferior and temporal);

– blurring of peripapillary nerve fiber layer;

– absence of spontaneous venous pulsation at the disc (appreciate in 80 % of the normal individuals);

– mild hyperemia of the disc;

– splinter hemorrhages in the peripapillary region may be present;

– visual fields are fairly normal.

2. Established (fully developed) papilledema

Symptoms

Patient may give history of transient visual obscurations in one or both eyes, lasting a few seconds, after standing. Visual acuity is usually normal. Pupillary reactions remain fairly normal. Ophthalmoscopy features:

– apparent optic disc edema is seen as its forward elevation above the plane of retina; usually up to 1–2 mm (1 mm elevation is equivalent to +3 diopters);

– physiological cup of the optic disc is obliterated;

– disc becomes markedly hyperemic and blurring of the margin is present all-around;

– multiple soft exudates and superficial hemorrhages may be seen near the disc;

– veins become tortuous and engorged;

– in advanced cases, the disc appears to be enlarged and circumferential greyish white folds may develop due to separation of nerve fibers by the edema;

– rarely, hard exudates may radiate from the fovea in the form of an incomplete star;

– visual fields show enlargement of blind spot.

3. Chronic or long standing (vintage) papilledema

Symptoms:

– visual acuity is variably reduced depending upon the duration of the papilledema;

– pupillary reactions are usually normal;

– ophthalmoscopy features: in this stage, acute hemorrhages and exudates resolve, and peripapillary edema is resorbed; the optic disc gives appearance of the dome of a champagne cork; the central cup remains obliterated; small druse like crystalline deposits (corpora amylase) may appear on the disc surface;

– visual fields: blind spot is enlarged and the visual fields begin to constrict.

4. Atrophic papilledema

Symptoms:

– atrophic papilledema develops after 6–9 months of chronic papilledema and is characterized by severely impaired visual acuity;

– pupillary reaction: light reflex is impaired;

– ophthalmoscopy features: it is characterized by greyish white discolouration and pallor of the disc due to atrophy of the neurons and associated gliosis. Prominence of the disc decreases in spite of persistent raised intracranial pressure. Retinal arterioles are narrowed and veins become less congested. Whitish sheathing develops around the vessels.

Treatment and prognosis

It is a neurological emergency and requires immediate hospitalization. As a rule, unless the causative disease is treatable or cerebral decompression is done, the course of papilledema is chronic and ultimate visual prognosis is bad.

Optic Atrophy

It refers to degeneration of the optic nerve, which occurs as an end result of any pathologic process that damages axons in the anterior visual system, i. e. from retinal ganglion cells to the lateral geniculate body.

Classification

A. Primary versus secondary optic atrophy. It is customary to divide the optic atrophy into primary and secondary.

1. *Primary optic atrophy* refers to the simple degeneration of the nerve fibers without any complicating process within the eye, e. g., syphilitic optic atrophy of tabs dorsalis.

2. *Secondary optic atrophy* occurs following any pathologic process which produces optic neuritis or papilledema.

B. Ophthalmoscopy classification. It is more useful to classify optic atrophy based on its ophthalmoscopy appearance.

Common types are as follows:

1. Primary (simple) optic atrophy.
2. Consecutive optic atrophy.
3. Glaucomatous optic atrophy.

4. Post-neurotic optic atrophy.

5. Vascular (ischemic) optic atrophy. The etiology and salient features of each type will be considered separately.

C. Ascending versus descending optic atrophy.

1. *Ascending optic atrophy* follows damage to ganglion cells or nerve fiber layer due to disease of the retina or optic disc. In it the nerve fiber degeneration progresses (ascends) from the eyeball towards the geniculate body.

2. *Descending* or *retrograde optic atrophy* proceeds from the region of the optic tract, chiasma or posterior portion of the optic nerve towards the optic disc.

Clinical features of optic atrophy

1. Loss of vision, may be of sudden or gradual onset (depending upon the cause of optic atrophy) and partial or total (depending upon the degree of atrophy). It is important to note that ophthalmoscopy signs cannot be correlated with the amount of vision.

2. Pupil is semi dilated and *direct light reflex* is very sluggish or absent. Swinging flash light test depicts Marcus Gunn pupil.

3. Visual field loss will vary with the distribution of the fibers that have been damaged. In general, the field loss is peripheral in systemic infections, central in focal optic neuritis and eccentric when the nerve or tracts are compressed.

4. Ophthalmoscopy appearance of the disc will vary with the type of optic atrophy. However, ophthalmoscopy features of optic atrophy in general are pallor of the disc and decrease in the number of small blood vessels (Kastenbaum index). The pallor is not due to atrophy of the nerve fibers but to loss of vasculature.

Ophthalmoscopy features of different types of optic atrophy are as described below:

1. *Primary optic atrophy*. Colour of the disc is chalky white or white with bluish hue. Its edges (margins) are sharply

outlined. Slight recession of the entire optic disc occurs in total atrophy. Lamina cribrosa is clearly seen at the bottom of the physiological cup. Major retinal vessels and surrounding retina are normal.

2. *Consecutive optic atrophy*. Disc appears yellow waxy. Its edges are not so sharply defined as in primary optic atrophy. Retinal vessels are attenuated.

3. *Post-neuritic optic atrophy*. Optic disc looks dirty white in colour. Due to gliosis its edges are blurred, physiological cup is obliterated and lamina cribrosa is not visible. Retinal vessels are attenuated and perivascular sheathing is often present.

4. *Glaucomatous optic atrophy*. It is characterised by deep and wide cupping of the optic disc and nasal shift of the blood vessels.

5. *Ischaemic optic atrophy*. Ophthalmoscopic features are pallor of the optic disc associated with marked attenuation of the vessels.

Treatment

The underlying cause when treated may help in preserving some vision in patients with partial optic atrophy. However, once complete atrophy has set in, the vision cannot be recovered.

Toxic Amblyopias

These include those conditions wherein visual loss results from damage to the optic nerve fibers due to the effects of exogenous (commonly) or endogenous (rarely) poisons. A few common varieties of toxic amblyopia are described here.

Tobacco amblyopia

It typically occurs in men who are generally pipe smokers, heavy drinkers and have a diet deficient in proteins and vitamin B complex; and thence also labelled as “tobacco-alcohol-amblyopia”.

Pathogenesis

The toxic agent involved is cyanide found in tobacco.

Clinical features

The condition usually occurs in men between 40 and 60 years and is characterized by bilateral gradually progressive impairment in the central vision. Patients usually complain of fogginess and difficulty in doing near work. Visual field examination reveals bilateral centrocecal scotomata with diffuse margins which are not easily defined. The defect is greater for red than the white colour. Fundus examination is essentially normal or there may be slight temporal pallor of the disc.

Treatment

It consists of complete cessation of tobacco and alcohol consumption, hydroxycobalamine 1000 µg intramuscular injections weekly for 10 weeks and care of general health and nutrition. Vasodilators have also been tried.

Prognosis

It is good, if complete abstinence from tobacco and alcohol is maintained. Visual recovery is slow and may take several weeks to months.

Ethyl alcohol amblyopia

It usually occurs in association with tobacco amblyopia. However, it may also occur in non-smokers, who are heavy drinkers suffering from chronic gastritis. The optic neuritis occurs along with the peripheral neuritis of chronic and debilitated alcoholics.

Clinical picture and treatment is similar to tobacco amblyopia, but the prognosis is not so good.

Methyl alcohol amblyopia

Unlike ethyl alcohol (which produces chronic amblyopia), poisoning by methyl alcohol (methanol) is typically acute, usually resulting in optic atrophy and permanent blindness.

Etiology

It usually occurs due to intake of wood alcohol or methylated spirit in cheap adulterated or fortified beverages. Sometimes, it may also be absorbed by inhalation of fumes in industries, where methyl alcohol is used as a solvent. Rarely, it may also be absorbed from the skin following prolonged daily use of liniments.

General symptoms of acute poisoning are headache, dizziness, nausea, vomiting, abdominal pain, delirium, stupor and even death. Presence of a characteristic odor due to excretion of formaldehyde in the breath or sweat is a helpful diagnostic sign.

Ocular features

Patients are usually brought with almost complete blindness, which is noticed after 2–3 days, when stupor weans off. Fundus examination in early cases reveals mild disc edema and markedly narrowed blood vessels. Finally, bilateral primary optic atrophy ensues.

Treatment

1. Gastric lavage to wash away the methyl alcohol should be carried out immediately and at intervals during the first few days, as the alcohol in the system is continuously returned to stomach.

2. Administration of alkali to overcome acidosis should be done in early stages. Soda bicarb may be given orally or intravenously (500 ml of 5 % solution).

3. Ethyl alcohol. It should also be given in early stages. It competes with the methyl alcohol for the enzyme alcohol dehydrogenase, thus preventing the oxidation of methanol to formaldehyde. It should be given in small frequent doses, 90 cc every 3 hours for 3 days.

4. Eliminative treatment by diaphoresis in the form of peritoneal dialysis is also helpful by washing the alcohol and formaldehyde from the system.

5. Prognosis is usually poor; death may occur due to acute poisoning. Blindness often occurs in those who survive.

Quinine amblyopia

It may occur even with small doses of the drug in susceptible individuals.

Clinical features

Patient may develop near total blindness. Deafness and tinnitus may be associated. The pupils are fixed and dilated. Fundus examination reveals retinal edema, marked pallor of the disc and extreme attenuation of retinal vessels. Visual fields are markedly contracted.

Anterior Ischaemic Optic Neuropathy (AION)

It refers to the segmental or generalized infarction of anterior part of the optic nerve.

Etiology

The AION results from occlusion of the short posterior ciliary arteries. Depending upon the etiology it may be typified as follows:

1. Idiopathic AION. It is the most common entity, thought to result from the atherosclerotic changes in the vessels.
2. Arteritic AION. It is the second common variety. It occurs in association with giant cell arteritis.
3. AION due to miscellaneous causes. It may be associated with severe anemia, collagen vascular disorders, following massive hemorrhage, papilledema, migraine and malignant hypertension.

Clinical features

Visual loss is usually marked and sudden. Fundus examination during acute stage may reveal segmental or diffuse edematous, pale or hyperemic disc, usually associated with splinter hemorrhages.

Visual fields show typical altitudinal hemianopia involving the inferior (commonly) or superior half.

Investigations: ESR and C-reactive protein levels are raised in patients with giant cell arteritis. Confirmation of the diagnosis may be done by temporal artery biopsy.

Treatment

Immediate treatment with heavy doses of corticosteroids (80 mg prednisolone daily) should be started and tapered by 10 mg weekly. Steroids in small doses (5 mg prednisolone) may have to be continued for a long time (3 months to one year).

Congenital and Developmental Disorders

Classification

1. Anomalies of the optic disc. These include crescents, situs in versus, congenital pigmentation, coloboma, druse and hypoplasia of the optic disc.

2. Anomalies of the nerve fibers, e. g., modulated (opaque) nerve fibers.

3. Anomalies of vascular elements, such as persistent hyaloid artery and congenital tortuosity of retinal vessels.

4. Anomalies of the retina proper. These include albinism, congenital night blindness, congenital day blindness, Oguchi disease, congenital retinal cyst, congenital retinal detachment and coloboma of the fundus.

5. Congenital anomalies of the macula are aplasia, hypoplasia and coloboma. A few important congenital disorders are described briefly.

Coloboma of the Optic Disc

It results from the failure in closure of the embryonic fissure. It occurs in two forms. The minor defect is more common and manifests as inferior crescent, usually in association with hypermetropic or astigmatic refractive error.

The actual optic disc is seen as a linear horizontal pinkish band confined to a small superior wedge. Defective vision and a superior visual field defect are usually associated.

Drusen of the Optic Disc

Drusen are intrapapillary refractile bodies, which usually lie deep beneath the surface of the disc tissue in childhood and emerge out by the early teens. Thus, in children they present as pseudopapilledema and by teens they can be recognized ophthalmoscopically as waxy pea-like irregular refractile bodies.

Hypoplasia of Optic Disc

Hypoplasia of the optic nerve may occur as an isolated anomaly or in association with other anomalies of the central nervous system. The condition is bilateral in 60 % of cases. It is associated with maternal alcohol use, diabetes and intake of certain drugs in pregnancy. It forms a significant cause of blindness at birth in developed countries.

Diagnosis of mild cases presents little difficulty. In typical cases the disc is small and surrounded by a yellowish and a pigmented ring, referred to as "double ring sign".

Symptomatic Disturbances of the Vision

Night Blindness (Nyctalopia)

Night (scotopic) vision is a function of rods. Therefore, the conditions in which functioning of these nerve endings is deranged will result in night blindness. These include vitamin A deficiency, tapetoretinal degenerations (e. g., retinitis pigmentosa), congenital high myopia, familial congenital night blindness and Oguchi disease. It may also develop in conditions of the ocular media interfering with the light rays in dim light (i. e. with dilated pupils). These include paracentral lenticular and corneal opacities. In advanced cases of primary

open angle glaucoma, dark adaptation may be so much delayed that patient gives history of night blindness.

Day Blindness (Hamarlopia)

It is a symptomatic disturbance of the vision, in which the patient is able to see better in dim light as compared to bright light of the day. Its causes are congenital deficiency of cones, central lenticular opacities (polar cataracts) and central corneal opacities.

Colour Blindness

An individual with normal colour vision is known as trichromate. In colour blindness, faculty to appreciate one or more primary colours is either defective (anomalous) or absent (anopia). It may be congenital or acquired.

Congenital colour blindness

It is an inherited condition affecting males more (3–4 %) than females (0.4 %). It may be of the following types:

1. Dyschromatopsia.
2. Achromatopsia.

Dyschromatopsia

Dyschromatopsia literally means colour confusion due to deficiency of mechanism to perceive colours. It can be classified into:

1. Anomalous trichromatism.
2. Dichromatism.

Anomalous trichromatic colour vision

Here, the mechanism to appreciate all the three primary colours is present, but is defective for one or two of them. It may be of following types:

1. *Protanomalous.* It refers to defective red colour appreciation.

2. *Deuteranomalous.* It means defective green colour appreciation.

3. *Tritanomalous.* It implies defective blue colour appreciation.

Dichromatic colour vision

In this condition faculty to perceive one of the three primary colours is completely absent. Such individuals are called dichromates and may have one of the following types of defects:

- *protanopia*, i. e. complete red colour defect;
- *deuteranopia*, i. e. complete defect for green colour;
- *tritanopia*, i. e. absence of blue colour appreciation.

Red-green deficiency (protanomalous, protanopia, deuteranomalous and deuteranope) is more common. Such a defect is a source of danger in certain occupations such as drivers, sailors and traffic police. Blue deficiency (tritanomalous and tritanopia) is comparatively rare.

Achromatopsia

It is an extremely rare condition presenting as cone monochromatism or rod monochromatism.

Cone monochromatism is characterised by presence of only one primary colour and thus the person is truly colour blind. Such patients usually have a visual acuity of 6/12 or better.

Rod monochromatism may be complete or incomplete. It is inherited as an autosomal recessive trait. It is characterized by:

- total colour blindness;
- day blindness (visual acuity is about 6/60);
- nystagmus;
- fundus is usually normal.

Acquired Colour Blindness

It may follow damage to macula or optic nerve. Usually, it is associated with a central scotoma or decreased visual acuity.

Blue-yellow impairment is seen in retinal lesions such as CSR, macular edema and shallow retinal detachment.

Red-green deficiency is seen in optic nerve lesions such as optic neuritis, Leber's optic atrophy and compression of the optic nerve.

Acquired blue colour defect (blue blindness) may occur in old age due to increased sclerosis of the crystalline lens. It is owing to the physical absorption of the blue rays by the increased amber coloured pigment in the nucleus.

Tests for Colour Vision

These tests are designed for:

1. Screening defective colour vision from normal.
2. Qualitative classification of colour blindness, i. e. protan, deuteran and tritan.
3. Quantitative analysis of degree of deficiency, i. e. mild, moderate or marked.

Amaurosis

It implies complete loss of sight in one or both eyes, in the absence of ophthalmoscopic or other marked objective signs.

Amaurosis fugax: it refers to a sudden, temporary and painless monocular visual loss occurring due to a transient failure of retinal circulation.

Common causes of amaurosis fugax are: carotid transient ischemic attacks (TIA), embolization of retinal circulation, papilledema, giant cell arteritis, Raynaud's disease, migraine, as a prodromal symptom of central retinal artery or carotid artery occlusion, hypertensive retinopathy, and venous stasis retinopathy. An attack of amaurosis fugax is typically

described by the patients as a curtain that descends from above or ascends from below to occupy the upper or lower halves of their visual fields.

Uremic amaurosis: it is a sudden, bilateral, complete loss of sight occurring probably due to the effect of certain toxic materials upon the cells of the visual centre in patients suffering from acute nephritis, eclampsia of pregnancy and renal failure. The visual loss is associated with dilated pupils which generally react to light. The fundi are usually normal except for the coincidental findings of hypertensive retinopathy, when associated. Usually, the vision recovers in 12–48 hours.

Amblyopia

It implies a partial loss of sight in one or both eyes, in the absence of ophthalmoscopy or other marked objective signs. It may be either congenital or acquired. Acquired amblyopia may be organic (toxic amblyopia) or functional. Functional amblyopia results from the psychical suppression of the retinal image. It may be anisometropia, strabismus or due to stimulus deprivation (amblyopia ex anopsia).

Cortical Blindness

Cortical blindness (visual cortex disease) is produced by bilateral occipital lobe lesions. Unilateral occipital lobe lesions typically produce contralateral macular sparing congruous homonymous hemianopia.

Ocular Manifestations of Diseases of the Central Nervous System

Ocular involvement in diseases of the central nervous system is not infrequent. A few common ocular lesions of these diseases are mentioned here.

Intracranial Infections

These include meningitis, encephalitis, brain abscess and neurosyphilis.

1. *Meningitis.* It may be complicated by papillitis, and paralysis of the third, fourth and sixth cranial nerves. Chronic chiasma arachnoiditis may produce bilateral optic atrophy. Tuberculous meningitis may be associated with choroidal tubercles.

2. Encephalitis. It may be complicated by papillitis and/or papilledema. Cranial nerve palsies are usually incomplete. Diplopia and ptosis are often present.

3. Brain abscess. It is frequently associated with papilledema. Focal signs depend upon the site of the abscess, and are thus similar to tumours.

4. *Neurosyphilis*. Ocular involvement is quite frequent.

Gummatous meningitis may be associated with papillitis, papilledema or post neurotic optic atrophy and cranial nerve palsies. (Third nerve is paralyzed in nearly 30 % cases, less frequently the fifth and sixth, and least frequently the fourth.) Tabs dorsalis and generalized paralysis of insane may be associated with primary optic atrophy, Argyll Robertson pupil, and internal and/or external ophthalmoplegia.

Intracranial Aneurysms

Intracranial aneurysms associated with ocular manifestations are located around the circle of Willis.

Intracranial Haemorrhages

Ophthalmic signs of intracerebral hemorrhage are tonic conjugate and dysconjugate deviations. Subarachnoid hemorrhage may produce retinal hemorrhages (especially sub hyaloid hemorrhage of the posterior pole), papilledema, and ocular palsies.

Intracranial Space-Occupying Lesions (ICSOL)

These include primary and secondary brain tumours, hematomas, granulomatous inflammations and parasitic cysts.

Demyelinating Diseases

These include multiple sclerosis, neuromyelitis optica and diffuse sclerosis. Ocular involvement may occur in all these conditions. Their salient features are as follows:

Multiple sclerosis: it is a demyelinating disorder of unknown etiology, affecting women more often than men, usually in the 15–50 years age group. Pathologically, the condition is characterized by a patchy destruction of the myelin sheaths throughout the central nervous system.

Clinical course of the condition is marked by remissions and relapses. In this condition, optic neuritis is usually unilateral.

Other ocular lesions include internuclear ophthalmoplegia and vestibular or cerebellar nystagmus.

Neuromyelitis optica (Devic's disease): it is characterized by bilateral optic neuritis associated with ascending myelitis, entailing a progressive quadriplegia and anaesthesia.

Unlike multiple sclerosis, this condition is not characterized by remissions and is not associated with ocular palsies and nystagmus.

Diffuse sclerosis (Schilder disease): it typically affects children and adolescents and is characterized by progressive demyelination of the entire white matter of the cerebral hemispheres. Ocular lesions include: optic neuritis (papillitis or retrobulbar neuritis), cortical blindness (due to destruction of the visual centres and optic radiations), ophthalmoplegia and nystagmus.

Idiopathic Intracranial Hypertension (IIH)

(Pseudotumour cerebri, benign intracranial hypertension)

Idiopathic intracranial hypertension (IIH) is known by the raised intracranial pressure in the absence of a mass lesion or of hydrocephalus. It is mostly related to impaired cerebrospinal fluid (CSF) absorption from the subarachnoid space. It is common in obese women of 20–40 years of age. The female to male ratio is between 3:1 and 8:1. Up to 90 % of patients are overweight. IIH may also be caused by drugs. Lumbar puncture (LP) is one of the treatment methods and is also useful for the diagnosis. If the opening pressure is >250 mm H_2O and LP shows no inflammatory cells with normal CSF protein and glucose levels, the diagnosis will be achieved. Cranial MRI shows normal ventricles and often an empty sellar. Venous sinus thrombosis should be excluded by the cranial MR venography.

Medical and lifestyle treatment: weight loss, Acetazolamide, Furosemide, Topiramate.

Surgical treatment: repeated lumbar puncture, optic nerve sheath decompressions, ventriculoperitoneal shunt placements, lumboperitoneal shunt placement.

Basic examination of the nervous optics disease:

- observation of external structures;
- extraocular movements and cranial nerves;
- dilating the pupil;
- visual acuity;
- slit-lamp examination;
- intraocular pressure (IOP);
- visual field (confrontation) testing;
- colour vision;
- ophthalmoscopy.

Other tests carried out in the ophthalmology department:

– visual field assessment;
– ultrasound;
– hess chart fluorescein angiography;
– optical coherence tomography (OCT);
– visually evoked potential (VEP), also called visually evoked response (VER) and visually evoked cortical potential (VECP);
– magnetic resonance imaging (MRI) scan.

TOPIC 13. DISEASES OF THE RETINA

Among the four leading causes of vision loss, two are retinal diseases – age-related macular degeneration (AMD), which affects people over age 50, and diabetic retinopathy, which affects people aged 40 and above.

Worldwide, the World Health Organization estimates 295 million people are visually impaired, 39 million of whom are blind. The National Institutes of Health, the National Eye Institute expect those numbers to double within the next 30 years.

Classification

1. Congenital and developmental disorders.
2. Traumatic lesions.
3. Inflammatory disorders:
   - *retinitis*;
   - *periphlebitis retinae*.
4. Vascular disorders:
   - *retinal artery occlusions*;
   - *retinal vein occlusions*;
   - *diabetic retinopathy*;
   - *hypertensive retinopathy*;
   - *sickle cell retinopathy*;
   - *retinopathy of prematurity*;
   - *retinal telengiectasia*;
   - *ocular ischaemic syndrome*.
5. Dystrophies and degenerations.
6. Macular disorders.
7. Retinal detachment.
8. Tumours.

Congenital and Developmental Disorders

Classification

1. Anomalies of the optic disc. These include crescents, situs inversus, congenital pigmentation, coloboma, drusen and hypoplasia of the optic disc.

2. Anomalies of the nerve fibres, e. g., medullated (opaque) nerve fibres.

3. Anomalies of vascular elements, such as persistent hyaloid artery and congenital tortuosity of retinal vessels.

4. Anomalies of the retina proper. These include albinism, congenital night blindness, congenital day blindness, Oguchi's disease, congenital retinal cyst, congenital retinal detachment and coloboma of the fundus.

5. Congenital anomalies of the macula are aplasia, hypoplasia and coloboma. A few important congenital disorders are described briefly.

Coloboma of the optic disc

It results from the failure in closure of the embryonic fissure. It occurs in two forms. The minor defect is more common and manifests as inferior crescent, usually in association with hypermetropic or astigmatic refractive error.

Drusen of the optic disc

Drusens are intrapapillary refractile bodies, which usually lie deep beneath the surface of the disc tissue in childhood and emerge out in the early teenagehood. Thus, in children they present as pseudo-papilledema and by teens they can be recognised ophthalmoscopically as waxy pea-like irregular refractile bodies.

Hypoplasia of the optic disc

Hypoplasia of the optic nerve may occur as an isolated anomaly or in association with other anomalies of the central nervous system. The condition is bilateral in 60 % of cases. It is associated with maternal alcohol use, diabetes and intake of

certain drugs in pregnancy. It forms a significant cause of blindness at birth in developed countries.

Inflammatory Disorders of the Retina

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

Retinitis

I. Non-specific retinitis.

It is caused by pyogenic organisms and may be either acute or subacute.

1. Acute purulent retinitis. It occurs as metastatic infection in patients with pyaemia. The infection usually involves the surrounding structures and soon converts into metastatic endophthalmitis or even panophthalmitis.

2. Subacute retinitis of Roth. It typically occurs in patients suffering from subacute bacterial endocarditis (SABE).

It is characterised by multiple superficial retinal haemorrhages, involving posterior part of the fundus. Most of the haemorrhages have a white spot in the centre (Roth's spots). Vision may be blurred due to involvement of the macular region or due to associated papillitis.

II. Specific retinitis.

It may be *bacterial* (tuberculosis, leprosy, syphilis and actinomycosis), *viral* (cytomegalic inclusion disease, rubella, herpes zoster), *mycotic*, *rickettsia* or *parasitic* in origin. Cytomegalovirus (CMV) retinitis, zoster retinitis, progressive outer retinal necrosis (PORN) caused by an aggressive variant of varicella zoster virus and acute retinal necrosis (ARN) caused by herpes simplex virus II (in patients under the age of 15 years) and by varicella zoster virus and herpes simplex

virus-I (in older individuals) have become more conspicuous in patients with AIDS (HIV infection).

Retinal Vasculitis

Inflammation of the retinal vessels may be primary (*Eales' disease*) or secondary to uveitis.

Eales' disease

It is an idiopathic inflammation of the peripheral retinal veins. It is characterised by recurrent vitreous haemorrhage, so it is also referred to as primary vitreous haemorrhage.

Etiology

It is not known exactly. Many workers consider it to be a hypersensitivity reaction to tubercular protein.

Clinical features

It is a bilateral disease, typically affecting young adult males. The common presenting symptoms are sudden appearance of floaters (black spots) in front of the eye or painless loss of vision due to vitreous haemorrhage.

Treatment

1. Medical treatment: a course of oral corticosteroids for extended periods is the main course of treatment during active inflammation. A course of antitubercular therapy has also been recommended in selective cases.

2. Laser photocoagulation of the retina is indicated at the stage of neovascularization.

3. Vitreoretinal surgery is required for non-resolving vitreous haemorrhage and tractional retinal detachment.

Vascular Disorders of Retina

Common vascular disorders of retina include: retinal artery occlusions, retinal vein occlusions, diabetic retinopathy, hypertensive retinopathy, sickle cell retinopathy, retinopathy of prematurity and retinal telangiectasia.

Retinal Artery Occlusion

Etiology

Occlusive disorders of retinal vessels are more common in patients suffering from hypertension and other cardiovascular diseases.

Causes

Common causes of retinal artery occlusion are:

– Atherosclerosis-related thrombosis at the level of lamina cribrosa is the most common cause (75 %) of CRAO.

– Emboli from the carotid artery and those of cardiac origin account for about 20 % cases of CRAO. Retinal arteritis with obliteration (associated with giant cell arteritis) and periarteritis (associated with polyarteritis nodosa, systemic lupus erythematosus, Wegner's granulomatosis and scleroderma) are other causes of CRAO.

– Angiospasm is a rare cause of retinal artery occlusion. It is commonly associated with amaurosis.

– Raised intraocular pressure may occasionally be associated with obstruction of retinal arteries for example due to tight cerclage in retinal detachment surgery.

– Thrombophilic disorders such as inherited defects of anticoagulants may occasionally be associated with CRAO in young individuals.

Clinical features

Clinically retinal artery occlusion may present as central retinal artery occlusion or branch artery occlusion. It is more common in males than females. It is usually unilateral but rarely may be bilateral (1 to 2 % cases).

Central retinal artery occlusion (CRAO)

It occurs due to obstruction at the level of lamina cribrosa.

Symptoms

Patient complains of sudden painless loss of vision.

Signs

Direct pupillary light reflex is absent.

On ophthalmoscopy examination retinal arteries are markedly narrowed but retinal veins look almost normal. Retina becomes milky white due to edema. Central part of the macular area shows cherry-red spot due to vascular choroid shining through the thin retina of this region. In eyes with a cilioretinal artery, part of the macular will remain normal. Blood column within the retinal veins is segmented (cattle trucking). After a few weeks the edema subsides, and atrophic changes occur which include grossly attenuated thread-like arteries and consecutive optic atrophy.

Treatment of the central retinal artery occlusion is unsatisfactory, as retinal tissue cannot survive ischemia for more than a few hours. The emergency treatment should include:

1. Immediate lowering of intraocular pressure by intravenous mannitol and intermittent ocular massage. It may aid the arterial perfusion and also help in dislodging the embolus. Even paracentesis of anterior chamber has been recommended for this purpose.

2. Vasodilators and inhalation of a mixture of 5 percent carbon dioxide and 95 percent oxygen (practically patient should be asked to breathe in a polythene bag) may help by relieving element of angiospasm.

3. Anticoagulants may be helpful in some cases.

4. Intravenous steroids are indicated in patients with giant cell arteritis.

Complications

In some cases “neovascular glaucoma” with incidence varying from 1 to 5 %, may occur as a delayed complication of the central retinal artery occlusion.

Branch retinal artery occlusion (BRAO)

It usually occurs following lodgment of embolus at a bifurcation. Retina distal to the occlusion becomes edematous with narrowed arterioles. Later on the involved area is atrophied leading to permanent sectoral visual field defect.

Retinal Vein Occlusion

It is more common than the artery occlusion. It typically affects elderly patients in sixth or seventh decade of life.

Etiology

1. Pressure on the vein by a sclerotic retinal artery where the two share a common adventitia (e. g., just behind the lamina cribrosa and at arteriovenous crossings).
2. Hyperviscosity of blood as in polycythemia, hyperlipidemia and macroglobulinemia.
3. Periphlebitis retinae which can be central or peripheral.
4. Raised intraocular pressure. Central retinal vein occlusion is more common in patients with primary open-angle glaucoma.
5. Local causes are orbital cellulitis, facial erysipelas and cavernous sinus thrombosis.

Classification

1. Central retinal vein occlusion (CRVO): it may be non-ischemic CRVO (venous stasis retinopathy) or ischemic CRVO (hemorrhagic retinopathy).
2. Branch retinal vein occlusion (BRVO)

Non-ischemic CRVO (venous stasis retinopathy)

It is the most common clinical variety (75 %). It is characterized by mild to moderate visual loss.

Fundus examination in early cases reveals mild venous congestion and tortuosity, a few superficial flame-shaped hemorrhages more in the peripheral than the posterior retina, mild papilledema and mild or no macular edema. In late stages

(after 6–9 months), there appears sheathing around the main veins, and a few cilioretinal collaterals around the disc. Retinal hemorrhages are partly absorbed. Macula may show chronic cystoid edema in moderate cases or may be normal in mild cases.

Treatment is usually not required. The condition resolves with almost normal vision in about 50 % cases. Visual loss in rest of the cases is due to chronic cystoid macular edema, for which no treatment is effective. However, a course of oral steroids for 8–12 weeks may be effective.

Ischemic CRVO

(Hemorrhagic retinopathy)

It refers to acute (sudden) complete occlusion of the central retinal vein. It is characterized by the marked sudden visual loss.

Fundus examination in early cases reveals massive engorgement, congestion and tortuosity of retinal veins, massive retinal hemorrhages (almost whole fundus is full of hemorrhages giving a "splashed-tomato" appearance), numerous soft exudates, and papilledema.

Macular area is full of hemorrhages and is severely edematous. In late stages, marked sheathing around veins and collaterals is seen around the disc.

Neovascularization may be seen at the disc (NVD) or in the periphery (NVE). Macula shows marked pigmentary changes and chronic cystoid edema. The pathognomonic features for differentiating ischemic CRVO from non-ischemic CRVO are presence of relative afferent pupillary defect (RAPD), visual field defects and reduced amplitude of b-wave of electroretinogram (ERG).

Complications

Rubeosis iridis and neovascular glaucoma (NVG) occur in more than 50 percent cases within 3 months (so also called

as 90 days glaucoma); a few cases develop vitreous hemorrhage and proliferative retinopathy.

Treatment

Pan-retinal photocoagulation (PRP) or cryo-application, if the media is hazy, may be required to prevent neovascular glaucoma in patients with widespread capillary occlusion. Photocoagulation should be carried out when most of the intraretinal blood is absorbed, which usually takes about 3–4 months.

Branch retinal vein occlusion (BRVO)

It is more common than the central retinal vein occlusion. It may occur at the following sites: main branch at the disc margin causing hemispheric occlusion, major branch vein away from the disc, at A–V crossing causing quadratic occlusion and small macular or peripheral branch occlusion. In branch vein occlusion edema and hemorrhages are limited to the area drained by the affected vein.

Vision is affected only when the macular area is involved. Secondary glaucoma occurs rarely in these cases. Chronic macular edema and neovascularization may occur as complications of BRVO in about one third cases.

Treatment

Grid photocoagulation may be required in patients with chronic macular edema. In patients with neovascularization, scatter photocoagulation should be carried out.

Hypertensive Retinopathy

It refers to fundus changes occurring in patients suffering from systemic hypertension.

Pathogenesis

Three factors which play role in the pathogenesis of hypertensive retinopathy are vasoconstriction, arteriosclerosis and increased vascular permeability.

1. Vasoconstriction. Primary response of the retinal arterioles to the raised blood pressure is narrowing (vasoconstriction) and is related to the severity of hypertension. It occurs in pure form in young individuals, but is affected by the pre-existing involutional sclerosis in older patients.

2. Arteriosclerotic changes which manifest as changes in arteriolar reflex and A–V nipping result from thickening of the vessel wall and are a reflection of the duration of hypertension. In older patients arteriosclerotic changes may preexist due to involutional sclerosis.

3. Increased vascular permeability results from hypoxia and is responsible for haemorrhages, exudates and focal retinal edema.

Grading of hypertensive retinopathy

Keith and Wegner (1939) have classified hypertensive retinopathy changes into the following four grades:

Grade I. It consists of mild generalized arteriolar attenuation, particularly of small branches, with broadening of the arteriolar light reflex and vein concealment.

Grade II. It comprises marked generalized narrowing and focal attenuation of arterioles associated with deflection of veins at arteriovenous crossings (Salus's sign).

Grade III. This consists of Grade II changes plus copper-wiring of arterioles, banking of veins distal to arteriovenous crossings (Bonnet sign), tapering of veins on either side of the crossings (Gunn sign) and right-angle deflection of veins (Salus's sign). Flame-shaped hemorrhage, cotton-wool spots and hard exudates are also present.

Grade IV. This consists of all changes of Grade III plus silver-wiring of arterioles and papilledema.

Clinical types

Clinically, hypertensive retinopathy may occur in four circumstances:

1. Hypertension with involutionary (senile) sclerosis. When hypertension occurs in elderly patients (after the age of 50 years) in the presence of involutionary sclerosis the fundus changes comprise augmented arteriosclerotic retinopathy.

2. Hypertension without sclerosis. It occurs in young people, where elastic retinal arterioles are exposed to the raised blood pressure for a short duration. There are few retinal signs. The arterioles are constricted, pale and straight with acute-angled branching. There are minimal signs of arteriovenous crossing. Occasionally small hemorrhages may be found. Exudates and papilledema are never seen.

3. Hypertension with compensatory arteriolar sclerosis. This condition is seen in young patients with prolonged benign hypertension usually associated with benign nephrosclerosis. The young arterioles respond by proliferative and fibrous changes in the media (compensatory arteriolar sclerosis). Advanced fundus changes in these patients have been described as "albuminuric or renal retinopathy".

4. Malignant hypertension. It is not a separate variety of hypertension, but is an expression of its rapid progression to a serious degree in a patient with relatively young arterioles undefended by fibrosis. The fundus picture is characterized by marked arteriolar narrowing, papilledema (an essential feature of malignant hypertension), retinal edema over the posterior pole, clusters of superficial flame-shaped hemorrhages and an abundance of cotton wool patches.

Retinopathy in Pregnancy-Induced Hypertension (PIH)

Pregnancy-induced hypertension previously known as "toxemia of pregnancy" is a disease of unknown etiology characterized by the raised blood pressure, proteinuria and generalized edema. Retinal changes are liable to occur in this condition when blood pressure rises above 160/100 mm of Hg and are marked when blood pressure rises above 200/130 mm

of Hg. Earliest changes consist of narrowing of nasal arterioles, followed by generalized narrowing. Severe persistent spasm of vessels causes retinal hypoxia characterized by appearance of "cotton wool spots" and superficial hemorrhages.

If pregnancy is allowed to continue, further progression of retinopathy occurs rapidly. Retinal edema and exudation is usually marked and may be associated with "macular star" or "flat macular detachment". Rarely, it may be complicated by bilateral exudative retinal detachment. Prognosis for retinal reattachment is good, as it occurs spontaneously within a few days of termination of pregnancy.

Management

Changes of retinopathy are reversible and disappear after the delivery, unless organic vascular disease is established. Therefore, in pre organic stage when patient responds well to conservative treatment, the pregnancy may justifiably be continued under close observation. However, the advent of hypoxic retinopathy (soft exudates, retinal edema and hemorrhages) should be considered an indication for termination of pregnancy; otherwise, permanent visual loss or even loss of life (of both mother and fetus) may occur.

Diabetic Retinopathy

It refers to retinal changes seen in patients with diabetes mellitus. With increase in the life expectancy of diabetics, the incidence of diabetic retinopathy (DR) has increased. In Western countries, it is the leading cause of blindness.

Risk factors associated with occurrence of DR are:

1. Duration of diabetes is the most important determining factor. Roughly 50 percent of patients develop DR after 10 years, 70 percent after 20 years and 90 percent after 30 years of onset of the disease.

2. Sex. Incidence is more in females than males (4:3).

3. Poor metabolic control is less important than duration, but is nevertheless relevant to the development and progression of DR.

4. Heredity. It is transmitted as a recessive trait without sex linkage. The effect of heredity is more on the proliferative retinopathy.

5. Pregnancy may accelerate the changes of diabetic retinopathy.

6. Hypertension, when associated, may also accentuate the changes of diabetic retinopathy.

7. Other risk factors include smoking, obesity and hyperlipidemia.

Classification

Diabetic retinopathy has been variously classified. Presently followed classification is as follows:

I. Non-proliferative diabetic retinopathy (NPDR):

– mild NPDR;
– moderate NPDR;
– severe NPDR;
– very severe NPDR.

II. Proliferative diabetic retinopathy (PDR).

III. Diabetic maculopathy.

IV. Advanced diabetic eye disease (ADED).

I. Non-proliferative diabetic retinopathy (NPDR)

Ophthalmoscopic features of NPDR include:

– micro aneurysms in the macular area (the earliest detectable lesion);

– retinal hemorrhages both deep (dot and blot hemorrhages) and superficial hemorrhages (flame-shaped);

– hard exudates-yellowish-white waxy-looking patches are arranged in clumps or in circinate pattern;

– retinal edema characterized by retinal thickening;

– cotton-wool spots (if > 8, there is high risk of developing PDR);

– venous abnormalities, beading, looping and dilatation;

– intra-retinal microvascular abnormalities (IRMA);

– dark-blot hemorrhages representing hemorrhagic retinal infarcts.

On the basis of severity of the above findings the NPDR has been further classified as under:

1. Mild NPDR:

– at least one micro-aneurysm or intra-retinal hemorrhage;

– hard/soft exudates may or may not be present.

2. Moderate NPDR:

– Moderate micro-aneurysms/intra-retinal hemorrhage.

– Early mild IRMA.

– Hard/soft exudates may or may not present.

3. Severe NPDR. Any one of the following (4–2–1 Rule):

– four quadrants of severe micro aneurysms/intra-retinal hemorrhages;

– two quadrants of venous beading;

– one quadrant of IRMA changes.

4. Very severe NPDR. Any two of the following (4–2–1 Rule):

– four quadrants of severe micro aneurysms/intra-retinal hemorrhages;

– two quadrants of venous beading;

– one quadrant of IRMA changes.

II. Proliferative diabetic retinopathy (PDR)

Proliferative diabetic retinopathy develops in more than 50 percent of cases after about 25 years of onset of the disease. Therefore, it is more common in patients with juvenile onset

diabetes. The hallmark of PDR is the occurrence of neovascularization over the changes of very severe non-proliferative diabetic retinopathy.

It is characterized by proliferation of new vessels from the capillaries, in the form of neovascularization at the optic disc (NVD) and/or elsewhere (NVE) in the fundus, usually along the course of the major temporal retinal vessels. These new vessels may proliferate in the plane of retina or spread into the vitreous as vascular fronds. Later on condensation of connective tissue around the new vessels results in formation of fibro vascular epiretinal membrane. Vitreous detachment and vitreous hemorrhage may occur in this stage.

Types

On the basis of high risk characteristics (HRCs) described by diabetic retinopathy study (DRS) group, the PDR can be further classified as below:

1. PDR without HRCs (Early PDR).
2. PDR with HRCs (Advanced PDR).

High risk characteristics (HRC) of PDR are as follows:

– NVD 1/4 to 1/3 of disc area with or without vitreous hemorrhage (VH) or pre-retinal hemorrhage (PRH);
– NVD < 1/4 disc area with VH or PRH;
– NVE > 1/2 disc area with VH or PRH.

III. Diabetic maculopathy

Changes in macular region need special mention, due to their effect on vision. These changes may be associated with non-proliferative diabetic retinopathy (NPDR) or proliferative diabetic retinopathy (PDR).

The diabetic macular edema occurs due to increased permeability of the retinal capillaries. It is termed as clinically significant macular edema (CSME) if one of the following criteria is present on the slit-lamp examination with 90D lens:

– thickening of the retina at or within 500 micron of the centre of the fovea;

– hard exudate at or within 500 micron of the centre of fovea associated with adjacent retinal thickening.

Clinico-angiographically diabetic maculopathy can be classified into four types:

1. Focal exudative maculopathy.

It is characterized by micro aneurysms, hemorrhages, macular edema and hard exudates which are usually arranged in a circinate pattern. Fluorescein angiography reveals focal leakage with adequate macular perfusion.

2. Diffuse exudative maculopathy.

It is characterized by diffuse retinal edema and thickening throughout the posterior pole, with relatively few hard exudates. Fluorescein angiography reveals diffuse leakage at the posterior pole.

3. Ischemic maculopathy.

It occurs due to microvascular blockage. Clinically it is characterized by marked visual loss with micro aneurysms, hemorrhages, mild or no macular edema and a few hard exudates. Fluorescein angiography shows areas of non-perfusion which in early cases are in the form of enlargement of fovea avascular zone (FAZ), later on areas of capillary dropouts are seen and in advanced cases precapillary arterioles are blocked.

4. Mixed maculopathy.

Combined features of ischemic and exudative maculopathy are present.

IV. Advanced diabetic eye disease

It is the end result of uncontrolled proliferative diabetic retinopathy. It is marked by complications such as:

– tractional retinal detachment;

– neovascular glaucoma;

– persistent vitreous hemorrhage.

Investigations:

– urine examination;

– blood sugar estimation;

– fundus fluorescein angiography should be carried out to elucidate areas of neovascularization, leakage and capillary nonperfusion.

Management

Screening for diabetic retinopathy: to prevent visual loss occurring from diabetic retinopathy, a periodic follow-up is very important for a timely intervention.

The recommendations for periodic fundus examination are as follows:

– every year, till there is no diabetic retinopathy or there is mild NPDR;

– every 6 months, in moderate NPDR;

– every 3 months, in severe NPDR;

– every 2 months, in PDR with no high risk characteristic.

Medical treatment

Besides laser and surgery to the eyes (as indicated and described below), the medical treatment also plays an essential role.

Medical treatment for diabetic retinopathy can be discussed as:

1. Control of systemic risk factors is known to influence the occurrence, progression and effect of laser treatment on DR.

The systemic risk factors which need attention are:

– strict metabolic control of blood sugar;

– lipid reduction;

– control of associated anemia;

– control of associated hyperproteinemia.

2. Role of pharmacological modulation. Pharmacological inhibition of certain biochemical pathways involved in the pathogenesis of retinal changes in diabetes is being evaluated. These include:

– protein kinase C (PKC) inhibitors;

– vascular endothelial growth factors (VEGF) inhibitors;

– aldose reductase and ACE inhibitors;

– antioxidants such as vitamin E.

3. Role of intravitreal steroids in reducing diabetic macular edema is also being stressed recently by following modes of administration:

– fluocinolone acetonide intravitreal implant;

– intravitreal injection of triamcinolone (2 to 4 mg);

Photocoagulation

It remains the mainstay in the treatment of diabetic retinopathy and maculopathy. Either argon or diode laser can be used. The protocol of laser application is different for macula and rest of the retina as follows:

1. Macular photocoagulation

Macula is treated by laser only if there is clinically significant macular edema (CSME). Laser treatment is contraindicated in ischemic diabetic maculopathy. In patients with PDR associated with CSME, macular photo-coagulation should be considered first, i. e. before PRP since the latter may worsen macular edema.

Macular photocoagulation includes two techniques:

Focal treatment with argon laser is carried out for all lesions (micro aneurysms, IRMA or short capillary segments) 500–3000 microns from the centre of the macula, believed to be leaking and causing CSME. Spot size of 100–200 μm of 0.1 second duration is used.

Grid treatment: grid pattern laser burns are applied in the macular area for diffuse diabetic macular edema.

2. Panretinal photocoagulation (PRP)

Panretinal photocoagulation or scatter laser consists of 1200–1600 spots, each 500 μm in size and 0.1 sec of duration. Laser burns: 2–3 disc areas are applied from the centre of the macula extending peripherally to the equator.

Surgical treatment

It is required in advanced cases of PDR. Pars plana vitrectomy is indicated for dense persistent vitreous hemorrhage, tractional retinal detachment, and epiretinal membranes. Associated retinal detachment also needs surgical repair.

Retinopathies of Blood Dyscrasias

These are seen in patients suffering from anemias, leukemias and polycythemias.

Anemic retinopathy

In anemia, retinal changes are liable to occur when hemoglobin level falls by 50 percent and are consistently present when it is below 35 percent (5 gm%).

Anemic retinopathy is characterized by pale arterioles and a pale general background of the fundus. Retinal veins are dilated. Superficial retinal and preretinal (sub hyaloid) hemorrhages may be seen in posterior half of the fundus. A few hemorrhages have white centres (Roth spots). Rarely, a few soft exudates (cotton-wool patches) may also be present.

Leukemic retinopathy

It is characterized by pale and orange fundus background with dilated and tortuous veins. In later stages, greyish white lines may be seen along the course of the veins (due to perivascular leukemic infiltration).

Retinopathy of Prematurity

Retinopathy of prematurity (ROP) is a bilateral proliferative retinopathy, occurring in premature infants with low birth weight who often have been exposed to high

concentration of oxygen. Earlier this disease was known as retrolental fibroplasia.

Dystrophies and Degenerations of Retina

A wide variety of dystrophies and degenerations of the retina have been described and variously classified. These lesions are beyond the scope of this chapter, only a common retinal dystrophy (retinitis pigmentosa), and a few peripheral retinal degenerations, some of the vitreoretinal degenerations are described here.

Retinitis Pigmentosa

This primary pigmentary retinal dystrophy is a hereditary disorder predominantly affecting the rods more than the cones.

Inheritance

Most common mode is autosomal recessive, followed by autosomal dominant. X-linked recessive is the least common.

Incidence

It occurs in 5 persons per 1 000 of the world population.

– *Age*. It appears in the childhood and progresses slowly, often resulting in blindness in advanced middle age.

– *Race*. No race is known to be exempt or prone to it.

– *Sex*. Males are more commonly affected than females in a ratio of 3:2.

– *Laterality*. Disease is almost invariably bilateral and both the eyes are equally affected.

Clinical features

I. Visual symptoms:

1. Night blindness. It is the characteristic feature and may present several years before the visible changes in the retina appear. It occurs due to degeneration of the rods.

2. Dark adaptation. Light threshold of the peripheral retina is increased; though the process of dark adaptation itself is not affected until very late.

3. Tubular vision occurs in advanced cases.

II. Fundus changes:

1. *Retinal pigmentary changes*. These are typically perivascular and resemble bone corpuscles in shape. Initially, these changes are found in the equatorial region only and later spread both anteriorly and posteriorly.

2. *Retinal arterioles* are attenuated (narrowed) and may become thread-like in late stages.

3. *Optic disc* becomes pale and waxy in later stages and ultimately consecutive optic atrophy occurs.

4. *Other* associated changes which may be seen are colloid bodies, choroidal sclerosis, cystoid macular edema, atrophic or cellophane maculopathy.

III. Visual field changes.

Annular or ring-shaped scotoma is a typical feature which corresponds to the degenerated equatorial zone of retina. As the disease progresses, scotoma increases anteriorly and posteriorly and ultimately only central vision is left (tubular vision). Eventually even this is also lost and the patient becomes blind.

IV. Electrophysiological changes.

Typical electrophysiological changes appear early in the disease before the subjective symptoms or the objective signs (fundus changes) appear:

1. Electro-retinogram (ERG) is subnormal or abolished.

2. Electro-oculogram (EOG) shows absence of light peak.

Associations of Retinitis Pigmentosa

I. Ocular associations. These include myopia, primary open angle glaucoma, microphthalmos, conical cornea and posterior subcapsular cataract.

II. Systemic associations. These are in the form of the following syndromes:

1. Laurence–Moon–Biedl syndrome. It is characterized by retinitis pigmentosa, obesity, hypogenitalism, polydactyly and mental deficiency.

2. Cockayne's syndrome. It comprises retinitis pigmentosa, progressive infantile deafness, dwarfism, mental retardation, nystagmus and ataxia.

3. Refsum's syndrome. It is characterized by retinitis pigmentosa, peripheral neuropathy and cerebellar ataxia.

4. Usher's syndrome. It includes retinitis pigmentosa and labyrinthine deafness.

5. Hallgren's syndrome. It comprises retinitis pigmentosa, vestibulo-cerebellar ataxia, congenital deafness and mental deficiency.

Atypical forms of retinitis pigmentosa

– *Retinitis pigmentosa sine pigmento*. It is characterized by all the clinical features of typical retinitis pigmentosa, except that there are no visible pigmentary changes in the fundus.

– *Sectorial retinitis pigmentosa*. It is characterized by involvement of only one sector of the retina.

– *Pericentric retinitis pigmentosa*. In this condition all the clinical features are similar to typical retinitis pigmentosa except that pigmentary changes are confined to an area, immediately around the macula.

– *Retinitis punctata albescens*. It is characterised by the presence of innumerable discrete white dots scattered over the fundus without pigmentary changes. Other features are narrowing of arterioles, night blindness and constriction of visual fields.

Treatment

It is most unsatisfactory; rather we can say that till date there is no effective treatment for the disease.

1. Measures to stop progression, which have been tried from time to time, without any breakthrough include:

vasodilators, placental extracts, transplantation of rectus muscles into suprachoroidal space, light exclusion therapy, ultrasonic therapy and acupuncture therapy. Recently vitamins A and E have been recommended to check its progression.

2. Low vision aids (LVA) in the form of "magnifying glasses" and "night vision device" may be of some help.

3. Rehabilitation of the patient should be carried out as per his socio-economic background.

4. Prophylaxis. Genetic counselling for no consanguineous marriages may help to reduce the incidence of disease. Further, affected individuals should be advised not to produce children.

Peripheral Retinal Degenerations

1. *Lattice degeneration*. It is the most important degeneration associated with retinal detachment. Its incidence is 6 to 10 % in general population and 15 to 20 % in myopic patients. It is characterized by white arborizing lines arranged in a lattice pattern along with areas of retinal thinning and abnormal pigmentation. Small round retinal holes are frequently present in it. The typical lesion is spindle-shaped, located between the ora serrata and the equator with its long axis being circumferentially oriented. It more frequently involves the temporal than the nasal, and superior than the inferior halves of the fundus.

2. *Snail tract degeneration*. It is a variant of lattice degeneration in which white lines are replaced by snow-flake areas which give the retina a white frostlike appearance.

3. *Acquired retinoschisis*. The term retinoschisis refers to splitting of the sensory retina into two layers at the level of the inner nuclear and outer plexiform layers. It occurs in two forms – the *congenital* and *acquired*. The latter, also called as *senile retinoschisis*, may rarely act as predisposing factor for primary retinal detachment.

Acquired retinoschisis is characterized by thin, transparent, immobile, shallow elevation of the inner retinal layers which typically produces absolute field defects – the fact which helps in differentiating it from the shallow retinal detachment which produces a relative scotoma. The condition is frequently bilateral and usually involves the lower temporal quadrants, anterior to the equator.

4. *White-with-pressure and white-without pressure*. These are not uncommonly associated with retinal detachment. "White-with-pressure" lesions are characterized by greyish translucent appearance of retina seen on scleral indentation. "White-without pressure" lesions are located in the peripheral retina and may be associated with lattice degeneration.

5. *Focal pigment clumps*. These are small, localized areas of irregular pigmentation, usually seen in the equatorial region. These may be associated with posterior vitreous detachment and/or retinal tear.

6. *Diffuse chorioretinal degeneration.* It is characterized by diffuse areas of retinal thinning and depigmentation of underlying choroid. It commonly involves equatorial region of highly myopic eyes.

7. *Peripheral cystoid retinal degeneration*. It is a common degeneration seen in the eyes of old people. It may predispose to retinal detachment in some very old people.

Vitreoretinal Degenerations

Vitreoretinal degenerations or vitreoretinopathies include:

- Wagner's syndrome;
- Stickler syndrome;
- Favre–Goldmann syndrome;
- familial exudative vitreoretinopathy;
- erosive vitreoretinopathy;

– dominant neovascular inflammatory vitreoretinopathy;

– dominant vitreoretinochoroidopathy.

Note : Characteristic features of some conditions are mentioned here.

Wagner's syndrome

Wagner's syndrome has an autosomal dominant (AD) inheritance with the following features:

– vitreous is liquefied with condensed membranes;

– retina shows narrow and sheathed vessels, and pigmented spots in the periphery;

– choroid may be atrophied;

– cataract may develop as late complication.

Stickler syndrome

Stickler syndrome, also known as hereditary arthro-ophthalmopathy, is an autosomal dominant connective tissue disorder characterized by the following features:

Ocular features:

– vitreous is liquefied and shows syneresis giving appearance of an optically-empty vitreous cavity;

– progressive myopia is very common radial lattice like degeneration associated with pigmentary changes and vascular sheathing;

– bilateral retinal detachment may occur in 30 % cases (commonest inherited cause of retinal detachment in children) ectopia lentis is occasionally associated;

– presenile cataract occurs in 50 % cases.

Orofacial abnormalities include flattered nasal bridge, maxillary hypoplasia, cleft palate and high arched palate.

Arthropathy is characterized by stiff, painful, prominent and hyper extensible large joints.

Other features include deafness and mitral valve prolapse.

Macular Disorders

Macula, being concerned with vision, has attracted the attention of many retina specialists. Consequently, many disorders have been defined and variously classified. A simple, etiological classification for a broad overview of the macular lesions is as follows:

A. Congenital anomalies. These include aplasia, hypoplasia and coloboma.

B. Hereditary dystrophies. These include Best's disease, Stargardt's disease, butterfly-shaped dystrophy, bull's eye dystrophy and central areolar dystrophy.

C. Acquired maculopathies include:

1. *Traumatic lesions*. These include macular edema, traumatic macular degeneration, macular hemorrhage and macular hole.

2. *Inflammations*. These are central chorioretinitis and photoretinitis (sunburn).

3. *Degenerations*. Important conditions are age related macular degeneration (ARMD), and myopic degeneration.

4. *Metabolic disorders*. These include diabetic maculopathy and sphingolipidosis.

5. *Toxic maculopathies*. These are chloroquine and phenothiazine-induced maculopathy.

6. *Miscellaneous acquired maculopathies*. A few common conditions are: central serous retinopathy (CSR), cystoid macular edema (CME), macular hole, and macular pucker. Only a few important macular lesions are described here.

Photoretinitis

Photoretinitis, also known as solar retinopathy or eclipse retinopathy, refers to retinal injury induced by direct or indirect sun viewing. Solar retinopathy is associated with religious sun gazing, solar eclipse observing, telescopic solar

viewing, sun bathing and sun watching in psychiatric disorders. Causes of photic retinopathy, other than solar retinopathy, are:

– welding arc exposure;
– lightening retinopathy;
– retinal phototoxicity from ophthalmic instruments like operating microscope.

Symptoms

These include persistence of negative after-image of the sun, progressing later into a positive scotoma and metamorphopsia. Unilateral or bilateral deceased vision (6/12–6/60) which develops within 1 to 4 hours after solar exposure, usually improves to 6/6–6/12 within six months.

Signs

Initially the fundus may appear normal. Shortly after exposure a small yellow spot with gray margin may be noted in the foveolar and parafoveolar region. The typical lesion, which appears later, consists of a central burnt-out hole in the pigment epithelium surrounded by aggregation of mottled pigment.

Ophthalmoscopically, it appears as a bean or kidney shaped pigmented spot with yellowish white centre in the foveal region. In worst cases, typical macular hole may appear.

Treatment

There is no effective treatment for photoretinitis, so emphasis should be on prevention. Eclipse viewing should be discouraged unless there is proper use of protective eye wear filters (which absorb UV and infrared wave lengths). Prognosis is guarded, since some scotoma and loss in visual acuity by one or two lines mostly persists.

Other Acquired Maculopathies

1. Central serous retinopathy.
2. Idiopathic macular hole.
3. Idiopathic premacular fibrosis.
4. Cystoid macular edema.
5. Myopic maculopathy.

Central Serous Retinopathy

Central serous retinopathy is an eye condition in which fluid builds up behind the retina and affects your vision. The retina is a thin, sensitive layer of tissue in the back of the eye. It converts light into neural signals that are sent to the brain. It helps you recognize the images you see.

The buildup of fluid can cause a partial detachment of the retina. Sometimes the condition, also known as *central serous chorioretinopathy* (CSC), corrects itself without treatment. But the sooner you recognize a change in your vision and have the problem diagnosed, the greater the odds of having a full recovery with no permanent vision loss.

Factors of central serous retinopathy:

– smoke-stack appearance;
– early hyperfluorescent spot;
– later dye passage into subretinal space and vertical ascend;
– subsequent lateral spread until entire area filled;
– ink-blot appearance (less common);
– early hyper fluorescent spot;
– subsequent concentric spread until entire area filled.

Symptoms

CSC usually affects one eye, though you could conceivably have the condition in both eyes during your lifetime. The first symptom you are likely to notice is blurry vision in the affected eye. Vision in that eye may also seem dim.

Other symptoms include:

– darkness may exist in your central vision;
– straight lines may look crooked;
– objects may seem farther away than they really are;
– white objects may have a brownish or grayish tinge, making them appear dull.

Depending on where the fluid buildup is located, you may not notice any change in vision. If the affected part of the retina doesn't include the macula, you may continue to see things properly and with detail. The macula is the part of the retina responsible for distinguishing the finer details of an object you're viewing.

Treatment

CSC is typically a self-limiting disease, and visual recovery usually occurs within a few weeks to months without treatment.

Patients who are taking corticosteroids of any kind should discontinue their use if possible, but only after checking with their prescribing physician to ensure it is safe to stop. Suddenly discontinuing high-dose steroid medications can cause medical problems.

Several therapies have been used to treat chronic CSC, including thermal laser treatments, oral medications, and eye injections. A "cold laser", called photodynamic therapy, is also effective and often used to focally treat the source of fluid leakage under the retina in chronic CSC.

With photodynamic therapy, a drug called verteporfin is injected into the arm, where it travels to the eye. The verteporfin is activated by shining a special cold laser on the source of leakage in chronic CSC. It may also prevent future recurrences in some eyes.

Depending on the severity and timeline of your symptoms, your doctor will choose the best treatment option, which often begins with a trial of observation. Early detection of CSC is very helpful, and most eyes with CSC can be treated successfully to avoid permanent vision loss.

Cystoid Macular Edema (CME)

It refers to collection of fluid in the outer plexiform (Henle's layer) and inner nuclear layer of the retina, centred on the foveola.

Etiology

It is associated with a number of disorders. A few common causes are as follows:

1. As postoperative complication following cataract extraction and penetrating keratoplasty.
2. Retinal vascular disorders, e. g., diabetic retinopathy and central retinal vein occlusion.
3. Intraocular inflammations, e. g., pars planitis, posterior uveitis, Behcet disease.
4. As a side-effect of drugs, e. g., following use of adrenaline eye drops, especially for aphakic glaucoma.
5. Retinal dystrophies, e. g., retinitis pigmentosa.

Pathogenesis

CME develops due to leakage of fluid following breakdown of inner blood-retinal barrier (i. e., leakage from the retinal capillaries).

Clinical features

1. Visual loss. Initially there is minimal to moderate loss of vision, unassociated with other symptoms. If edema persists, there may be permanent decrease in vision.
2. Ophthalmoscopy in clinically established cases reveals a typical "Honey-comb appearance" of macula (due to multiple cystoid oval spaces). CME is best examined with a fundus contact lens on slit-lamp or +90D lenses.
3. Fundus fluorescein angiography demonstrates leakage and accumulation of dye in the macular region which in a well-established case presents a "flower petal appearance".

Myopic Maculopathy

Short-sightedness is characterized by a longer than normal eyeball. Due to this the retina is more stretched and therefore more fragile. In a patient suffering from acute short-sightedness, myopic maculopathy appears when the central retina shows degenerative anomalies due to an excessively long eyeball and with an atrophic retina.

A severe and rapid drop in visual acuity can appear when abnormal blood vessels develop in the centre of the retina. In this case you are in the same situation as wet senile macular degeneration.

Age-Related Macular Degeneration

Age-Related Macular Degeneration (AMD) is the leading cause of severe vision loss in adults over age 50. The Centres for Disease Control and Prevention estimate that 1.8 million people have AMD and another 7.3 million are at substantial risk for vision loss from AMD.

Caucasians are at higher risk for developing AMD than other races. Women also develop AMD at an earlier age than men.

This eye disease occurs when there are changes to the macula, a small portion of the retina that is located on the inside back layer of the eye. AMD is a loss of central vision that can occur in two forms: “dry” (atrophic) and “wet” (exudative).

Most people with macular degeneration have the dry form, for which there isn’t any known treatment. The less common wet form may respond to laser procedures and medication injections, if diagnosed and treated early.

Symptoms and diagnosis of AMD

In its early stages, the following signs of macular degeneration can go unnoticed:

– gradual loss of ability to see objects clearly;

– shape of objects appears distorted;
– straight lines look wavy or crooked;
– loss of clear colour vision;
– a dark or empty area in the centre of vision.

If you experience any of the above signs or symptoms, contact your doctor of optometry immediately for a comprehensive eye examination. Your optometrist will perform a variety of tests to determine if you have macular degeneration or any other eye health problems.

Central vision that is lost to macular degeneration cannot be restored. However, low-vision devices, such as telescopic and microscopic lenses, can maximize existing vision.

Treatment of AMD

With "dry" macular degeneration, the tissue of the macula gradually becomes thin and stops working properly. There is no cure for dry AMD, and any loss in central vision cannot be restored.

However, researchers and doctors believe there is a link between nutrition and the progression of dry AMD. Making dietary changes and taking nutritional supplements can slow vision loss.

Less common, "wet" macular degeneration occurs when fluids leak from newly formed blood vessels under the macula. This leakage blurs central vision. Vision loss can be rapid and severe.

If detected early, wet AMD can be treated with laser treatment, which is often called *photocoagulation*. A highly focused beam of light seals the leaking blood vessels that are damaging the macula. Or in photodynamic therapy (PDT), a medication is injected into the bloodstream, which is then activated by shining a laser into the eye.

Medication can also be injected into the back of the eye to slow the growth of leaky blood vessels. None of these are permanent cures, but they can help minimize vision loss.

Researchers have linked eye-friendly nutrients such as lutein and zeaxanthin, vitamin C, vitamin E and zinc to reducing the risk of certain eye diseases, including macular degeneration.

Clinical types

1. Non-exudative or *atrophic ARMD*. It is also called dry or geographic ARMD and is responsible for 90 percent cases. It typically causes mild to moderate, gradual loss of vision. Patients may complain of distorted vision, difficulty in reading due to central shadowing. Ophthalmoscopically, it is characterised by occurrence of drusens (colloid bodies), pale areas of retinal pigment epithelium atrophy and irregular or clustered pigmentation.

Drusens appear as small discrete, yellowish-white, slightly elevated spots. In later stages, there occurs enlargement of the atrophic areas within which the larger choroidal vessels may become visible (geographic atrophy).

2. Exudative ARMD. It is also called wet or neovascular ARMD. It is responsible for only 10 percent cases of ARMD but is associated with comparatively rapidly progressive marked loss of vision. Typically, the course of exudative ARMD rapidly passes through many stages. These include:

– stage of drusen formation;

– stage of retinal pigment epithelium (RPE) detachment;

– stage of choroidal neovascularisation (CNV);

– stage of haemorrhagic detachment of RPE;

– stage of haemorrhagic detachment of neurosensory retina;

– stage of disciform (scarring) macular degeneration.

Diagnosis

Clinical diagnosis is made from the typical signs described above, which are best elucidated on examination of the macula by slit-lamp biomicroscopy with a +90D/+78D non-contact lens or Mainster contact lens.

Fundus fluorescein angiography and indocyanine green angiography help in detecting choroidal neovascularization (CNV) in relation to foveal avascular zone. Being subfoveal, juxta foveal or extrafoveal CNV may be classical or occult.

Treatment

There is no effective treatment for non-exudative ARMD. However, some treatment options are available for exudative ARMD.

Treatment modalities available to treat exudative (neovascular) ARMD are:

– Argon green-laser photocoagulation is the treatment of choice for extrafoveal choroidal neovascular membrane (CNVM).

– Photodynamic therapy (PDT) is the treatment of choice for subfoveal and juxtafoveal classic CNVM. In PDT, vertiporfin, a photosensitizer or light activated dye is injected intravenously. The area of CNVM is then exposed to light from a diode laser source at a wavelength (689 nm) that corresponds to absorption peak of the dye. The light-activated dye then causes disruption of cellular structures and occlusion of CNVM with minimum damage to adjacent RPE, photoreceptors and capillaries.

– Transpupillary thermotherapy (TTT) with a diode laser (810 nm) may be considered for subfoveal occult CNVM. PDT is definitely better than TTT but is very costly.

– Surgical treatment in the form of submacular surgery to remove CNVM and macular translocation surgery are being evaluated.

– Pharmacologic modulation with antiangiogenic agent like interferon alfa-29, and inhibitor of vascular endothelial growth factor (VEGF) is under experimental trial.

Retinal Detachment

It is the separation of neurosensory retina proper from the pigment epithelium. Normally these two layers are loosely attached to each other with a potential space in between. Hence, actually speaking the term retinal detachment is a misnomer and it should be *retinal separation*.

Classification

The retinal detachment can be classified into three types:

1. Rhegmatogenous retinal detachment (primary retinal detachment).
2. Exudative retinal detachment (solid retinal detachment).
3. Traction retinal detachment (secondary retinal detachment).

Rhegmatogenous or primary retinal detachment

It is usually associated with a retinal break (hole or tear) through which subretinal fluid (SRF) seeps and separates the sensory retina from the pigmentary epithelium.

Etiology

It is still not clear exactly. The predisposing factors and the proposed pathogenesis are as follows:

Pathogenesis of rhegmatogenous retinal detachment (RRD): the retinal breaks responsible for RRD are caused by the interplay between the dynamic vitreoretinal traction and predisposing degeneration in the peripheral retina. Dynamic vitreoretinal traction is induced by rapid eye movements especially in the presence of PVD, vitreous synersis, aphakia and myopia.

Once the retinal break is formed, the liquified vitreous may seep through it separating the sensory retina from the pigment epithelium. As the subretinal fluid (SRF) accumulates, it tends to gravitate downwards. The final shape and position of RD is determined by location of retinal break, and the anatomical limits of optic disc and ora serrata.

Risk factors for developing rhegmatogenous retinal detachment include:

– lattice degeneration (thinning in the peripheral retina, or the area outside of the central retina);
– high myopia (extreme near-sightedness);
– advanced age;
– family history of retinal tears or retinal detachment;
– previous retinal detachment;
– previous eye surgery such as cataract surgery;
– trauma.

Clinical features

Symptoms

There's no pain associated with retinal detachment. Primary symptoms include:

– blurred vision;
– partial vision loss, which makes it seem as if a curtain has been pulled across your field of vision, with a dark shadowing effect;
– sudden flashes of light that appear when looking to the side;
– suddenly seeing many floaters, which are small bits of debris that appear as black flecks or strings floating before your eye

Prodromal symptoms

These include dark spots (floaters) in front of the eye (due to rapid vitreous degeneration) and photopsia, i. e., sensation of flashes of light (due to irritation of retina by vitreous movements).

Symptoms of detached retina

These are as follows:

1. Localised relative loss in the field of vision (of detached retina) is noticed by the patient in early stage which progresses to a total loss when peripheral detachment proceeds gradually towards the macular area.

2. Sudden painless loss of vision occurs when the detachment is large and central. Such patients usually complain of sudden appearance of a dark cloud or veil in front of the eye.

Complications

These usually occur in long-standing cases and include proliferative vitreoretinopathy (PVR), complicated cataract, uveitis and phthisis bulbi.

Treatment

Basic principles and steps of RD surgery are:

1. Sealing of retinal breaks.

2. SRF drainage.

3. To maintain chorioretinal apposition for at least a couple of weeks.

Exudative or solid retinal detachment

It occurs due to the retina being pushed away by a neoplasm or accumulation of fluid beneath the retina following inflammatory or vascular lesions.

Etiology

Its common causes can be grouped as follows:

1. Systemic diseases. These include: toxaemia of pregnancy, renal hypertension, blood dyscrasias and polyarteritis nodosa.

2. Ocular diseases. These include:

– inflammations such as Harada's disease, sympathetic ophthalmia, posterior scleritis, and orbital cellulitis;

– vascular diseases such as central serous retinopathy and exudative retinopathy of Coats;

– neoplasms, e. g., malignant melanoma of choroid and retinoblastoma;

– sudden hypotony due to perforation of globe and intraocular operations.

Clinical features

Exudative retinal detachment can be differentiated from a simple primary detachment by:

– absence of photopsia, holes/tears, folds and undulations;

– the exudative detachment is smooth and convex; at the summit of a tumour it is usually rounded and fixed and may show pigmentary disturbances;

– occasionally, pattern of retinal vessels may be disturbed due to presence of neovascularization on the tumour summit;

– shifting fluid characterized by changing position of the detached area with gravity is the hallmark of exudative retinal detachment;

– on transillumination test a simple detachment appears transparent while solid detachment is opaque.

Treatment

– Exudative retinal detachment due to transudate, exudate and haemorrhage may undergo spontaneous regression following absorption of the fluid. Thus, the treatment should be for the causative disease.

– Presence of intraocular tumours usually requires enucleation.

Traction or secondary retinal detachment

It occurs due to retina being mechanically pulled away from its bed by the contraction of fibrous tissue in the vitreous (vitreoretinal tractional bands).

Etiology

It is associated with the following conditions:

– post-traumatic retraction of scar tissue especially following penetrating injury;

– proliferative diabetic retinopathy;

– post-haemorrhagic retinitis proliferans;

– retinopathy of prematurity;

– plastic cyclitis;

– sickle cell retinopathy;

– proliferative retinopathy in Eales' disease.

Causes and symptoms

Several conditions may cause retinal detachment:

– scarring or shrinkage of the vitreous can pull the retina inward;

– small tears in the retina allow liquid to seep behind the retina and push it forward;

– injury to the eye can simply knock the retina loose;

– bleeding behind the retina, most often due to diabetic retinopathy or injury, can push it forward;

– retinal detachment may be spontaneous; this occurs more often in the elderly or in very nearsighted (myopic) eyes;

– cataract surgery causes retinal detachment 2 % of the time;

– tumours can cause the retina to detach.

Treatment

Reattaching the retina to the inner surface of the eye requires making a scar that will hold it in place and then bringing the retina close to the scarred area. The scar can be made from the outside, through the sclera, using either a laser or a freezing cold probe (cryopexy). Bringing the retina close to the scar can be done in two ways. A tiny belt tightened around the eyeball will bring the sclera in until it reaches the retina. This procedure is called scleral buckling and may be done under general anesthesia.

Using this procedure permits the repair of retinal detachments without entering the eyeball. Sometimes, the eye

must be entered to pump in air or gas, forcing the retina outward against the sclera and its scar. This is called pneumatic retinopexy and can generally be done under local anesthesia.

If all else fails, and especially if there is disease in the vitreous, the vitreous may have to be removed in a procedure called vitrectomy. This can be done through tiny holes in the eye, through which equally tiny instruments are placed to suck out the vitreous and replace it with saline, a salt solution. The procedure must maintain pressure inside the eye so that the eye does not collapse.

Tumours of Retina

Tumours of retina have become a subject of increasing interest to clinical ophthalmologists as well as ocular pathologists. Their classification is given here and only a few of common interests are described.

Classification

A. Primary tumours

1. Neuroblastic tumours. These arise from sensory retina (retinoblastoma and astrocytoma) and pigment epithelium (benign epithelioma and melanotic malignant tumours).

2. Mesodermal angiomata, e. g., cavernous haemangioma.

3. Phakomatoses. These include: angiomatosis retinae (von Hippel–Lindau disease), tuberous sclerosis (Bourneville's disease), neurofibromatosis (von Recklinghausen's disease and encephalo-trigeminal angiomatosis (Sturge–Weber syndrome).

B. Secondary tumours

1. Direct extension, e. g., from malignant melanoma of the choroid.

2. Metastatic carcinomas from the gastrointestinal tract, genitourinary tract, lungs, and pancreas.

3. Metastatic sarcomas.

4. Metastatic malignant melanoma from the skin.

Retinoblastoma

Retinoblastoma is the most common intraocular malignancy afflicting children. The estimated incidence varies by country from 3.4 to 42.6 cases per million live births. Retinoblastoma typically affects young children with the highest incidence in patients less than 4 years old.

It occurs equally in males and females. On presentation, approximately 60 % of cases are unilateral, and the remaining 40 % are bilateral. Patients diagnosed with retinoblastoma are categorized by whether the mutation is germline or somatic. Laterality generally, but imperfectly, predicts whether the retinoblastoma is secondary to a germline or somatic mutation.

Retinoblastoma may arise as hereditary and nonhereditary forms.

1. *Hereditary or familial cases.* In such cases first hit (mutation) occurs in one of the parental germ cells before fertilization. This means mutation will occur in all somatic cells (predisposing to develop even nonocular tumour). Second hit (mutation) occurs late in postzygote phase and affects the second allele, resulting in development of retinoblastoma. Some facts about hereditary retinoblastoma are:

– accounts for 40 % of all cases;

– all bilateral cases and about 15 % of the unilateral cases are hereditary;

– most hereditary cases are multifocal;

– some hereditary cases have trilateral retinoblastoma (i. e., have associated pinealoblastoma);

– inheritance is autosomal dominant and the risk of transmitting the gene mutation is 50 %; 40 % of offspring of a surviver of heraditary retinoblastoma will develop the tumour because of high penetrance;

– there are 40 % chances of developing tumour in a sibling of a child with bilateral retinoblastoma (with unaffected parents).

2. *Non-hereditary or sporadic cases*. In nonhereditary cases both hits (mutations) occur in the embryo after fertilization and in the same retinal cell.

Some facts about non-hereditary (somatic) retinoblastoma are:

– accounts for 60 % of all cases; all non-hereditary cases are unilateral and unifocal and account for 85 % of all the unilateral cases of retinoblastoma;

– patient is not predisposed to get second nonocular cancer;

– tumour is not transmissible.

Clinical picture

It may be divided into four stages:

I. Quiescent stage

It lasts for about 6 months to one year. During this stage, child may have any of the following features:

1. Leukocoria or yellowish-white pupillary reflex (also called as amaurotic cat's eye appearance) is the commonest feature noticed in this stage.

2. Squint, usually convergent, may develop in some cases.

3. Nystagmus is a rare feature, noticed in bilateral cases.

4. Defective vision. Very rarely, when the tumour arises late (3–5 years of age), the child may complain of defective vision.

5. Ophthalmoscopy features of tumour. In the early stages, before the appearance of leukocoria, fundus examination after full mydriasis may reveal the growth.

Ophthalmoscopy signs in two types of retinoblastoma are as follows:

1. *Endophytic retinoblastoma:* it grows inwards from the retina into the vitreous cavity.

2. *Exophytic retinoblastoma*. it grows outwards and separates the retina from the choroid.

II. Glaucomatous stage

It develops when retinoblastoma is left untreated during the quiescent stage. This stage is characterized by severe pain, redness, and watering.

Signs

Eyeball is enlarged with apparent proptosis, conjunctiva is congested, and cornea become hazy, intraocular pressure is raised.

Occasionally, picture simulating severe, acute uveitis usually associated with pseudohypopyon and/or hyphaema may be the presenting mode (retinoblastoma masquerading as iridocyclitis).

III. Stage of extraocular extension

Due to progressive enlargement of tumour, the globe bursts through the sclera, usually near the limbus or near the optic disc. It is followed by rapid fungation and involvement of extraocular tissues resulting in marked proptosis.

IV. Stage of distant metastasis

It is characterized by the involvement of distant structures as follows:

1. Lymphatic spread first occurs in the preauricular and neighbouring lymph nodes.

2. Direct extension by continuity to the optic nerve and brain is common.

3. Metastasis by blood stream involves cranial and other bones. Metastasis in other organs, usually the liver, is relatively rare.

Treatment

1. *Tumour destructive therapy.*

When tumour is diagnosed at an early stage I, i. e. when tumour is involving less than half of retina and the optic nerve is not involved (usually in the second eye of bilateral cases), it may be treated conservatively by any one or more of the following tumour destructive methods depending upon the size

and location of the tumour. The recommendations for sequential aggressive local therapy (SALT) comprising multimodality therapy are as follows:

– chemo reduction followed by local therapy (cryotherapy, thermochemotherapy or brachytherapy) is recommended for large tumours (>12 mm in diameter);

– radiotherapy (external beam radiotherapy, i. e. EBRT or brachytherapy) combined with chemotherapy is recommended for medium size tumour <12 mm in diameter and <8mm in thickness);

– cryotherapy is indicated for a small tumour (<4.5 mm indiameter and <2.5 mm in thickness) located anterior to equator;

– laser photocoagulation is used for a small tumour located posterior to equator <3 mm from fovea;

– thermotherapy with diode laser is used for a small tumour located posterior to equator away from macula.

2. *Enucleation.*

It is the treatment of choice when:

– tumour involves more than half of the retina;

– optic nerve is involved;

– glaucoma is present and anterior chamber is involved.

3. *Palliative therapy is given in the following cases where prognosis for life is dismal in spite of aggressive treatment:*

– retinoblastoma with orbital extension;

– retinoblastoma with intracranial extension;

– retinoblastoma with distant metastasis.

Prognosis

1. If untreated the prognosis is almost always bad and the patient invariably dies. Rarely spontaneous regression with resultant cure and shrinkage of the eyeball may occur due to

necrosis followed by calcification; suggesting role of some immunological phenomenon.

2. Prognosis is fair (survival rate 70–85 %) if the eyeball is enucleated before the occurrence of extraocular extension.

3. Poor prognostic factors are:

– optic nerve involvement;
– undifferentiated tumour cells;
– massive choroidal invasion.

Enucleation

It is excision of the eyeball. It can be performed under local anaesthesia in adults and under general anaesthesia in children.

Indications

1. Absolute indications are retinoblastoma and malignant melanoma.

2. Relative indications are painful blind eye, mutilating ocular injuries, anterior staphyloma and phthisis bulbi.

TOPIC 14. OCULAR MANIFESTATIONS OF SYSTEMIC DISEASES

Ocular involvement in systemic disorders is quite frequent. It is imperative for the ophthalmologists as well as physicians to be well conversant with these. Ocular lesions of the common systemic disorders are enumerated and a few important ones are described here.

Ocular Manifestations of Nutritional Deficiences

1. Deficiency of vitamin A. Ocular manifestations of vitamin A deficiency are referred to as xerophthalmia.

2. Deficiency of vitamin B1 (thiamine). It can cause corneal anaesthesia, conjunctival and corneal dystrophy and acute retrobulbar neuritis.

3. Deficiency of vitamin B2 (riboflavin). It can produce photophobia and burning sensation in the eyes due to conjunctival irritation and vascularization of the cornea.

4. Deficiency of vitamin C. It may be associated with hemorrhages in the conjunctiva, lids, anterior chamber, retina and orbit. It also delays wound healing.

5. Deficiency of vitamin D. It may be associated with zonular cataract, papilledema and increased lacrimation.

Categories of systemic diseases

1. *Congenital* (Down syndrome, Marfan syndrome, myotonic dystrophy, tuberous sclerosis, metabolic disorders involving lysosomal storage and carbohydrate metabolism, and neurofibromatosis).
2. *Traumatic*.
3. *Vascular* (systemic hypertension, embolic disease, central retinal vein occlusion, migraine, blood dyscrasias, hyperviscosity syndromes, sickle cell anemia)
4. *Neoplastic* (metastatic carcinoma).
5. *Autoimmune*.
6. *Idiopathic* (multiple sclerosis).

7. *Infectious* (AIDS).
8. *Metabolic*/*endocrine* (diabetes).
9. *Drugs*/*toxins*.

Xerophthalmia

The term xerophthalmia is now reserved (by a joint WHO and USAID Committee, 1976) to cover all the ocular manifestations of vitamin A deficiency, including not only the structural changes affecting the conjunctiva, cornea and occasionally retina, but also the biophysical disorders of retinal rods and cones functions.

Etiology

It occurs either due to dietary deficiency of vitamin A or its defective absorption from the gut. It has long been recognized that vitamin A deficiency does not occur as an isolated problem but is almost invariably accompanied by protein-energy malnutrition (PEM) and infections.

Clinical features

1. X N (night blindness). It is the earliest symptom of xerophthalmia in children. It has to be elicited by taking detailed history from the guardian or relative.

2. X1A (conjunctival xerosis). It consists of one or more patches of dry, lusterless, non-wettable conjunctiva, which has been well described as "emerging like sand banks at receding tide" when the child ceases to cry.

3. X1B (Bitot's spots). It is an extension of the xerotic process seen in stage X1A. The Bitot's spot is a raised, silvery white, foamy, triangular patch of keratinized epithelium, situated on the bulbar conjunctiva in the inter-palpebral area.

4. X2 (corneal xerosis). The earliest change in the cornea is punctate keratopathy which begins in the lower nasal quadrant, followed by haziness and/or granular pebbly dryness. Involved cornea lacks lustre.

5. X3A and X3B (corneal ulceration/keratomalacia). Stromal defects occur in the late stage due to colliquative necrosis and take several forms. Small ulcers (1–3 mm) occur peripherally; they are characteristically circular, with steep margins and are sharply demarcated. Large ulcers and areas of necrosis may extend centrally or involve the entire cornea. If appropriate therapy is instituted immediately, stromal defects involving less than one-third of corneal surface (X3A) usually heal, leaving some useful vision. However, larger stromal defects (X3B) commonly result in blindness.

6. XS (corneal scars). Healing of stromal defects results in corneal scars of different densities and sizes which may or may not cover the pupillary area. A detailed history is required to ascertain the cause of corneal opacity.

7. XFC (Xerophthalmic fundus). It is characterized by typical seed-like, raised, whitish lesions scattered uniformly over the part of the fundus at the level of optic disc.

Treatment

It includes local ocular therapy, vitamin A therapy and treatment of underlying general disease.

Ocular Manifestations of Systemic Infections

A. Viral infections

Measles. Ocular lesions are: catarrhal conjunctivitis, Koplik's spots on conjunctiva, corneal ulceration, optic neuritis and retinitis.

Mumps. Ocular involvement may occur as conjunctivitis, keratitis, acute dacryoadenitis and uveitis.

Rubella. Ocular lesions seen in rubella (German measles) are congenital microphthalmos, cataract, glaucoma, chorioretinitis and optic atrophy.

Whooping cough. There may be subconjunctival hemorrhages and rarely orbital hemorrhage leading to proptosis.

Ocular Involvement in AIDS

AIDS (Acquired Immune Deficiency Syndrome) is caused by Human immunodeficiency virus (HIV) which is an RNA retrovirus.

Modes of spread include:

– sexual intercourse with an infected person;

– use of infected hypodermic needles;

– transfusion of infected blood;

– transplacental spread to fetus from the infected mothers.

Pathogenesis of AIDS

The HIV infects T-cells, T helper cells, macrophages and B-cells and thus interferes with the mechanism of production of immune bodies thereby causing immunodeficiency. Immune deficiency renders the individuals prone to various infections and tumours, which involve multiple systems and finally cause death.

Ocular manifestations

These occur in about 75 percent of patients and sometimes may be the presenting features of AIDS in an otherwise healthy person or the patient may be a known case of AIDS when his eye problems occur.

Ocular lesions of AIDS may be classified as follows:

1. *Retinal microvasculopathy.*

2. *Usual ocular infections.* These are also seen in healthy people, but occur with greater frequency and produce more severe infections in patients with AIDS. These include:

– Herpes zoster ophthalmicus.

– Herpes simplex infections.

– Toxoplasmosis (chorioretinitis).

– Ocular tuberculosis, syphilis.

– Fungal corneal ulcers.

3. *Opportunistic infections of the eye*. These are caused by microorganisms which do not affect normal patients. They

can infect someone whose cellular immunity is suppressed by HIV infection or by other causes such as leukemia. These include:

– cytomegalovirus (CMV) retinitis;
– candida endophthalmitis;
– cryptococcal infections and pneumocystis carinii;
– choroiditis.

4. *Unusual neoplasms*. Kaposi's sarcoma is a malignant vascular tumour which may appear on the eyelid or conjunctiva as multiple nodules. It is seen in about 3 percent cases of AIDS. Burkitt's lymphoma of the orbit is also seen in a few patients.

5. *Neuro-ophthalmic lesions*. These are thought to be due to CMV or other infections of the brain.

B. Bacterial infections

Septicaemia. Ocular involvement may occur in the form of metastatic retinitis, uveitis or endophthalmitis.

Diphtheria. There may occur: membranous conjunctivitis, corneal ulceration, paralysis of accommodation and paralysis of extraocular muscles.

Brucellosis. It may involve the eye in the form of iritis, choroiditis and optic neuritis.

Gonococcal ocular lesions are: ophthalmia neonatorum, acute purulent conjunctivitis in adults and corneal ulceration.

Meningococcal infection may be associated with: metastatic conjunctivitis, corneal ulceration, paresis of extraocular muscles, optic neuritis and metastatic endophthalmitis or panophthalmitis.

Typhoid fever. It may be complicated by optic neuritis and corneal ulceration due to lagophthalmos.

Tuberculosis. Ocular lesions are granulomatous conjunctivitis, phlyctenular keratoconjunctivitis, interstitial

keratitis, non-granulomatous and granulomatous uveitis, Eales' disease, optic atrophy (following chiasma arachnoiditis secondary to meningitis), and papilledema (due to raised intracranial pressure following intracranial tuberculoma).

Syphilitic lesions (acquired) seen in primary stage are conjunctivitis and chancre of conjunctiva. In secondary stage there may occur iridocyclitis. Tertiary stage lesions include chorioretinitis and gummata in the orbit. Neurosyphilis is associated with optic atrophy and pupillary abnormalities. Ocular lesions of congenital syphilis are: interstitial keratitis, iridocyclitis and chorioretinitis.

Leprosy. Ocular lesions of leprosy include cutaneous nodules on the eyelids, madarosis, interstitial keratitis, exposure keratitis, granulomatous uveitis and dacryocystitis.

C. Parasitic infections

Toxoplasmosis is known to produce necrotising chorioretinitis.

Taenia echinococcus infestation may manifest as hydatid cyst of the orbit, vitreous and retina.

Taenia solium infestation. Cysticercus cysts are known to involve conjunctiva, vitreous, retina, orbit and extra-ocular muscles.

Toxocara infestation may be associated with endophthalmitis.

Onchocerciasis is a common cause of blindness in African countries. Its ocular features include sclerosing keratitis, uveitis, chorioretinitis and optic neuritis invariably ending in optic atrophy.

D. Fungal infections

Systemic fungal infections may be associated with corneal ulceration and endophthalmitis.

Ocular Manifestations of Common Endocrinal and Metabolic Disorders

Diabetes mellitus

Ocular involvement in diabetes is very common. Structure-wise ocular lesions are as follows:

1. Lids: xanthelasma and recurrent sty or internal hordeolum.

2. Conjunctiva: telangiectasia, sledging of the blood in conjunctival vessels and sub conjunctival hemorrhage.

3. Cornea: pigment dispersal at back of cornea, decreased corneal sensations (due to trigeminal neuropathy), punctate keratopathy, Descemet's folds, higher incidence of infective corneal ulcers and delayed epithelial healing due to abnormality in epithelial basement membrane.

4. Iris: rubeosis iridis (neovascularization).

5. Lens: snow-flake cataract in patients with IDDM, posterior subcapsular cataract, early onset and early maturation of senile cataract.

6. Vitreous: vitreous hemorrhage and fibrovascular proliferation secondary to diabetic retinopathy.

7. Retina: diabetic retinopathy and lipaemia retinalis.

8. Intraocular pressure: increased incidence of POAG, neovascular glaucoma and hypotony in diabetic ketoacidosis (due to increased plasma bicarbonate levels).

9. Optic nerve: optic neuritis.

10. Extraocular muscles: ophthalmoplegia due to diabetic neuropathy.

11. Changes in refraction: hypermetropic shift in hypoglycemia, myopic shift in hyperglycemia and decreased accommodation.

Adverse Ocular Effects of Common Systemic Drugs

C.V.S. drugs

– Digitalis: disturbance of colour vision, scotomas.
– Quinidine: optic neuritis (rare).
– Thiazides: xanthopsia (yellow vision), myopia.
– Carbonic anhydrase inhibitors: ocular hypotony, transient myopia.
– Amiodarone: corneal deposits.
– Oxprenolol: photophobia, ocular irritation.

Central nervous system drugs

– Barbiturates: extraocular muscle palsies with diplopia, ptosis, and cortical blindness.
– Chloral hydrate: diplopia, ptosis, miosis.
– Phenothiazine's: deposits of pigment in conjunctiva, cornea, lens and retina, oculogyric crisis.
– Amphetamines: widening of palpebral fissure, dilatation of pupil, paralysis of ciliary muscle with loss of accommodation.
– Monoamine oxidase inhibitors: nystagmus, extraocular muscle palsies, optic atrophy.
– Tricyclic agents: pupillary dilatation (glaucoma risk), cycloplegia.
– Phenytoin: nystagmus, diplopia, ptosis, slight blurring of vision (rare).
– Neostigmine: nystagmus, miosis.
– Morphine: miosis.
– Haloperidol: capsular cataract.
– Lithium carbonate: exophthalmos, oculogyric crisis.
– Diazepam: nystagmus.

Hormones/Female sex hormones

– Retinal artery thrombosis.
– Retinal vein thrombosis.
– Papilledema.
– Ocular palsies with diplopia.

– Nystagmus.
– Optic neuritis and atrophy.
– Retinal vasculitis.
– Scotoma.
– Migraine.
– Mydriasis.
– Cyloplegia.
– Macular edema.

Corticosteroids

– Cataract (posterior subcapsular).
– Local immune suppression causing susceptibility to viral (herpes simplex), bacterial and fungal infections.
– Steroid-induced glaucoma.

Antibiotics

– Chloramphenicol: optic neuritis and optic atrophy.
– Streptomycin: optic neuritis.
– Tetracycline: pseudo tumour cerebri, transient myopia.

Antimalarial Chloroquine

– Macular changes (Bull's eye maculopathy).
– Central scotomas.
– Pigmentary degeneration of the retina.
– Chloroquine keratopathy.
– Ocular palsies.
– Ptosis.
– Electroretinography depression.

TOPIC 15. INJURY OF THE VISUAL ORGANS

Ocular Trauma

Eye injuries occur more often in combination with other injuries (in cases of polytrauma) than in isolation. Life-threatening injuries should always be treated before ophthalmological treatment is started.

Classification of Ocular Injuries by Mechanism of Injury

1. ***Mechanical injuries***
– Extraocular foreign bodies.
– Blunt trauma.
– Perforating injuries.
– Perforating injuries with retained IOFB.
– Sympathetic ophthalmia.
2. ***Non-mechanical injuries***
3. ***Chemical injuries***
– Acid burns.
– Alkali burns.
4. ***Thermal injuries***
5. ***Electrical injuries***
6. ***Radiation injuries***
– Ultraviolet radiations.
– Infrared radiations.
– Ionizing radiation injuries.

Mechanical Injuries

Mechanical injuries can be grouped as under:
– retained extraocular foreign bodies;
– blunt trauma (contusion injuries);
– penetrating and perforating injuries;
– penetrating injuries with retained intraocular foreign bodies.

New ocular trauma terminologies

Before going into details of these mechanical injuries, it will be worthwhile to become familiar with the new ocular trauma terminology system. The term eyewall has been restricted for the outer fibrous coat (cornea and sclera) of the eyeball. The new definitions proposed by the "American Ocular Trauma Society" for mechanical ocular injuries are as follows:

1. Closed-globe injury is one in which the eyewall (sclera and cornea) does not have a full thickness wound but there is intraocular damage.

It includes:

– *Contusion.* It refers to closed-globe injury resulting from blunt trauma. Damage may occur at the site of impact or at a distant site.

– *Lamellar laceration.* It is a closed Globe injury characterized by a partial thickness wound of the eyewall caused by a sharp object or blunt trauma.

2. Open-globe injury is associated with a full thickness wound of the sclera or cornea or both. It includes rupture and laceration of the eyewall:

– *Rupture* refers to a full-thickness wound of eyewall caused by the impact of blunt trauma. The wound occurs due to markedly raised intraocular pressure by an inside-out injury mechanism.

– *Laceration* refers to a full-thickness wound of eyewall caused by a sharp object. The wound occurs at the impact site by an outside-in mechanism. It includes:

- penetrating injury: refers to a single laceration of eyewall caused by a sharp object;
- perforating injury: refers to two full thickness lacerations (one entry and one exit) of the eyewall caused by a sharp object or missile; the two wounds must have been caused by the same agent;

· intraocular foreign body injury is technically a penetrating injury associated with retained intraocular foreign body; however, it is grouped separately because of different clinical implications.

Extraocular Foreign Bodies

Extraocular foreign bodies are quite common in industrial and agricultural workers. Even in day-today life, these are not uncommon.

Common sites

A foreign body may be impacted in the conjunctiva or cornea:

– on the conjunctiva, it may be lodged in the sulcus subtarsalis, fornices or bulbar conjunctiva;

– in the cornea, it is usually embedded in the epithelium or superficial stroma and rarely into the deep stroma.

Symptoms

A foreign body produces immediate:

– discomfort, profuse watering and redness in the eye;

– pain and photophobia are more marked in corneal foreign body than the conjunctival;

– defective vision occurs when it is lodged in the centre of cornea.

Treatment

Extraocular foreign bodies should be removed as early as possible.

1. *Removal of conjunctival foreign body*. A foreign body lying loose in the lower fornix, sulcus subtarsalis or in the canthi may be removed with a swab stick or clean handkerchief even without anesthesia. Foreign bodies impacted in the bulbar conjunctiva need to be removed with the help of a hypodermic needle after topical anesthesia.

2. *Removal of corneal foreign body*. Eye is anaesthetized with topical instillation of 2 to 4 percent xylocaine and the patient is made to lie supine on an examination table. Lids are separated with universal eye speculum; the patient is asked to look straight upward, and light is focused on the cornea. First, an attempt is made to remove the foreign body with the help of a wet cotton swab stick. If it fails, then foreign body spud or hypodermic needle is used. Extra care is taken while removing a deep corneal foreign body, as it may enter the anterior chamber during maneuvering. If such a foreign body happens to be magnetic, it is removed with a hand-held magnet. After removal of foreign body, pad and bandage with antibiotic eye ointment is applied for 24 to 48 hours. Antibiotic eye drops are instilled 3–4 times a day for about a week.

Blunt Trauma

Modes of injury:

– direct blow to the eyeball with a fist, ball or blunt instruments like sticks and big stones;

– accidental blunt trauma to the eyeball may also occur in roadside accidents, automobile accidents, injuries by agricultural and industrial instruments/machines as well as fall upon the projecting blunt objects.

Traumatic lesions of blunt trauma:

– closed-globe injury;
– globe rupture;
– extraocular lesions.

Closed-globe injury

Either there is no corneal or scleral wound at all (contusion) or it is only of partial thickness (lamellar laceration). Contusion injuries may vary in severity from a simple corneal abrasion to an extensive intraocular damage.

Lesions seen in closed-globe injury are briefly enumerated here.

Cornea

1. *Simple abrasions*. These are very painful and diagnosed by fluorescein staining. These usually heal up within 24 hours with "pad and bandage" applied after instilling antibiotic ointment.

2. *Recurrent corneal erosions* (recurrent keratalgia). These may sometimes follow simple abrasions, especially those caused by fingernail trauma. Patient usually gets recurrent attacks of acute pain and lacrimation on opening the eye in the morning. This occurs due to abnormally loose attachment of epithelium to the underlying Bowman's membrane.

Treatment

Loosely attached epithelium should be removed by debridement and "pad and bandage" applied for 48 hours, so that firm healing is established.

3. *Partial corneal tears* (lamellar corneal laceration). These may also follow a blunt trauma.

4. *Blood staining of cornea*. It may occur occasionally from the associated hyphema and raised intraocular pressure. Cornea becomes reddish brown or greenish in colour and in later stages simulates dislocation of the clear lens into the anterior chamber. It clears very slowly from the periphery towards the centre; the whole process may take even more than two years.

5. *Deep corneal opacity*. It may result from edema of corneal stroma or occasionally from folds in the Descemet's membrane.

Sclera

Partial thickness, scleral wounds (lamellar scleral lacerations) may occur alone or in association with other lesions of closed-globe injury.

Anterior chamber

1. *Traumatic hyphema* (blood in the anterior chamber). It occurs due to injury to the iris or ciliary body vessels.

2. *Exudates*. These may collect in the anterior chamber following traumatic uveitis.

Iris, pupil and ciliary body

1. *Traumatic miosis*. It occurs initially due to irritation of ciliary nerves. It may be associated with spasm of accommodation.

2. *Traumatic mydriasis* (iridoplegia). It is usually permanent and may be associated with traumatic cycloplegia.

3. *Rupture of the pupillary margin* is a common occurrence in closed-globe injury.

4. *Radiating tears in the iris stroma*, sometimes reaching up to ciliary body, may occur occasionally.

5. *Iridodialysis*, i. e. detachment of iris from its root at the ciliary body occurs frequently. It results in a D-shaped pupil and a black biconvex area seen at the periphery.

6. *Ant flexion of the iris*. It refers to rotation of the detached portion of iris, in which its posterior surface faces anteriorly. It occurs following extensive iridodialysis.

7. *Retro flexion of the iris*. This term is used when the whole iris is doubled back into the ciliary region and becomes invisible.

8. *Traumatic aniridia* or *congenital aniridia*. In this condition, the completely torn iris (from the ciliary body) sinks to the bottom of anterior chamber in the form of a minute ball.

9. *Angle recession* refers to the tear between longitudinal and circular muscle fibers of the ciliary body. It is characterized by deepening of the anterior chamber and widening of the ciliary body band on gonioscopy. Later on, it is complicated by glaucoma.

10. *Inflammatory changes*. These include traumatic iridocyclitis, haemophthalmus, post-traumatic iris atrophy and pigmentary changes.

Treatment

It consists of atropine, antibiotics and steroids. In the presence of ruptures of pupillary margins and subluxation of lens, atropine is contraindicated.

Lens

It may show the following changes:

1. *Vossius ring*. It is a circular ring of brown pigment seen on the anterior capsule. It occurs due to striking of the contracted pupillary margin against the crystalline lens. It is always smaller than the size of the pupil.

2. *Concussion cataract*. It occurs mainly due to imbibition of aqueous and partly due to direct mechanical effects of the injury on lens fibers. It may assume any of the following shapes:

– discrete sub epithelial opacities are of most common occurrence;

– early rosette cataract (punctate): it is the most typical form of concussion cataract; it appears as feathery lines of opacities along the star-shaped suture lines, usually in the posterior cortex;

– late rosette cataract: it develops in the posterior cortex 1 to 2 years after the injury; its sutural extensions are shorter and more compact than the early rosette cataract;

– traumatic zonular cataract: it may also occur in some cases, though rarely;

– diffuse (total) concussion cataract: it is of frequent occurrence;

– early maturation of senile cataract may follow blunt trauma.

3. *Traumatic absorption of the lens*. It may occur sometimes in young children resulting in aphakia.

4. *Subluxation of the lens*. It may occur due to partial tear of zonules. The subluxated lens is slightly displaced but still present in the pupillary area. On dilatation of the pupil its edge may be seen. Depending upon the site of zonular tear subluxation may be vertical (upward or downward), or lateral (nasal or temporal).

5. *Dislocation of the lens*. It occurs when rupture of the zonules is complete. It may be intraocular (commonly) or extraocular (sometimes). Intraocular dislocation may be anterior or posterior chamber.

Vitreous

1. *Liquefaction* and *appearance of clouds* of fine pigmentary opacities (the most common change).

2. *Detachment of the vitreous* either anterior or posterior.

3. *Vitreous hemorrhage*.

4. *Vitreous herniation* in the anterior chamber may occur with subluxation or dislocation of the lens.

Choroid

1. *Rupture of the choroid*. The rupture of choroid is concentric to the optic disc, and is situated temporal to it. Rupture may be single or multiple. On fundus examination, the choroidal rupture looks like a whitish crescent (due to underlying sclera) with fine pigmentation at its margins. Retinal vessels pass over it.

2. *Choroidal hemorrhage* may occur under the retina (subretinal) or may even enter the vitreous if retina is also torn.

3. *Choroidal detachment* is also known occur following blunt trauma.

Retina

1. *Commotio retinae* (Berlin's edema). It is of common occurrence following a blow on the eye. It manifests as milky white cloudiness involving a considerable area of the posterior pole with a "cherry-red spot" in the fovea region. It may

disappear after some days or may be followed by pigmentary changes.

2. *Retinal haemorrhages*. These are quite common following concussion trauma. Multiple haemorrhages including flame-shaped and preretinal (subhyaloid) D-shaped haemorrhage may be associated with traumatic retinopathy.

3. *Retinal tears*. These may follow a contusion, particularly in the peripheral region, especially in the eyes already suffering from myopia or senile degenerations.

4. *Traumatic proliferative retinopathy* (Retinitis proliferans). It may be secondary to vitreous hemorrhage, forming tractional bands.

5. *Retinal detachment*. It may follow retinal tears or vitreo-retinal tractional bands.

6. *Concussion changes at macula*. Traumatic macular edema is usually followed by pigmentary degeneration. Sometimes, a macular cyst is formed, which on rupture may be converted into a lamellar or full thickness macular hole.

Intraocular pressure changes in closed-globe injury

1. *Traumatic glaucoma*.

2. *Traumatic hypotony*. It may follow damage to the ciliary body and may even result in phthisis bulbi.

Traumatic changes in the refraction

1. Myopia may follow ciliary spasm or rupture of zonules or anterior shift of the lens.

2. Hypermetropia and loss of accommodation may result from damage to the ciliary body (cycloplegia).

Globe Rupture

Globe rupture is a full-thickness wound of the eyewall caused by a blunt object. Globe rupture may occur in two ways:

1. *Direct rupture* may occur, though rarely, at the site of injury.

2. *Indirect rupture* is more common and occurs because of the compression force. The impact results in momentary increase in the intraocular pressure and an inside-out injury at the weakest part of the eyewall, i. e. in the vicinity of canal of Schlemm concentric to the limbus.

The super nasal limbus is the most common site of the globe rupture (countercoup effect is the lower temporal quadrant being most exposed to trauma). Rupture of the globe may be associated with prolapse of uveal tissue, vitreous loss, intraocular hemorrhage and dislocation of the lens.

Treatment

A badly damaged globe should be enucleated. In less severe cases, repair should be done under general anesthesia. Postoperatively atropine, antibiotics and steroids should be used.

Extraocular Lesions

Extraocular lesions caused by blunt trauma are as follows:

1. *Conjunctival lesions*. Subconjunctival hemorrhage occurs very commonly. It appears as a bright red spot. Chemosis and lacerating wounds of conjunctiva (tears) are also not uncommon.

2. *Eyelid lesion*. Ecchymosis of eyelids is of frequent occurrence. Because of loose subcutaneous tissue, blood collects easily into the lids and produces “blackeye”. There may be laceration and avulsion of the lids. Traumatic ptosis may follow damage to the levator muscle.

3. *Lacrimal apparatus lesions*. These include dislocation of lacrimal gland and lacerations of lacrimal passages, especially the canaliculi.

4. *Optic nerve injuries*. These are commonly associated with fractures of the base of skull. These may be in the form of traumatic papillitis, lacerations of the optic nerve, optic nerve

sheath hemorrhage and avulsion of the optic nerve from back of the eye.

5. *Orbital injury*. There may occur fractures of the orbital walls; commonest being the "blow-out fracture" of the orbital floor. Orbital hemorrhage may produce sudden proptosis. Orbital emphysema may occur following ethmoidal sinus rupture.

Penetrating and Perforating Injuries

As mentioned earlier, penetrating injury is defined as a single full-thickness wound of the eyewall caused by a sharp object. While perforating injury refers to two full-thickness wounds (one entry and one exit) of the eyewall caused by a sharp object or missile.

These can cause severe damage to the eye and so should be treated as serious emergencies.

Modes of injury

1. Trauma by sharp and pointed instruments like needles, knives, nails, arrows, screw-drivers, pens, pencils, compasses, glass pieces and so on.

2. Trauma by foreign bodies travelling at very high speed such as bullet injuries and iron foreign bodies in lathe workers.

Effects of penetrating/perforating injury

Damage to the ocular structures may occur by the following effects:

1. *Mechanical effects* of the trauma or physical changes. These are discussed later in detail.

2. *Introduction of infection.* Sometimes, pyogenic organisms enter the eye during perforating injuries, multiply there and can cause varying degree of infection depending upon the virulence and host defense mechanism.

These include: ring abscess of the cornea, sloughing of the cornea, purulent iridocyclitis, endophthalmitis or

panophthalmitis. Rarely tetanus and infection by gas-forming organisms (Clostridium welchii) may also occur.

3. *Post-traumatic iridocyclitis*. It is of frequent occurrence and if not treated properly can cause devastating damage.

4. *Sympathetic ophthalmitis*. It is a rare but most dangerous complication of a perforating injury.

Intraocular Foreign Bodies (IOFB)

Intraocular foreign bodies are seen in 18–40 % of penetrating ocular injuries. Such injuries usually occur at workplace where hammers, chisels or other metal tools striking metals are used. Majority of people injured are working men between 20 and 40 years of age.

Modes of damage

A penetrating/perforating injury with retained foreign body may damage the ocular structures by the following modes:

- mechanical effects;
- introduction of infection;
- reaction of foreign bodies;
- post-traumatic iridocyclitis;
- sympathetic ophthalmic.

Mechanical effects depend upon the size, velocity and type of the foreign body. Foreign bodies greater than 2 mm in size cause extensive damage. The lesions caused also depend upon the route of entry and the site up to which a foreign body has travelled.

Intraocular infection is the real danger to the eyeball. Fortunately, small flying metallic foreign bodies are usually sterile due to the heat generated on their commission. However, pieces of the wood and stones carry a great chance of infection. Unfortunately, once intraocular infection is

established it usually ends in endophthalmitis or even panophthalmitis.

Reactions of the inorganic foreign bodies

Depending upon its chemical nature the following 4 types of reactions are noted in the ocular tissues:

1. No reaction is produced by the inert substances which include glass, plastic, porcelain, gold, silver and platinum.
2. Local irrigative reaction leading to encapsulation of the foreign body occurs with lead and aluminum particles.
3. Suppurative reaction is excited by pure copper, zinc, nickel and mercury particles.
4. Specific reactions are produced by iron (Siderosis bulbi) and copper alloys (Chalcosis).

Siderosis Bulbi

It refers to the degenerative changes produced by an iron foreign body. Siderosis bulbi usually occurs after 2 months to 2 years of the injury. However, earliest changes have been reported after 9 days of trauma.

Mechanism

The iron particle undergoes electrolytic dissociation by the current of rest and its ions are disseminated throughout the eye. These ions combine with the intracellular proteins and produce degenerative changes. In this process, the epithelial structures of the eye are most affected.

Clinical manifestations

1. The anterior epithelium and capsule of the lens are involved firstly. Here, the rusty deposits are arranged radially in a ring. Eventually, the lens becomes cataracts.
2. Iris: it is first stained greenish and later on turns reddish brown.

3. Retina develops pigmentary degeneration which resembles retinitis pigmentosa.

4. Secondary open angle type of glaucoma occurs due to degenerative changes in the trabecular meshwork.

Chalcosis

It refers to the specific changes produced by the alloy of copper in the eye.

Mechanism

Copper ions from the alloy are dissociated electrolytic ally and deposited under the membranous structures of the eye. Unlike iron ions these do not enter into a chemical combination with the proteins of the cells and thus produce no degenerative changes.

Clinical manifestations

1. Kayser–Fleischer ring. It is a golden-brown ring which occurs due to deposition of copper under peripheral parts of the Descemet's membrane of the cornea.

2. Sunflower cataract. It is produced by deposition of copper under the posterior capsule of the lens. It is brilliant golden green in colour and arranged like the petals of a sun flower.

3. Retina. It may show deposition of golden plaques at the posterior pole which reflect the light with a metallic sheen.

Reaction of organic foreign bodies

The organic foreign bodies such as wood and other vegetative materials produce a proliferative reaction characterized by the formation of giant cells. Caterpillar hair produces ophthalmia nodosa, which is characterized by a severe granulomatous iridocyclitis with nodule formation.

Management of retained intraocular foreign bodies (IOFB)

Diagnosis

It is a matter of extreme importance particularly as the patient is often unaware that a particle has entered the eye. To come to a correct diagnosis the following steps should be taken:

1. *History.* A careful history about the mode of injury may give a clue about the type of IOFB.

2. *Ocular examination.* A thorough ocular examination with slit-lamp including gonioscopy should be carried out. The signs which may give some indication about IOFB are: *subconjunctival hemorrhage*, *corneal scar*, *holes in the iris*, and *opaque track through the lens*. With clear media, sometimes IOFB may be seen on ophthalmoscopy in the vitreous or on the retina. IOFB lodged in the angle of anterior chamber may be visualized by gonioscopy.

3. *Plain X-rays orbit.* Antero-posterior and lateral views are indispensable for the location of IOFB, as most foreign bodies are radio opaque.

4. *Localization of IOFB.* Once IOFB is suspected clinically and later confirmed, on fundus examination and/or X-rays, its exact localization is mandatory to plan the proper removal.

Treatment

– complete ocular examination including detailed retinal examination;

– suture of corneal or scleral entrance wound;

– IOFB removal using magnet or forceps, anterior or posterior approach depending on location of IOFB;

– possible pars plana vitrectomy;

– possible lensectomy if traumatic cataract;

– possible repair of retinal detachment.

Sympathetic Ophthalmia

Sympathetic ophthalmia is a serious bilateral granulomatous panuveitis which follows a penetrating ocular trauma. The injured eye is called *exciting eye* and the fellow eye which also develops uveitis is called *sympathizing eye*. Very rarely, sympathetic ophthalmia can also occur following an intraocular surgery.

Incidence of sympathetic ophthalmia has tremendously decreased in the recent years due to meticulous repair of the injured eye utilizing microsurgical techniques and use of the potent steroids.

Etiology of sympathetic ophthalmia is still not known exactly. However, the facts related with its occurrence are as follows:

Predisposing factors:

1. It almost always follows a penetrating wound.
2. Wounds in the ciliary region (the so-called dangerous zone) are more prone to it.
3. Wounds with incarceration of the iris, ciliary body or lens capsule are more vulnerable.
4. It is more common in children than in adults.
5. It does not occur when actual suppuration develops in the injured eye.

Pathogenesis

Various theories have been put forward. Most accepted one is allergic theory, which postulates that the uveal pigment acts as allergen and excites plastic uveitis in the sound eye.

Clinical picture

I. *Exciting (injured) eye.* It shows clinical features of persistent low-grade plastic uveitis, which include ciliary congestion, lacrimation and tenderness. Keratic precipitates may be present at the back of cornea (dangerous sign).

II. *Sympathizing (sound) eye.* It is usually involved after 4–8 weeks of injury in the other eye. Earliest reported case is

after 9 days of injury. Most of the cases occur within the first year. However, delayed and very late cases are also reported.

Sympathetic ophthalmia, almost always, manifests as acute plastic iridocyclitis. Rarely, it may manifest as neuroretinitis or choroiditis.

Clinical picture of the iridocyclitis in sympathizing eye can be divided into two stages:

1. Prodromal stage. Sensitivity to light (photophobia) and transient indistinctness of near objects (due to weakening of accommodation) are the earliest symptoms.

Signs: in this stage the first sign may be the presence of retrolental flare and cells or the presence of a few keratic precipitates (KPs) on back of the cornea. Other signs include mild ciliary congestion, slight tenderness of the globe, fine vitreous haze and disc edema which is seen occasionally.

2. Fully-developed stage. It is clinically characterized by typical signs and symptoms consistent with acute plastic iridocyclitis.

Treatment when sympathetic ophthalmitis has already supervened:

– if the case is seen shortly after the onset of inflammation (i. e. during prodromal stage) in the sympathizing eye, and the injured eye has no useful vision, this useless eye should be excised at once;

– conservative treatment of sympathetic ophthalmia on the lines of iridocyclitis should be started immediately, as follows:

1. Corticosteroids should be administered by all routes, i. e. systemic, periocular injections and frequent instillation of topical drops.

2. In severe cases, immunosuppressant drugs should be started without delay.

3. Topical atropine should be instilled three times a day.

Note. The treatment should be continued for a long time

Prognosis

If sympathetic ophthalmia is diagnosed early (during prodromal stage) and immediate treatment with steroids is started, a useful vision may be obtained. However, in advanced cases, prognosis is very poor, even after the best treatment.

Chemical Injuries

Chemical injuries are by no means uncommon. These vary in severity from a trivial and transient irritation of little significance to complete and sudden loss of vision.

Modes of injury

These usually occur due to external contact with chemicals under the following circumstances:

1. Domestic accidents, e. g., with ammonia, solvents, detergents and cosmetics.
2. Agricultural accidents, e. g., due to fertilizers, insecticides, toxins of vegetable and animal origin.
3. Chemical laboratory accidents, with acids and alkalis.
4. Deliberate chemical attacks, especially with acids to disfigure the face.
5. Chemical warfare injuries.
6. Self-inflicted chemical injuries are seen in malingerers and psychopaths.

Types

– alkali burns;
– acid burns.

Alkali burns

Alkali burns are among the most severe chemical injuries known to the ophthalmologists. Common alkalis responsible for burns are: lime, caustic potash or caustic soda and liquid ammonia (most harmful).

Mechanisms of damage produced by alkali include:

1. Alkali dissociates and saponified fatty acids of the cell membrane and, therefore, destroys the structure of a cell membrane of the tissues.

2. Being hygroscopic, they extract water from the cells, a factor which contributes to the total necrosis.

3. They combine with lipids of cells to form soluble compounds, which produce a condition of softening and gelatinization. The above effects result in an increased deep penetration of the alkali into the tissues. Alkali burns, therefore, spread widely, their action continues for some days and their effects are difficult to circumscribe. Hence, prognosis in such cases must always be guarded.

Clinical picture

It can be divided into three stages:

1. *Stage of acute ischemic necrosis.* In this stage:

– conjunctiva shows marked edema, congestion, widespread necrosis and a copious purulent discharge;

– cornea develops widespread sloughing of the epithelium, edema and opalescence of the stroma;

– iris becomes violently inflamed and in severe cases both iris and ciliary body are replaced by granulation tissue.

2. *Stage of reparation.* In this stage conjunctival and corneal epithelium regenerate, there occurs corneal vascularization and inflammation of the iris subsides.

3. *Stage of complications.* This is characterized by development of symblepharon, recurrent corneal ulceration and development of complicated cataract and secondary glaucoma.

Acid burns

Acid burns are less serious than alkali burns. Common acids responsible for burns are: sulfuric acid, hydrochloric acid and nitric acid.

Chemical effects

The strong acids cause instant coagulation of all the proteins which then act as a barrier and prevent deeper penetration of the acids into the tissues. Thus, the lesions become sharply demarcated.

Ocular lesions:

1. *Conjunctiva.* There occurs immediate necrosis followed by sloughing. Later on symblepharon is formed due to fibrosis.

2. *Cornea.* It is also necrosis and sloughed out. The extent of damage depends upon the concentration of acid and the duration of contact. In severe cases, the whole cornea may slough out followed by staphyloma formation.

Treatment of chemical burns

1. *Immediate and thorough wash with the available clean water or saline.*

2. *Chemical neutralization.* It should be carried out when the nature of offending chemical is known. For example, acid burns should be neutralized with weak alkaline solutions (such as sodium bicarbonate) and alkali burns with weak acidic solutions (such as boric acid or mix). Ethylene diamine tetra acetic acid (EDTA) 1 % solution can also be used as neutralizing agent.

3. *Mechanical removal of contaminant.* If any particles are left behind, particularly in the case of lime, these should be removed carefully with a swab stick.

4. *Removal of contaminated and necrotic tissue*. Necrosis conjunctiva should be excised. Contaminated and necrosis corneal epithelium should be removed with a cotton swab stick.

5. *Maintenance of favourable conditions* for rapid and uncomplicated healing by frequent application of topical atropine, corticosteroids and antibiotics.

6. *Prevention of symblepharon* can be done by using a glass shell or sweeping a glass rod in the fornices twice daily.

7. *Treatment of complications*:

– secondary glaucoma should be treated by topical 0.5 % timolol instilled twice a day along with oral acetazolamide 250 mg 3–4 times a day;

– corneal opacity may be treated by keratoplasty;

– treatment of symblepharon (symblepharectomy).

Thermal injuries

Thermal injuries are usually caused by fire, or hot fluids. The main brunt of such injuries lies on the lids. Conjunctiva and cornea may be affected in severe cases.

Treatment for burns of lids is on general lines. When cornea is affected, it should be treated with atropine, steroids and antibiotics.

Electrical injuries

The passage of strong electric current from the area of eyes may cause the following lesions:

1. Conjunctiva becomes congested.

2. Cornea develops punctate or diffuse interstitial opacities.

3. Iris and ciliary body are inflamed.

4. Lens may develop "electric cataract" after 2–4 months of accident.

5. Retina may show multiple hemorrhages.

6. Optic nerve may develop neuritis.

Radiational injuries

1. *Ultraviolet radiations*. These may cause photo-ophthalmia and may be responsible for senile cataract.

2. *Infrared radiations*. These may cause solar macular burns **(**photo retinitis).

3. *Ionizing radiation injuries*. These are caused following radiotherapy to the tumours in the vicinity of the eyes. The common ocular lesions include:

– radiation keratoconjunctivitis;
– radiation dermatitis of lids;
– radiation cataract.

Photo-Ophthalmia

Photo-ophthalmia refers to occurrence of multiple epithelial erosions due to the effect of ultraviolet rays especially from 311 to 290μ.

Causes

1. Exposure to bright light of a short circuit.
2. Exposure to a naked arc light as in industrial welding and cinema operators.
3. Snow blindness due to reflected ultraviolet rays from snow surface.

Clinical features

– typically, patient presents with severe burning pain, lacrimation, photophobia, blepharospasm, swelling of palpebral conjunctiva and retrotarsal folds;

– there is history of exposure to ultraviolet rays 45 hours earlier;

– on fluorescein staining multiple spots are demonstrated on both corneas.

Prophylaxis

Crooker's glass which cuts off all infrared and ultraviolet rays should be used by those who are prone to exposure, e. g., welding workers, cinema operators.

Treatment

1. Cold compresses.
2. Pad and bandage with antibiotic ointment for 24 hours, heals most of the cases.
3. Oral analgesics may be given if pain is intolerable.
4. Single dose of tranquillizer may be given to apprehensive patients.

Photoretinitis

Photoretinitis, also known as solar retinopathy or eclipse retinopathy, refers to retinal injury induced by direct or indirect sun viewing. Solar retinopathy is associated with religious sun gazing, solar eclipse observing, telescopic solar viewing, sun bathing and sun watching in psychiatric disorders. Causes of photic retinopathy, other than solar retinopathy, are:

– welding arc exposure;
– lightening retinopathy;
– retinal phototoxicity from ophthalmic instruments like operating microscope.

Pathogenesis

Solar radiations damage the retina through:

– photochemical effects produced by UV and visible blue light;

– thermal effects may enhance the photochemical effects. The long visible wave length and infrared rays from the sun are absorbed by the pigment epithelium producing a thermal effect. Therefore, severity of lesion varies directly with the degree of pigmentation of the fundus, duration of exposure and the climatic conditions during exposure.

Symptoms

These include persistence of negative after-image of the sun, progressing later into a positive scotoma and metamorphopsia. Unilateral or bilateral deceased vision (6/12–6/60) which develops within 1 to 4 hours after solar exposure, usually improves to 6/6–6/12 within six months.

Treatment

There is no effective treatment for photoretinitis, so emphasis should be on prevention. Eclipse viewing should be discouraged unless there is proper use of protective eye wear filters (which absorb UV and infrared wave lengths).

Prognosis is guarded, since some scotoma and loss in visual acuity by one or two lines mostly persists.

TOPIC 16. OPHTHALMIA ONCOLOGY

Incidence and Epidemology

Tumours of the eye and orbit are rare. Male to female incidence is similar.

In adults, ***melanoma*** is the most common primary intraocular cancer, followed by ***lymphoma***.

In children ***retinoblastoma*** is the most common tumour followed by ***medulloepithelioma metastases***; secondary intraocular tumours are more common than primary tumours and typically come from breast or lung cancers.

Benign Orbital Tumours

1. Pterygium.
2. Chorodial hemangiomas.
3. Orbital pseudotumours.
4. Thyroid associated orbitopathy.

Malignant Orbital Tumours

1. Metastatic carcinoma to the uvea.
2. Malignant melenoma of the uvea.
3. Retinoblastoma.

The most common malignant orbital tumours in adults include:

Lymphoma is the most common type of malignant orbital tumour in the adults. Occurs mainly in the lacrimal gland, but can occur in any other orbital structure.

Optic nerve glioma is extremely rare tumour that begins in the optic nerve and spreads to the orbit.

Sarcoma: almost any type of sarcoma can involve the orbit:

- angiosarcoma;
- fibrosarcoma;
- osteosarcoma;
- chondrosarcoma;
- liposarcoma;

– malignant fibrous histiocytoma.

Secondary carcinomas can also be secondary cancers that have spread to the orbit from nearby structures, such as the eyeball (intraocular tumours), eyelid, conjunctiva, sinuses or nasal cavity.

Cancers from other parts of the body, such as the breast, lung, prostate, brain and kidney, can also spread (metastasize) to the orbit.

Metastasis from melanoma can also occur but it is uncommon.

TNM Staging

– TX – primary tumour cannot be assessed;
– T0 – no evidence of primary tumour;
– T1 – tumour is 15 mm (0.6 inch) or less in size;
– T2 – tumour is more than 15 mm in size. It has not spread into the globe of the eye or the bony wall of the orbit;
– T3 – tumour is any size and has spread into the orbital tissues or bony walls of the orbit;
– T4 – tumour has spread into one or more of the following:
· the globe;
· periorbital structures, such as the eyelids or temporal fossa;
· nasal cavity and paranasal sinuses;
· central nervous system (the brain and spinal cord);
– NX – regional lymph nodes cannot be assessed;
– N0 – no regional lymph node metastasis;
– N1 – regional lymph node metastasis;
– M0 – no distant metastasis;
– M1 – distant metastasis.

Signs and symptoms:

– proptosis is the most important sign;
– blurred vision;
– diplopia;

– strabismus;
– whitish or yellowish glow through the pupil;
– decreasing/loss of vision;
– red and painful eye;
– pain.

Management:

– systemic examination;
– ophthalmic examination;
– vascular study-orbital venography;
– carotid angiography, MR angiography;
– routine blood investigation;
– imaging of bony structure-digital X ray;
– ocular ultrasonography;
– CT scan & MRI;
– biopsy.

Treatment

Orbital tumour may be treated by:

– surgery;
– chemotherapy;
– external beam radiotherapy;
– plaque brachytherapy.

Lacrimal Gland Tumours

Lacrimal gland tumours are seen more frequently in the third decade of life, and the second bimodal peak is in the teenage years.

The lacrimal gland is a bilobed eccrine secretory gland, which is situated in the super temporal orbit. The 2 lobes of the lacrimal gland are:

– *orbital lobe*;
– *palpebral lobe*.

Lacrimal gland swelling can be classified broadly into *inflammatory* and *neoplastic* subtypes.

Inflammatory etiologies include:

– *dacryoadenitis*;

– *sarcoidosis*.

Most of the neoplastic lesions in the lacrimal gland are epithelial in origin, 50 % are classified as *benign* and 50 % – as *malignant pseudotumours*.

Benign lesions:

– pleomorphic adenomas;
– reactive lymphoid hyperplasia;
– oncocytomas.

These lesions are slowly growing masses more commonly found in adults in their forth to fifth decades of life.

Malignant lesions:

– adenoid cystic carcinoma: comprising 50 % of malignant tumours of lacrimal gland and 25 % of all lacrimal gland tumours;
– adenocarcinoma;
– squamous cell carcinoma;
– mucoepidermoid carcinoma;
– lymphomas.

TNM staging

– TX: the primary tumour cannot be assessed;
– T0: no evidence of primary tumour;
– T1: the tumour is 2 centimeters (cm) or smaller and may or may not extend outside of the lacrimal gland to the orbital soft tissue;
– T2: the tumour is between 2 cm and 4 cm and likely extends to the orbital soft tissue;
– T3: the tumour is greater than 4 cm and likely extends to the orbital soft tissue;
– T4: the tumour has invaded the periosteum (the membrane of connective tissue that covers the bone) or the orbital bone;
– T4a: the tumour has invaded the periosteum;
– T4b: the tumour has invaded the orbital bone;

– T4c: the tumour has extended beyond the orbit to adjacent structures, including the brain and sinuses;
– NX: the regional lymph nodes cannot be evaluated;
– N0: there is no regional lymph node metastasis;
– N1: there is regional lymph node metastasis;
– MX: tistant metastasis cannot be evaluated;
– M0: there is no distant metastasis;
– M1: there is metastasis to other parts of the body.

Signs and symptoms

– Malignant lesions characteristically present with a subacute course of proptosis and temporal sensory loss in the distribution of the lacrimal nerve in one third of patients.

– Diplopia and diminished visual acuity can be seen in rapid progressive lesions.

– Benign lesions commonly present with painless inferonasal globe displacement and fullness of the superotemporal lid and orbit.

Diagnosis

– CT scan of *benign epithelial lesions*, such as pleomorphic adenomas, reveals a well-circumscribed, pseudoencapsulated lesion in the superotemporal fossa.

– In contrast, *malignant epithelial lesions*, such as adenoid cystic carcinoma, usually present as an irregular mass, producing bony erosion (70 %) and occasional calcification (20 %).

Treatment

– *Surgery*: it is the mainstay treatment of lacrimal gland tumours.

– *Radiotherapy*: it is most often used for lacrimal gland lymphoma; the dosage of radiation used, the site and type of the tumour significantly affect the risks of side effects.

– *Chemotherapy*.

– *Immunotherapy*: rituximab (rituxan) is the most common targeted therapy used in the treatment of a lacrimal gland tumour.

Side effects

Cataracts are a very common side effect of radiation therapy to the eye area.

Also, loss of eyelashes and/or a dry eye can occur with external-beam radiation therapy.

Other side effects include radiation retinopathy.

Radiation optic neuropathy involves nerve damage in the eye.

Malignant Melanoma of the Uvea

Uvea is the primary matrix of eye. It is continuous with in the iris, ciliary body, and choroid.

Uveal melanoma is the most common primary intraocular malignancy in adults and account for 5 % of all melanomas.

Etiology

– low socio economic status;
– UV light exposure;
– choroidal melanomas are located posterior to the ciliary body;
– iris melanomas are the most easily visible, they tend to be smallest at diagnosis, and lest likely to metastasis;
– ciliary body melanomas are least visible as they are hidden behind iris, and more likely to metastasize.

Complications

The most frequently encountered complications are:

– exudative retinopathy;
– neovascular glaucoma;
– vitreous hemorrhage;
– radiation retinopathy;
– cataract.

TOPIC 17. FIRST AID IN OPHTHAMOLOGY

Assess Acuity in Both Eyes

Trauma to the eye clearly needs urgent treatment. Equally important is the fact that even a trivial injury to the only good eye of a pair is potentially far more serious than it would be if both eyes have good equal acuity.

The acuity of the uninjured eye is far more important at the outset than that of the injured eye because it indicates the level of care that is needed, perhaps making all the difference between admission and outpatient treatment. The acuity should be recorded in all cases, for both clinical and medicolegal reasons.

As general principles local anesthetics should not be used more than once, though general sedation is often sensible. Atropine drops 1 % or mydrilate, which dilate the pupil, often give great relief from pain.

Chemical and Thermal Burns: Immediate First Aid

Immediate first aid is needed in all forms of chemical burns. Speed is essential, and the eyes should be washed out with copious amounts of any clean water. This may be difficult because pain causes spasm of the lids, but it must be attempted, and within seconds if possible.

Many chemical burns have late complications, notably lime burns because of retained particles. Therefore all burns that affect the globe require early hospital attention. Late complications include adhesions of the globe to the lid, so that liberal and frequent applications of chloramphenicol ointment or drops are desirable.

It is difficult to know whether to cover the eyes, but the retention of toxic substances and danger of adhesions will be less if the eye remains uncovered or only loosely covered.

In many corneal conditions extremely painful iris spasm may often occur. This is indicated by constriction of the pupil, but even without this sign the patient is likely to be much more comfortable if mydrilate 1 % or atropine 1 % is instilled. Dark glasses are helpful.

Many chemicals have specific antidotes, but searching for these may cause delay. A supply of antidotes is clearly necessary in industrial practice, where there may be particular risks from specific toxic agents. The antidote to lime, for example, is a saturated solution of glucose.

Thermal burns seldom affect only the globe, but general sedation and antibiotic drop or ointment into the eye is sensible treatment.

When only the lids are injured externally the ultimate dangers are less and less immediate. Burnt lids should be treated like any other burn of the skin, though eventually scarring in more severe cases may lead to complications affecting the globe.

Sharp Wounds: General Management

When serious damage of the eye is feared the less examination of the eye that is carried out is better. If the globe is likely to have been damaged attempts to determine the extent of the wound may extend the damage. The patient should be immobilized, given a general analgesic if he/she is in severe pain, and transferred rapidly to the casualty department of a hospital that has facilities for eye surgery. No pressure should be put on the eye, so any covering should be light.

Penetrating wounds, which are often surprisingly painless, need urgent attention because the longer repair is delayed the more danger there is of the intraocular contents being disturbed or extruded and the greater the chance of infection.

Penetrating Wounds: State of Pupil as an Important Sign

An important sign of penetration is the state of the pupil. Instillation of local anesthetics, such as Amethocaine, may be necessary to make this examination, which is harmless in a co-operative patient. The use of such anesthetic should not be continued to relieve pain, however, as it may delay healing or promote further damage. A penetrating wound is usually indicated by deformed (oval) and sluggish pupil. If the pupil is deformed, however slight other signs may be, penetration must be assumed until it has been definitely excluded. Nevertheless, a round pupil does not indicate the penetration has not occurred.

Injuries from flying particles, during operations such grinding or chipping with hammer and chisel, are particularly misleading. In grinding, high-velocity metal particles may penetrate the eye and eventually lead to blindness with little or no apparent external injury or pain. In chipping it is often falsely assumed that a fragment from the object being chiseled has entered the eye, whereas it is just as common for a particle from one of the tools to have done so. It is important to find a foreign body in these cases and essential to take an X-ray picture. Retained intraocular foreign bodies may cause blindness many years later.

Removing Foreign Bodies: Harder than It Looks

The pain in superficial wounds may be much greater than with penetrating wounds, especially if the cornea is affected. Superficial foreign bodies are most often found either inside the upper or lower lid or on the cornea. The sensation of a foreign body under the lid, scraping the cornea with each blink, is identical with that of a scratch on the cornea without a foreign body. A good search for a foreign and its removal in the surgery will spare the patient a visit to hospital. A local anesthetic is usually needed.

Most foreign bodies look black against the lid or sclera but are often difficult to identify against the background of the iris or the pupil. A drop of fluoresce in dye will help to identify the lesion through washing round a corneal foreign body or staining a corneal abrasion. It is difficult to find quite large foreign bodies under the upper lid without exposing the inner surface of the lid, which is often impossible, even under local anesthesia. If it proves impossible the patient should be referred to an ophthalmologist if the symptoms or history of a foreign body are present.

Removing a foreign body from the lower lid is comparatively easy if the patient is co-operative. The patient's head should be firmly supported, and a local anesthetic (amethocaine 1 %) should be given if the foreign body is on the cornea. It may also be helpful if the foreign body is under the lids.

A good instrument to use in removing a foreign body is a piece of white postcard cut into a triangular shape. Gripped by one angle of the triangle the opposing sharp point will remove all superficial foreign bodies with far less trauma than a metallic instrument especially if the particle has to be pursued across the eye after loosened.

Any foreign bodies that resist this technique need more expert removal with more dangerous instruments. Removal should be followed by immediate instillation of an antibiotic (chloramphenicol), as the procedure is not sterile. If the cornea has been affected it is wise to pad the eye for about two hours or longer if the pain returns. A bright light, self-retaining lid speculum and magnification are all aids. Follow-up is advisable if the affected eye is the only useful one, and whenever possible acuity should be recorded in both eyes before treatment and in the injured eye after healing.

Scratches on the cornea commonly accompany foreign bodies and attempts at their removal. At home a baby's

fingernail, or stakes and twigs in the garden are most to be feared. All abrasions strain with fluorescein. Superficial corneal abrasions heal quickly in about 48 hours with a firm pad to stop blinking also has an analgesic effect. Local anesthesia delays healing but dilating the pupil also relieves pain. In all cases when fluorescein staining indicates epithelial damage antibiotic drops twice a day should be used until healing has occurred. Persistence of an abrasion or pain beyond 48 hours may well indicate complications.

Blunt Injuries:
Test Acuity and Exclude Internal Haemorrhage

Blunt injuries are characterized by the familiar black eye, which is caused by impact with objects larger than the orbit. The visual damage is unlikely to be as bad as external appearances suggest. The commonest complication of these injuries is double vision, which needs to be tested for at the full range of vision as soon as the eye is open.

Recovery tends to be spontaneous except when the orbital wall is fractured. Then urgent treatment is needed to reconstitute the normal bony structure, which will prevent permanent limitation of movement caused by secondary adhesions or fibrosis of the extra ocular muscles. Relief from diplopia may always be obtained by covering one eye, though at the expense of depth perception in patients with previously good binocular vision.

Ocular effects include haemorrhage into all part of the globe: intraocular haemorrhages will obviously usually diminish visual acuity. Such haemorrhages need differentiation by an ophthalmologist, and physical rest is sensible until their nature and source has been established. Later complications include secondary retinal detachment. In most cases it is sensible to instill mydrilate two or three times a day until the absence of intraocular damage is proved.

The most dangerous injuries apart from those caused by traffic accidents are those caused by shotgun and air gun pellets, squash balls and champagne corks-projectiles that are small in relation to the orbit. Fireworks and all games with bows and arrows are dangerous to children. Such incident is largely preventable by good discipline. Certain types of swimming goggle can cause dangerous injuries.

A dilated paralysed pupil is often the only sign of a blunt injury. This usually recovers in days or weeks but is often associated with haemorrhage into the vitreous, which must be excluded in every case.

Burns from Sun and Snow

Like its light rays the sun's heart rays are focused on the retina. Intense light initiates a defence mechanism to protect the retina by closing the eyes. Children who have competed with each other to see who could keep their eyes open longest while looking straight at the sun have often burnt their retinas and suffered a considerable permanent loss of vision. This is a variety of the eclipse blindness produced by watching the sun in eclipse with unprotected eyes when focusing of the heat rays is not prevented by a response to the sun's light. In sleep the retina is protected by the globe rolling upwards into the orbit. Ambient sunlight is harmless.

Ultraviolet radiation in the so-called "arc eye" or in snow blindness may cause extremely distressing superficial effects some time after exposure and occurs if the eyes are not protected by tinted shields. Danger to sight is, however minimal. Emergency treatment is of little avail, though bandaging with cold compresses gives partial relief of the pain, and adrenaline drops will reduce congestion temporarily. The use of amethocaine is justified, and its effect is sometimes dramatic. Atropine drops are useful if the pupils are painfully constricted as they reduce the element of spasm.

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Навчальне видання

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ПРАКТИЧНА ОФТАЛЬМОЛОГІЯ

Навчальний посібник

(Англійською мовою)

Художнє оформлення обкладинки Є. В. Нікітюка
Редактор Л. В. Штихно
Комп'ютерне верстання С. Е. Лекішвілі

Формат 60×84/16. Ум.-друк. арк. 22,79. Обл.-вид. арк. 20,66. Тираж 300 пр. Зам. №

Видавець і виготовлювач
Сумський державний університет,
вул. Римського-Корсакова, 2, м. Суми, 40007
Свідоцтво суб'єкта видавничої справи ДК № 3062 від 17.12.2007.