Eye Diseases and Ophthalmology

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Chapter 1

Eye Disease

The World Health Organization publishes a classification of known diseases and injuries called the International Statistical Classification of Diseases and Related Health Problems or ICD-10. This list uses that classification.

H00-H06 Disorders of eyelid, lacrimal system and orbit

- (H00.0) Hordeolum ("stye" or "sty") — a bacterial infection of sebaceous glands of eyelashes.
- (H00.1) Chalazion — a cyst in the eyelid (usually upper eyelid)
- (H01.0) Blepharitis — inflammation of eyelids and eyelashes; characterized by white flaky skin near the eyelashes
- (H02.0) Entropion and trichiasis
- (H02.1) Ectropion
- (H02.2) Lagophthalmos
- (H02.3) Blepharochalasis
- (H02.4) Ptosis
- (H02.6) Xanthelasma of eyelid
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- (H04.2) Epiphora
- (H06.2*) Dysthyroid exophthalmos it is shown that if your eye comes out that it will shrink because the optic fluids drain out

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(H30) Chorioretinal inflammation

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        - choroiditis
        - retinitis
        - retinochoroiditis
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        - choroiditis
        - retinitis
        - retinochoroiditis

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    - Choroiditis
    - Retinitis

    - Retinochoroiditis

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(H31) Other disorders of choroid

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(H32) Chorioretinal disorders in diseases classified elsewhere

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- (H40-H42) Glaucoma — optic neuropathy

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- (H43.9) Floaters — shadow-like shapes which appear singly or together with several others in the field of vision

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- (H47.3) Optic disc drusen — globules progressively calcify in the optic disc, compressing the vasculature and optic nerve fibers

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    - (H49.3-4) Ophthalmoparesis — the partial or total paralysis of the eye muscles
    - (H49.4) Progressive external ophthalmoplegia — weakness of the external eye muscles
    - (H50.0, H50.3) Esotropia — the tendency for eyes to become cross-eyed
    - (H50.1, H50.3) Exotropia — the tendency for eyes to look outward
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    - (H52.1) Myopia (Nearsightedness) — distant objects appear blurred
    - (H52.2) Astigmatism — the cornea or the lens of the eye is not perfectly spherical, resulting in different focal points in different planes

    - (H52.3) Anisometropia — the lenses of the two eyes have different focal lengths
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- (H53.5) Color blindness — the inability to perceive differences between some or all colors that other people can distinguish
    - (H53.5) Achromatopsia / Maskun — a low cone count or lack of function in cone cells
- (H53.6) Nyctalopia (Nightblindness) — a condition making it difficult or impossible to see in the dark
- (H54) Blindness — the brain does not receive optical information, through various causes
    - (H54/B73) River blindness — blindness caused by long-term infection by a parasitic worm (rare in western societies)
    - (H54.9) micro-opthalmia/coloboma — a disconnection between the optic nerve and the brain and/or spinal cord

H55-H59 Other disorders of eye and adnexa

- (H57.9) Red eye — conjunctiva appears red typically due to illness or injury
- (H58.0) Argyll Robertson pupil — small, unequal, irregularly shaped pupils

Other codes

The following are not classified as diseases of the eye and adnexa (H00-H59) by the World Health Organization:

- (B36.1) Keratomycosis — fungal infection of the cornea
- (E50.6-E50.7) Xerophthalmia — dry eyes, caused by vitamin A deficiency
- (Q13.1) Aniridia — a rare congenital eye condition leading to underdevelopment or even absence of the iris of the eye

Chapter 2

Stye

Stye

A stye on an eyelid

ICD-10	H00.0
ICD-9	373.11
DiseasesDB	12583
MedlinePlus	001009
eMedicine	emerg/755
MeSH	D006726

An external **stye** is an infection of the sebaceous glands of Zeis at the base of the eyelashes, or an infection of the apocrine sweat glands of Moll. External styes form on the outside of the lids and can be seen as small red bumps. Internal styes are infections of the meibomian sebaceous glands lining the inside of the eyelids. They also cause a red bump underneath the lid with only generalized redness and swelling visible on the outside. Styes are similar to chalazia, but tend to be of smaller size and are more painful and usually produce no lasting damage. Styes are characterized by an acute onset and

usually short in duration (7–10 days without treatment) compared to chalazia that are chronic and usually do not resolve without intervention.

Causes

Styes are commonly caused by a *Staphylococcus aureus* bacterial infection, or by the blocking of an oil gland at the base of the eyelash. Although they are particularly common in infants, styes are experienced by people of all ages. Styes can be triggered by poor nutrition, sleep deprivation, lack of hygiene or rubbing of the eyes. Sharing of washcloths or face towels should be curtailed to avoid spreading the infection between individuals. Styes can last from one to two weeks without treatment, or as little as four days if treated properly.

Medical professionals will sometimes lance a particularly persistent or irritating stye with a needle to accelerate its draining. A stye's expansion can also be fought with erythromycin ophthalmic ointment. Medical professionals may also treat styes with other antibiotics, such as chloramphenicol or amoxicillin. Chloramphenicol is used successfully in many parts of the world, but contains a black box warning in the United States due to concerns about aplastic anemia, which on rare occasions can be fatal. Erythromycin ointment enjoys widespread use, and may add to comfort and aid in preventing secondary infections. However, it is poorly absorbed when used topically, and usually requires oral dosing to reach the infection with therapeutic levels inside of a stye. Azasite, a topical eye drop form of azithromycin, does appear to penetrate eyelid tissues fairly well, and may be a topical treatment for styes used in the future.

If a stye bursts, care must be taken to cleanse the wound to prevent reinfection.

Signs and symptoms

The first sign of a stye is a small, yellowish spot at the center of the bump that develops as pus expands in the area.

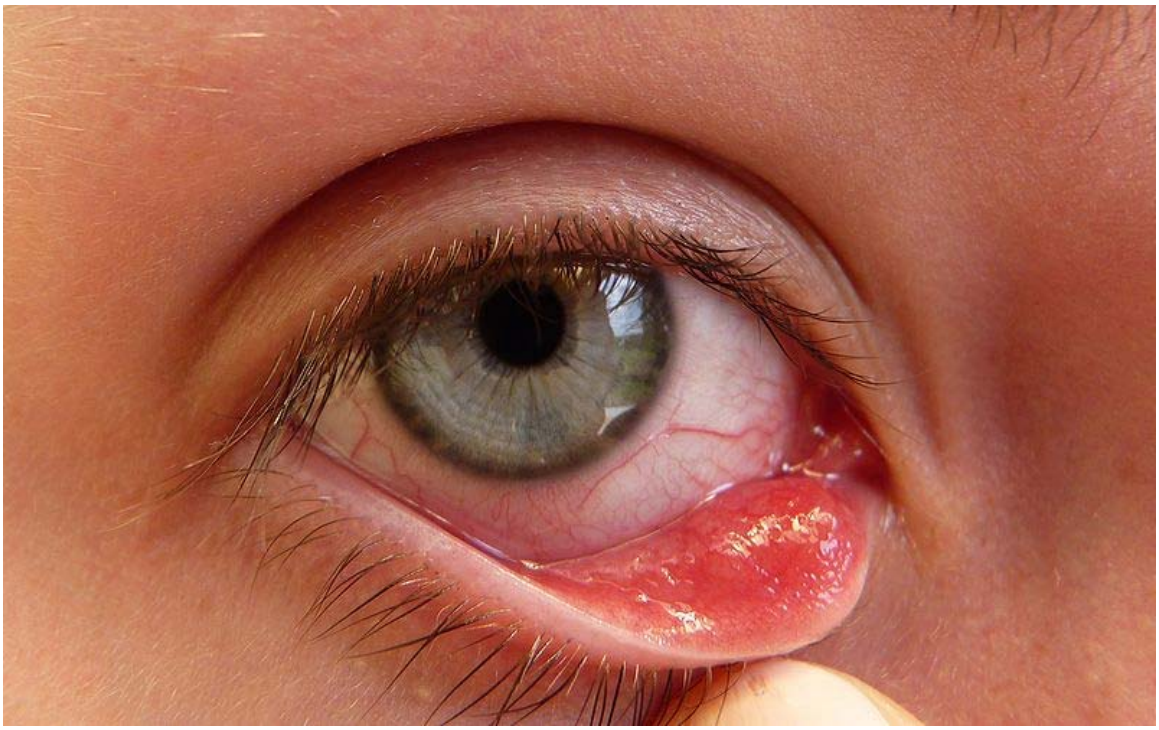
Other stye symptoms may include:

- A lump on the top or bottom eyelid
- Localized swelling of the eyelid
- Localized pain
- Redness
- Tenderness to touch
- Crusting of the eyelid margins
- Burning in the eye
- Droopiness of the eyelid
- Scratchy sensation on the eyeball
- Blurred vision
- Mucous discharge in the eye
- Irritation of the eye

- Light sensitivity
- Tearing
- Discomfort during blinking
- Sensation of a foreign body in the eye

Treatment

As styes usually go away within a week, most patients do not require medical treatment. The first steps in treating styes include applying cool compresses on the affected eye four to six times a day for approximately 15 minutes until the pain subsides. After about 3–5 days, once the eyelid is no longer painful, warm compresses can help the drainage and hasten the curing process. Warm compresses during initial swelling will increase swelling.



Stye on lower part of a person's eye

As a part of self-care at home, patients may cleanse the affected eyelid with tap water or with a mild, nonirritating soap or shampoo (such as baby shampoo) to help clean crusted discharge. Cleansing must be done gently and while the eyes are closed to prevent eye injuries.

Patients are highly advised to not squeeze or puncture the stye, as serious infection can occur as a result. The infection could spread to the surrounding tissues and areas.

Eye stye sufferers should avoid eye makeup (e.g., eyeliner), lotions and wearing contact lenses, since these can aggravate and spread the infection (sometimes to the cornea).

Medical treatment can also be provided by a doctor and it is aimed on relieving the symptoms. Pain relievers such as acetaminophen may be prescribed and in some cases, antibiotics may be needed. Antibiotics are normally given to patients with multiple styes or with styes that do not seem to heal, and to patients who also suffer from blepharitis or rosacea. Commonly, the ophthalmologist prescribes oral or intravenous antibiotics, such as doxycycline, only when the infection has spread. Topical antibiotic ointments or antibiotic/steroid combination ointments can also be administered in stye treatment.

Surgery is the last resort in stye treatment. Styes that do not respond to any type of therapies are usually surgically removed. Stye surgery is performed by an ophthalmologist, and generally under local anesthesia. The procedure consists of making a small incision on the inner or outer surface of the eyelid, depending if the stye is pointing externally or not. After the incision is made, the pus is drained out of the gland, and very small and unnoticeable sutures are used to close the lesion. It is common for the removed stye to be biopsied to rule out the possibility of skin cancer.

Complications

Stye complications occur in very rare cases. However, the most frequent complication of styes is progression to a chalazion that causes cosmetic deformity, corneal irritation, and often requires surgical removal. Complications may also arise from the improper surgical lancing, and mainly consist of disruption of lash growth, lid deformity or lid fistula. Styes that are too large may interfere with one's vision.

Eyelid cellulitis is another potential complication of eye styes, which is a generalized infection of the eyelid. Progression of a stye to a systemic infection (spreading throughout the body) is extremely rare, and only a few instances of such spread have been recorded.

Prognosis

Although styes are harmless in most cases and complications are very rare, styes often recur. They do not cause intraocular damage, meaning they do not affect the eye. Styes normally heal on their own by rupturing within few days to a week, causing the relief of symptoms. Few people require surgery as part of stye treatment. With adequate treatment, styes tend to heal quickly and without arising any type of complications.

The prognosis is better if one does not attempt to squeeze or puncture the stye, as infection may spread to adjacent tissues. A stye usually will heal within a few days to a week, but if it does not improve or it worsens within two weeks, a doctor's opinion should be sought. Also, patients are recommended to call a doctor if they encounter problems with vision, the eyelid bumps becomes very painful, the stye bleeds or reoccurs or the eyelid or eye becomes red.

Prevention

Stye prevention is closely related to proper hygiene. Proper hand washing can not only reduce the risks of developing styes, but also all other types of infections.

Upon awakening, application of a warm washcloth to the eyelids for one to two minutes may be beneficial in decreasing the occurrence of styes by liquefying the contents of the oil glands of the eyelid and thereby preventing blockage. Some studies suggest oral flaxseed supplementation to prevent the occurrence of styes.

To prevent developing styes, it is recommended to never share cosmetics or cosmetic eye tools with other people. People should also keep their eye tools clean and generally practice proper eye hygiene. Women are recommended to remove makeup every night before going to sleep and discard old or contaminated eye makeup.

Chapter 3

Chalazion

Chalazion

Eyelid affected by Chalazion

ICD-10	H00.1
ICD-9	373.2
DiseasesDB	6009
MedlinePlus	001006
eMedicine	emerg/94 oph/243
MeSH	D017043

A **chalazion** also known as a **meibomian gland lipogranuloma**, is a cyst in the eyelid that is caused by inflammation of a blocked meibomian gland, usually on the upper eyelid. Chalazia differ from styes (hordeola) in that they are subacute, nontender, and usually painless nodules. They may become acutely inflamed but, unlike a stye, chalazia usually point inside the lid rather than on the lid margin. A chalazion or meibomian cyst could take months to fully heal with treatment and could take years to heal without any major complications.

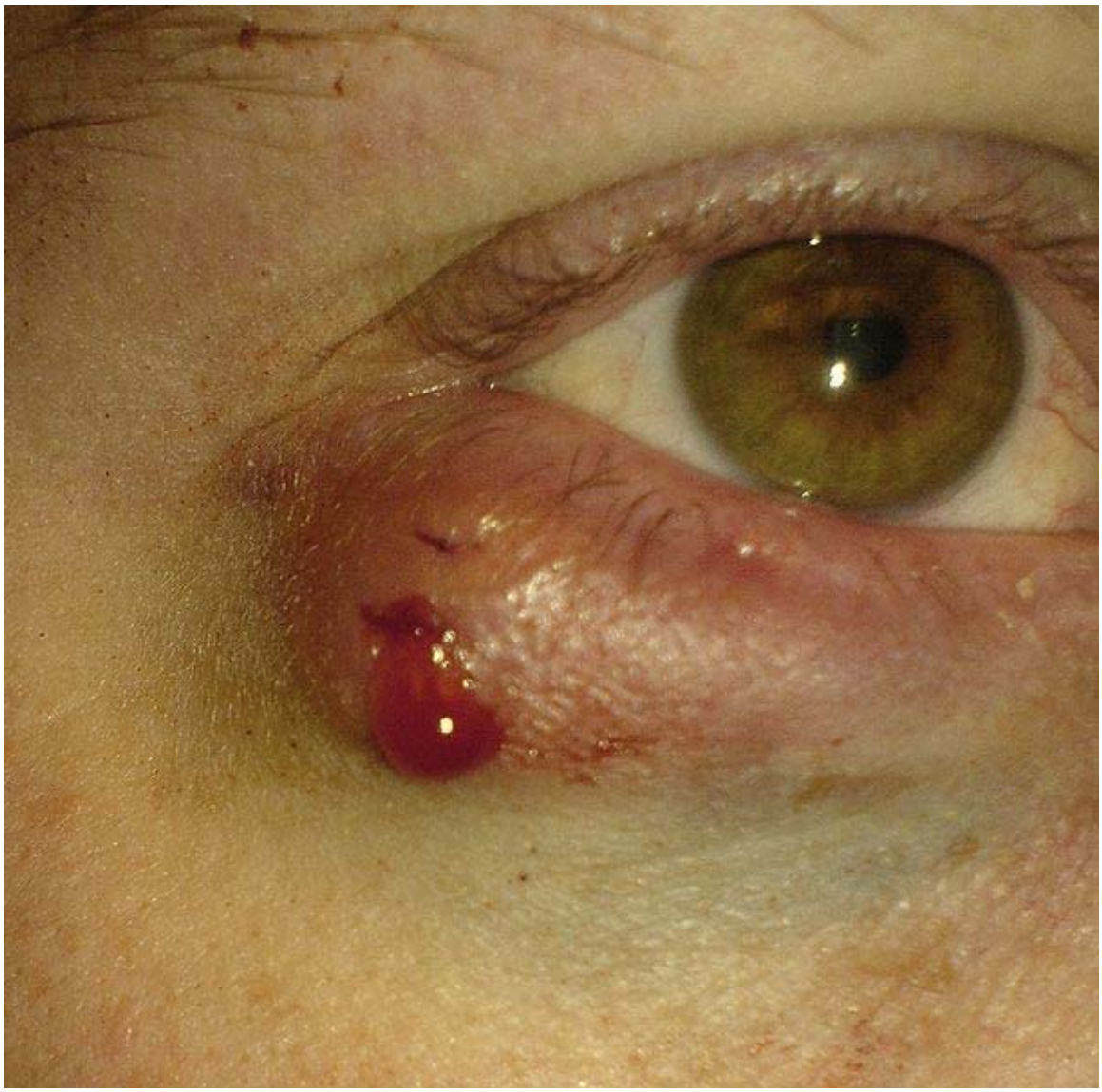
Signs and symptoms

- Swelling on the eyelid
- Eyelid tenderness
- Sensitivity to light
- Increased tearing

- Heaviness of the eyelid

A chalazion or meibomian cyst can sometimes be mistaken for a stye.

Treatment



A large chalazion ca. 20 minutes upon excision. This bipartite chalazion was removed via two separate incisions. Further along the lower eyelid, signs of chronic inflammation (Blepharitis) are visible.

Topical antibiotic eye drops or ointment (e.g. chloramphenicol or fusidic acid) are sometimes used for the initial acute infection, but are otherwise of little value in treating a chalazion. Chalazia will often disappear without further treatment within a few months and virtually all will resorb within two years.

If they continue to enlarge or fail to settle within a few months, then smaller lesions may be injected with a corticosteroid or larger ones may be surgically removed using local anesthesia. This is usually done from underneath the eyelid to avoid a scar on the skin. If the chalazion is located directly under the eyelid's outer tissue, however, an excision from above may be more advisable so as not to inflict any unnecessary damage on the lid itself. Eyelid epidermis usually mends well, without leaving any visible scar. Depending on the chalazion's texture, the excision procedure varies: while fluid matter can easily be removed under minimal invasivion, by merely puncturing the chalazion and exerting pressure upon the surrounding tissue, hardened matter usually necessitates a larger incision, through which it can be scraped out. Any residual matter should be metabolized in the course of the subsequent healing process, generally aided by regular appliance of dry heat. The excision of larger chalazia may result in visible hematoma around the lid, which will wear off within three or four days, whereas the swelling may persist for longer. Chalazion excision is an ambulant treatment and normally does not take longer than fifteen minutes. Nevertheless, owing to the risks of infection and severe damage to the eyelid, such procedures should only be performed by a medical professional.

A home remedy is to have a hot, wet flannel, and rub gently, until the heat has reached the cyst. this helps to reduce the size, and eventually it will be rid of. It is rare that a chalazion will recur and they will usually be biopsied to rule out the possibility of a tumour.

Complications

A large chalazion can cause astigmatism due to pressure on the cornea.

As laser eye surgery involves shaping the cornea by burning parts of it away, weakening its structure, post operation patients can be left predisposed to deformation of the cornea from small chalazia.

Complications including, but not limited to hypopigmentation may occur with corticosteroid injection.

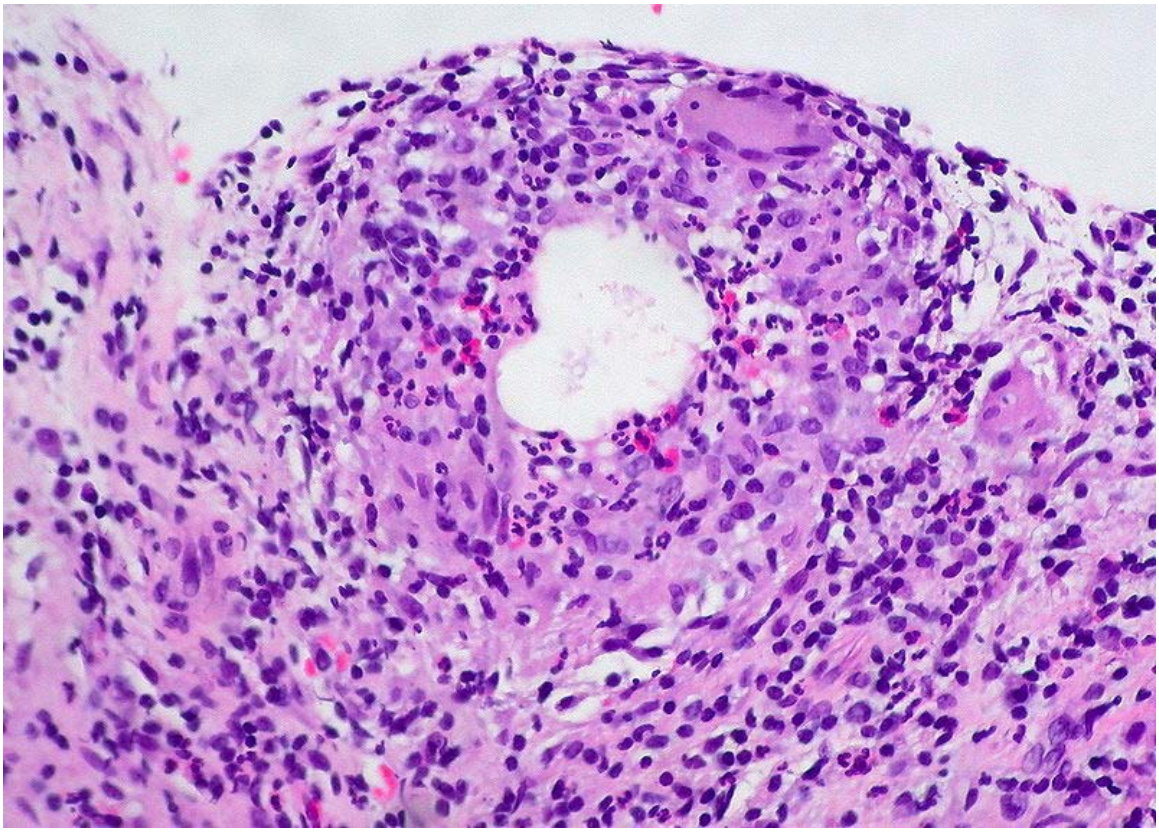
Recurring chalazia in the same area may sometimes be a symptom of sebaceous cell carcinoma, albeit rarely. This is a type of cancer.

Sometimes, as a last resort, surgery is performed. The eyelid is injected with a local anesthetic, a clamp is put on the eyelid, then the eyelid is turned over, an incision is made on the inside of the eyelid, and the chalazion is drained and scraped out. A scar on the upper lid can cause discomfort as some patients feel the scar as they blink. Of course as surgeries are intrusive and damage healthy tissue (e.g. leaving behind scar tissue or possibly even causing blepharitis), given other options, less intrusive treatment is always preferable. Similarly, chalazia may recur once the eye is predisposed and surgical intervention each time is not possible. So surgery should be considered only as a last resort, performed on as few as 5% of all chalazia patients.

Chalazion surgery

Chalazion surgery is normally performed by an ophthalmologist at an eye hospital. This type of surgery is a simple procedure which is generally performed as a day operation and the patient does not need to remain in the hospital for further medical care.

Chalazion removal surgery is performed under local or general anesthesia. Commonly, general anesthesia is administered in children to make sure they stay still and no injury to the eye occurs. Local anesthesia is used in adults and it is applied with a small injection into the eyelid. The discomfort of the injection is minimized with the help of an anesthetic cream which is applied locally.



Classic lipogranulomatous response seen in a well-developed chalazion

The chalazion may be removed in two ways, depending on the size of cyst. Relatively small chalazia are removed through a small cut at the back of the eyelid. The surgeon lifts the eyelid so he can have access to the back of its surface and makes an incision of approximately 3mm just on top of the chalazion. The lump is then removed and pressure is applied for a few minutes to stop any oozing of blood that may occur because of the operation. Surgery of small chalazia does not require stitches as the cut is at the back of the eyelid and therefore the cut cannot be seen and the cosmetic result is excellent.

Larger chalazia are removed through an incision in front of the eyelid. Larger chalazia usually push on the skin of the eyelid and this is the main reason why doctors prefer removing them this way. The cut is not larger than 3 mm and it is performed on top of the chalazion. The lump is removed and then pressure is applied on the incision so oozing is prevented. This type of surgery is closed with very fine stitches. They are hardly visible and they are usually removed within a week after the surgery has been performed. Although chalazia are rarely dangerous, every removed chalazion is sent to the laboratory to be examined under a microscope because very rarely it can harbor cancer.

When surgery for chalazion is considered, patients who take aspirin or any medication that contains aspirin are advised to stop taking them one week before the procedure as they may cause bleeding. There are several tests taken prior the surgery to make sure the patient is in good condition for the operation.

In rare cases, patients are kept overnight in the hospital after chalazion surgery. These include cases in which complications occurred and the patient needs to be closely monitored. In most cases however, patients are able to go home after the operation has ended.

The recovery process is easy and quite fast. Most patients experience some very minor discomfort in the eye which can be easily controlled by taking painkilling medication. Patients are however recommended to avoid getting water in the eye for up to 10 days after surgery, they may wash, bathe or shower but they must be careful in keeping the area dry and clean. Makeup may be worn after one month after surgery. Patients who wear contact lenses are recommended to not wear on in the operated side for at least eight weeks to prevent infection and potential complications.

Commonly, patients receive eye drops to prevent infection and swellings in the eye and pain medication that will help them cope with the pain and discomfort in the eyelid and eye. One can use paracetamol/tylenol rather than aspirin to control the pain. Also, after surgery, a pad and protective plastic shield are used to apply pressure on the eye in order to prevent leakage of blood after the operation and which may be removed 6 to 8 hours after the procedure.

People who underwent chalazion surgery are normally asked to check up their operation three to four weeks after surgery has been performed. They may start driving the day after surgery and they may get back to work in one or two days.

Chalazion surgery is a safe procedure and complications occur very seldom. Serious complications that require another operation to be fixed are also very rare. Among potential complications, although rare, there is infection, bleeding or the recurrence of the chalazion.

Chapter 4

Blepharitis

Blepharitis

An infant with mild blepharitis on his right side

ICD-10	H01.0
ICD-9	373.0
DiseasesDB	1455
eMedicine	oph/81
MeSH	D001762

Blepharitis is an ocular condition characterized by chronic inflammation of the eyelid, the severity and time course of which can vary. Onset can be acute, resolving without treatment within 2–4 weeks (this can be greatly reduced with lid hygiene), but more generally is a long standing inflammation varying in severity. It may be classified as seborrhoeic, staphylocccal, mixed, posterior or meiobomitis, or parasitic.

Signs and symptoms

Signs and symptoms that are associated with the chronic inflammation can be;

- Redness of the eyelids.
- Flaking of skin on the lids.
- Crusting at the lid margins, this is generally worse on waking.
- Cysts at the lid margin (hordeolum).

- Red eye.
- Debris in the tear film, seen under magnification (improved contrast with use of fluorescein drops).
- Gritty sensation of the eye.
- Reduced vision.

Common signs and symptoms of blepharitis also include itching, irritation and burning as well as a foreign body sensation. Some patients experience eye dryness, which can cause a certain degree of discomfort.

People who wear contact lenses usually have more trouble in coping with their symptoms because although they need contact-lenses, they cannot wear them. Many such patients complain of being unable to wear their lenses for long periods of time or that the lenses are causing them even more irritation of the eye.

Also, the lids may become red and may have ulcerative, non-healing areas which may actually bleed. Blepharitis does not tend to cause problems with the patient's vision whatsoever, but due to a poor tear film, one may experience blurred vision.

Eye redness and swelling tend to appear in more advanced cases, and they are rarely primary symptoms. The symptoms can slightly vary based on the exact cause of the condition. Blepharitis due to an allergy can cause dark lids, symptom which is known as "allergic shiner" and which tends to be more frequent in children rather than adults. Infectious blepharitis is accompanied by a yellow- or green-colored discharge which is more abundant in the morning and which leads to stuck lids. Blepharitis may also cause eyelid matting or "gluing" of the lashes. Not least, dandruff may occur on the scalp or on the eyebrows.

Other blepharitis symptoms include sensitivity to light, eyelashes that grow abnormally or even loss of eyelashes. Also, the tears might seem frothy or bubbly in nature and mild scarring might occur to the eyelids. The symptoms and signs of blepharitis are often erroneously ascribed by the patient as being due to "recurrent conjunctivitis".

Blepharitis that localizes in the skin of the eyelids may cause styes or chalazia, which appear like red bumps, sometimes with a yellow spot if infection is present. Although pain is not common among blepharitis symptoms, if the condition persists or becomes painful, the individual is recommended to seek medical attention.

Chronic blepharitis may result in damage of varying severity which may have a negative effect upon vision and therefore upon the eyeglass prescription.

Infectious blepharitis can cause hard crusts around the eyelashes which leave small ulcers that may bleed or ooze after cleaning.

As a general rule, blepharitis whose symptoms do not improve despite good hygiene consisting of proper cleaning and care of the eye area, should be referred to a doctor.

Staphylococcal blepharitis

Staphlycoccal blepharitis is caused by infection of the anterior portion of the eyelid by Staphylococcal bacteria. As the infection progresses, the sufferer may begin to notice a foreign body sensation, matting of the lashes, and burning. Usually, the primary care physician will prescribe topical antibiotics for staphylococcal blepharitis, as this is an acute condition and should heal quickly. The condition can sometimes lead to a chalazion or a stye.

Staphylococcal blepharitis is a more severe condition which may start in childhood and continue through adulthood. It is commonly recurrent and it requires special medical care. The prevalence of Staphylococcus aureus in the conjunctival sac and on the lid margin varies among countries, apparently according to climate.

In cases of blepharitis caused by Staphylococcus aureus, the presence of a collarette (a ring-like formation around the lash shaft) can be observed. This is the main sign of this particular condition. Other specific symptoms include loss of eyelashes or broken eyelashes.

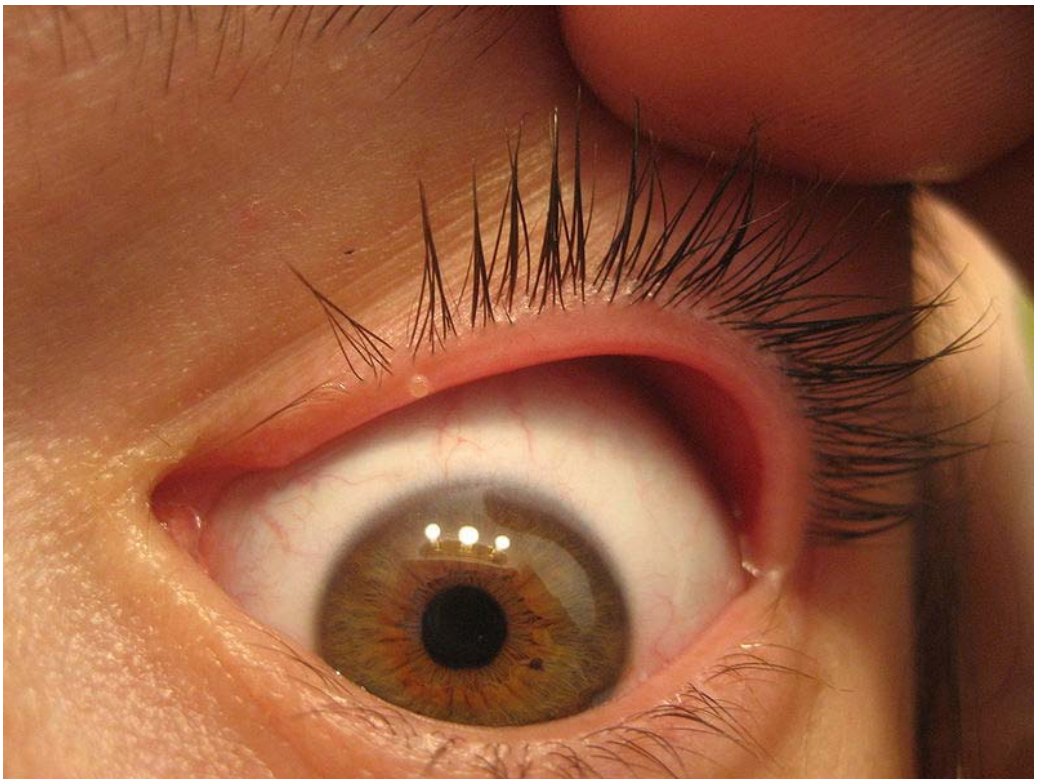
Staphylococcal blepharitis is mainly diagnosed upon the patient's medical history and a bacterial culture. However, since blepharitis is a condition that is completely understood, diagnosis usually consists of establishing an accurate medical history of the patient and a proper physical examination. Blepharitis is diagnosed primarily upon physical examination and rarely further tests are needed. Yet, it is recommended that older patients or those who are at risk of developing certain skin conditions undergo biopsy in order to remove the possibility of tumor.

This type of blepharitis is more likely to occur in individuals who are exposed to bacteria. Staphylococcal blepharitis, unlike the other types of this condition, seems to be seen more frequently in women than men.

It is important that this type of blepharitis is properly treated because otherwise the infection may spread to other parts of the eye or to the scarring of the cornea. Staphylococcal blepharitis is normally treated with antibiotics such as Chloramphenicol ointment. Fusidic acid is usually the choice of antibiotics in cases when Chloramphenicol is contraindicated. Antibiotics are given for at least four weeks and up to six weeks, which is considered enough so the infection is completely cured. Also, blepharitis treatment includes a short course of topical steroids which are administered to control the inflammation.

Staphylococcal blepharitis, along with the other types of blepharitis treatment is only effective if given at the same time with extra cautious eyelid hygiene. This consists of eyelid proper cleaning and removing crusts and debris and maintaining the hygiene by avoiding expired make up or cosmetics that are used around the eye. Particularly in blepharitis caused by S. aureus, patients are recommended to keep the eyelids area clean to avoid spreading the infection when oozing and bleeding occur.

Posterior blepharitis or *rosacea-associated* blepharitis



"Internal hordeolum"

The most common type of blepharitis is often found in people with a Rosacea skin type. The oil glands in the lid of rosacea sufferers secrete a modified oil which leads to inflammation at the meibomian gland openings which are found at the edge of the lid.

Treatment and management

The single most important treatment principle is a daily routine of lid margin hygiene, as described below. Such a routine needs to be convenient enough to be continued for life to avoid relapses as blepharitis is often a chronic condition. But it can be acute, and one episode does not mean it is a life-long condition.

A typical lid margin hygiene routine consists of four steps:

1. Softening of lid margin debris and oils: Apply a warm wet compress to the lids - such as a washcloth with hot water - for about two minutes. New, dry, warm compress masks can be conveniently warmed in a micro-wave oven and maintain a comfortable 40C temperature for 10 minutes while the waxy oils blocking the glands are cleared.
2. Mechanical removal of lid margin debris: At the end of a shower routine, wash your face with a wash cloth. Use facial soap or non-burning baby shampoo (make sure to dilute the soap solution 1/10 with water first). Gently and repeatedly rub

along the lid margins while eyes are closed. Too much soap or shampoo may remove the essential oily layer of the eyes' own tear film and create further problems with dry eye discomfort.
3. Antibiotic reduction of lid margin bacteria (at the discretion of your physician): After lid margin cleaning, spread small amount of prescription antibiotic ophthalmic ointment with finger tip along lid fissure while eyes closed. Use prior to bed time as opposed to in the morning to avoid blurry vision.
4. Avoid the use of eye make-up until symptoms subside.

An alternative after washing is to coat the eyelids with a good quality hair conditioner. Leave in place for several minutes then rinse. The conditioner seems to break the bacteria / crusting /irritation cycle, and daily use can eliminate the symptoms.

The following guide is very common but is more challenging to perform by visually disabled or frail patients as it requires good motor skills and a mirror. Compared with the above, it does not bear any advantages:

1. Apply hot compresses to both eyes for five minutes once to twice per day. The "hot wet flannel" treatment is often recommended, however this does not stay hot for long enough, is very messy & potentially unhygienic. A very effective alternative is a flax filled mask which is heated in a microwave oven.
2. After hot compresses, in front of a mirror, use a moist cotton bud soaked in a cup of water with a drop of baby shampoo, or a drop of antibiotic such as Chloramphenicol. Rub along the lid margins while tilting the lid outward with the other hand.
3. In front of mirror, place small drop of antibiotic ophthalmic ointment (e.g. erythromycin) in lower conjunctival sac while pulling lid away from eye with other hand.

Often the above is advised together with mild massage to mechanically empty glands located at the lid margin (Meibomian glands, Zeis glands, Moll glands).

Dermatologists treat blepharitis similarly to seborrhoeic dermatitis by using a safe topical anti-inflammatory medication like sulfacetamide or brief courses of a mild topical steroid. Although anti-fungals like ketoconazole (Nizoral) are commonly prescribed for seborrhoeic dermatitis, dermatologists and optometrists usually do not prescribe anti-fungals for seborrhoeic blepharitis.

If these conventional treatments for blepharitis do not bring relief, patients may consider allergy testing and ocular antihistamines. Allergic responses to dust mite feces and other allergens can cause lid inflammation, ocular irritation, and dry eyes. Prescription ocular antihistamines and over-the-counter ocular antihistamines are very safe and can bring almost immediate relief to patients whose lid inflammation is caused by allergies.

Chapter 5

Pterygium (Conjunctiva)

Pterygium (conjunctiva)

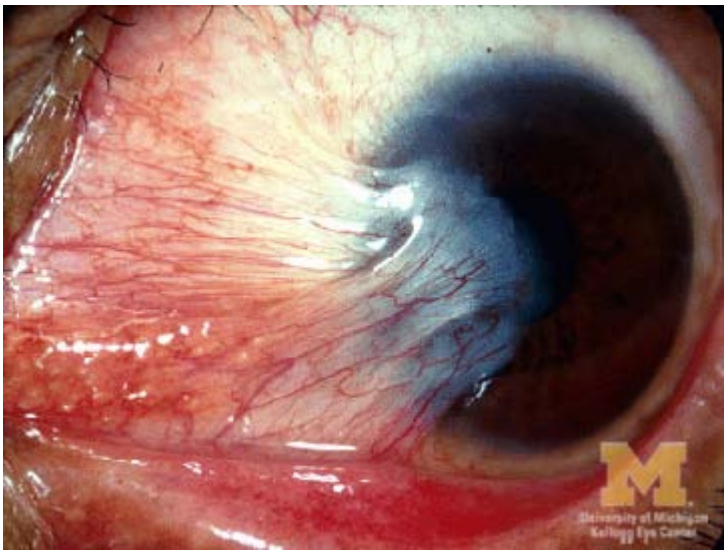
Pterygium removal surgery

ICD-10	H11.0
ICD-9	372.4
DiseasesDB	10916
MedlinePlus	001011
eMedicine	oph/542
MeSH	D011625

Pterygium (Surfer's Eye) most often refers to a benign growth of the conjunctiva. A pterygium commonly grows from the nasal side of the sclera. It is associated with, and thought to be caused by ultraviolet-light exposure (e.g., sunlight), low humidity, and dust. The predominance of pterygia on the nasal side is possibly a result of the sun's rays passing laterally through the cornea, where it undergoes refraction and becomes focused on the limbic area. Sunlight passes unobstructed from the lateral side of the eye, focusing on the medial limbus after passing through the cornea. On the contralateral (medial) side,

however, the shadow of the nose medially reduces the intensity of sunlight focused on the lateral/temporal limbus.

Pathology



Pterygium growing onto the cornea

Pterygium in the conjunctiva is characterized by elastotic degeneration of collagen (actinic elastosis) and fibrovascular proliferation. It has an advancing portion called the head of the pterygium, which is connected to the main body of the pterygium by the neck. Sometimes a line of iron deposition can be seen adjacent to the head of the pterygium called *Stocker's line*. The location of the line can give an indication of the pattern of growth.

The exact cause is unknown, but it is associated with excessive exposure to wind, sunlight, or sand. Therefore, it is more likely to occur in populations that inhabit the areas near the equator, as well as windy locations. In addition, pterygia are twice as likely to occur in men than women.

Some research also suggests a genetic predisposition due to an expression of vimentin, which indicates cellular migration by the keratoblasts embryological development, which are the cells that give rise to the layers of the cornea. These cells also exhibit an increased P53 expression likely due to a deficit in the tumor suppressor gene. These indications give the impression of a migrating limbus because the cellular origin of the pterygium is actually initiated by the limbal epithelium.

The pterygium is composed of several segments:

- Fuchs' Patches (minute gray blemishes that disperse near the pterygium head)
- Stocker's Line (a brownish line composed of iron deposits)
- Hood (fibrous nonvascular portion of the pterygium)

- Head (apex of the pterygium, typically raised and highly vascular)
- Body (fleshy elevated portion congested with tortuous vessels)
- Superior Edge (upper edge of the triangular or wing-shaped portion of the pterygium)
- Inferior Edge (lower edge of the triangular or wing-shaped portion of the ptyerygium).

Prevention

As it is associated with excessive sun or wind exposure, wearing protective sunglasses with side shields and/or wide brimmed hats and using artificial tears throughout the day may help prevent their formation or stop further growth. Surfers and other water-sport athletes should wear eye protection that blocks 100% of the UV rays from the water, as is often used by snow-sport athletes.

Symptoms

Symptoms of pterygium include persistent redness, inflammation, foreign body sensation, tearing, which can cause bleeding, dry and itchy eyes. In advanced cases the pterygium can affect vision as it invades the cornea with the potential of obscuring the optical center of the cornea and ***inducing' astigmatism*****and corneal scarring.**

Treatment

Today a variety of options are available for the management of pterygium, from irradiation, to conjunctival auto-grafting or amniotic membrane transplantation, along with glue and suture application. As it is a benign growth, pterygium typically does not require surgery unless it grows to such an extent that it covers the pupil, obstructing vision or presents with acute symptoms. Some of the irritating symptoms can be addressed with artificial tears. However, no reliable medical treatment exists to reduce or even prevent pterygium progression. Definitive treatment is achieved only by surgical removal. Long-term follow up is required as pterygium may recur even after complete surgical correction.

If there is recurrence after surgery or if recurrence of pterygium is thought to be vision threatening, it is possible to use strontium (^{90}Sr) plaque therapy. ^{90}Sr is a radioactive substance that produces beta particles, which penetrate a very short distance into the cornea at the site of the operation. It suppresses the regrowth of blood vessels that occur with return of the pterygium. The treatment requires some local anaesthetic in the eye and is best done at the time of, or on the same day as the pterygium excision.

The ^{90}Sr plaque is a concave metal disc about 1-1.5 cm in diameter that is hollow and filled with an insoluble strontium salt. The side placed on the eye is a very thin and delicate silver film that will contain the strontium but allow the beta particles to escape. The dose of radiation to the conjunctiva is controlled by the time that the plaque is left in contact with the surface. The integrity of the plaque surfaces is paramount to prevent

exposure to patients and so is wipe tested to see if radioactive matter is escaping. Obviously this test must be done very very gently.

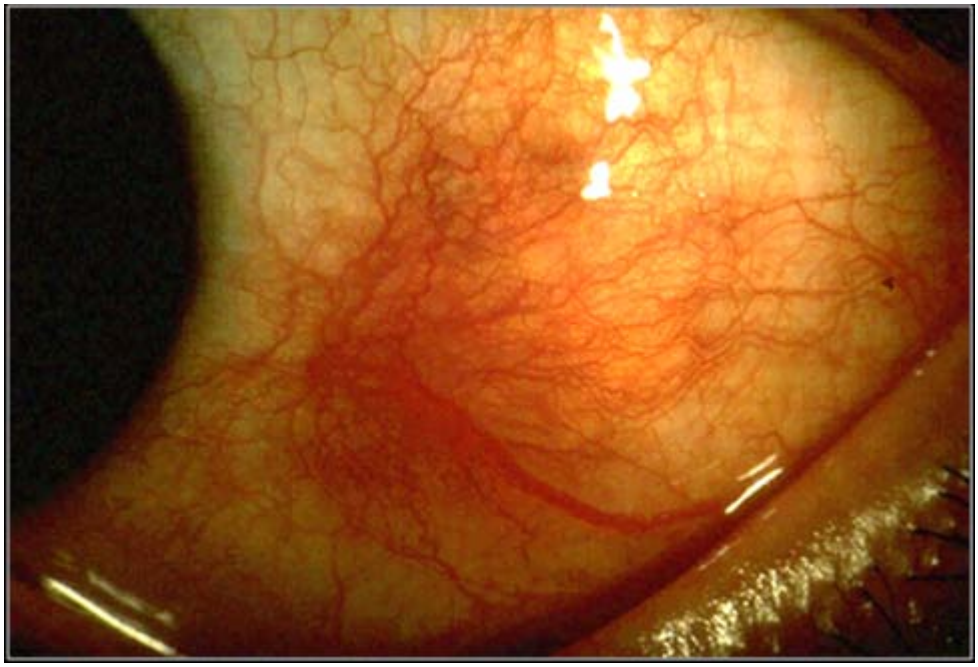
Conjunctival auto-grafting is a surgical technique that is effective and safe procedure for pterygium removal. When the pterygium is removed, the tissue that covers the sclera known as the conjunctiva is also extracted. Auto-grafting replaces the bare sclera with tissue that is surgically removed from the inside of the patients' upper eyelid. That "self-tissue" is then transplanted to the bare sclera and is fixated using sutures, tissue adhesive, or glue adhesive.

Amniotic membrane transplantation is an effective and safe procedure for pterygium removal. Amniotic membrane transplantation offers practical alternative to conjunctival auto graft transplantation for extensive pterygium removal. Amniotic membrane transplantation is tissue that is acquired from the innermost layer of the human placenta and has been used to replace and heal damaged mucosal surfaces including successful reconstruction of the ocular surface. It has been used as a surgical material since the 1940s, and has been shown to have a strong anti-adhesive effect. Using an amniotic graft facilitates epithelialization, and has anti-inflammatory as well as surface rejuvenation properties. Amniotic membrane transplantation can also be fixated to the sclera using sutures, or glue adhesive. Amniotic membrane transplantation with Tisseel glue application and Mitomycin-C has shown excellent cosmetic outcomes with a surface free of redness, stitching, or patches, which makes the ocular surface suitable for vision correction surgery sooner.

Chapter 6

Scleritis and Subconjunctival Hemorrhage

Scleritis



Scleritisis: a inflammation of entire thickness of the sclera.

Scleritis	
ICD-10	H15.0
ICD-9	379.0
DiseasesDB	11898
MedlinePlus	001003 scleritis. *001019* episcleritis

eMedicine emerg/521 oph/642

MeSH D015423

Scleritis is a serious inflammatory disease that affects the white outer coating of the eye, known as the sclera. The disease is often contracted through association with other diseases of the body, such as Wegener's granulomatosis or rheumatoid arthritis; it can also be attained through disorders of menstruation. For this reason, scleritis occurs frequently among young women. There are three types of scleritis: **diffuse scleritis** (the most common), **nodular scleritis**, and **necrotizing scleritis** (the most severe). Scleritis may be the first symptom of onset of connective tissue disease.

Episcleritis is inflammation of the episclera, a less serious condition that seldom develops into scleritis.

Signs and symptoms

Symptoms of the disease include:

- Redness of the sclera and conjunctiva, sometimes changing to a purple hue
- Severe ocular pain (not present in episcleritis) which may radiate to the temple or jaw
- Photophobia and tearing
- Decrease in visual acuity, possibly leading to blindness
- a slight loss of vision of the eye

Diagnosis

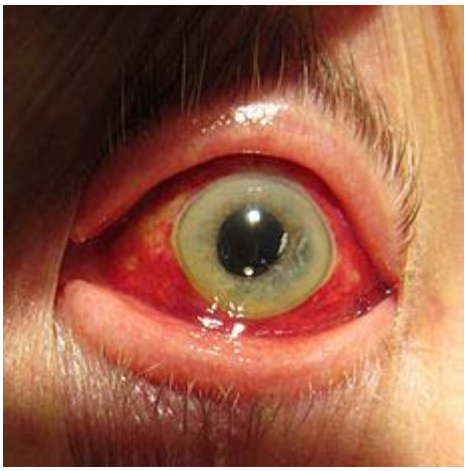
Scleritis is best detected by examining the sclera in daylight; retracting the lids helps determine the extent of involvement. Other aspects of the eye exam (i.e. visual acuity testing, slit lamp examination, etc) can be normal. Ancillary tests CT scans, MRIs, and ultrasonographies can be helpful, but do not replace the physical examination.

Treatment

In very severe cases of necrotizing scleritis, eye surgery must be performed to repair damaged corneal tissue in the eye and preserve the patient's vision. For less severe cases, nonsteroidal anti-inflammatory drugs, such as ibuprofen, are prescribed for pain relief. Scleritis itself is treated with an oral medication containing corticosteroids and an eye solution. In some cases, antibiotics are prescribed. Simply using eye drops will not treat scleritis. In more aggressive cases of scleritis, chemotherapy (such as systemic immunosuppressive therapy with such drugs as cyclophosphamide or azathioprine) may be used to treat the disease. If not treated, scleritis can cause blindness.

Subconjunctival hemorrhage

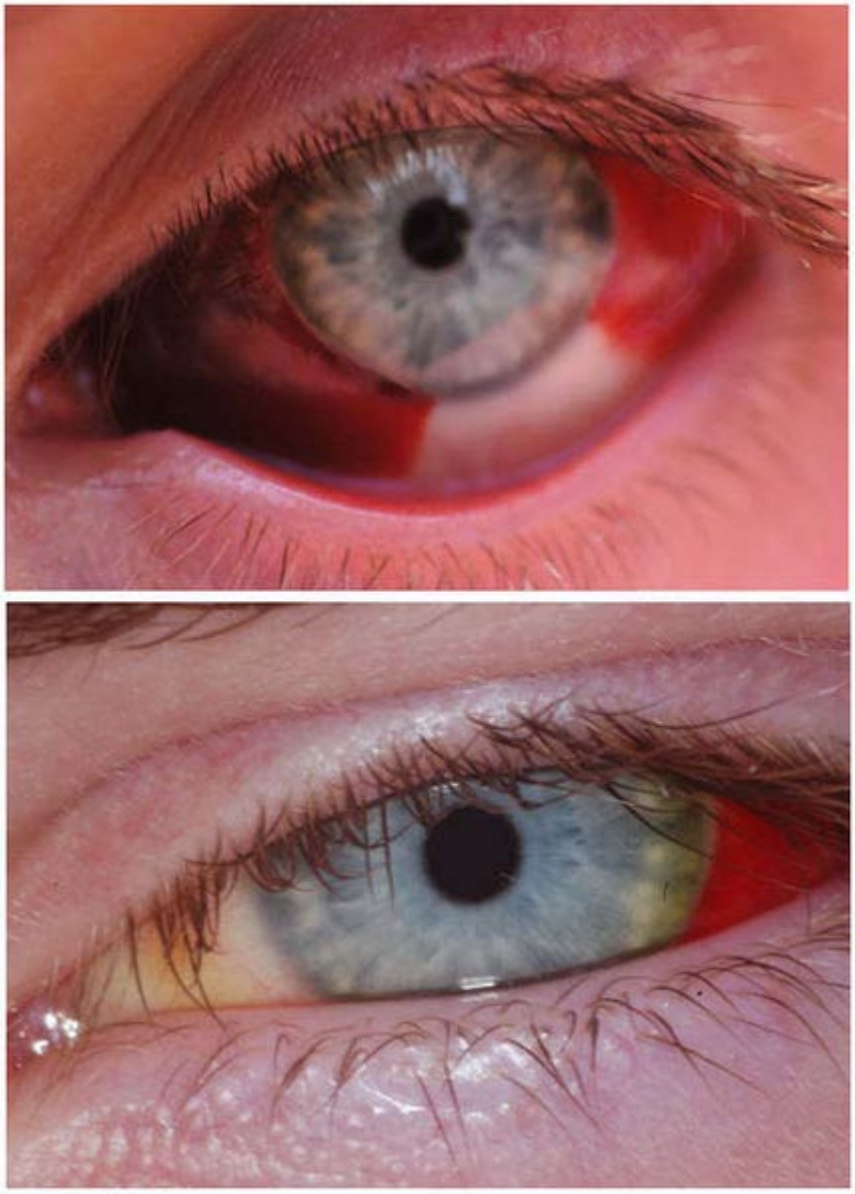
Subconjunctival hemorrhage



Subconjunctival hemorrhage causing red coloration as result of ruptured blood vessel in the eye.

ICD-10 H11.3

ICD-9 372.72



(Top) A stress induced subconjunctival hemorrhage in the left eye one week after hemorrhaging. (Bottom) Same hemorrhage four weeks after hemorrhaging. Some of the blood in the sclera has turned yellow, like a bruise.

A **subconjunctival hemorrhage** (or **subconjunctival haemorrhage**) also known as hyposphagma, is bleeding underneath the conjunctiva. The conjunctiva contains many small, fragile blood vessels that are easily ruptured or broken. When this happens, blood leaks into the space between the conjunctiva and sclera.

Whereas a bruise typically appears black or blue underneath the skin, a subconjunctival hemorrhage initially appears bright red underneath the transparent conjunctiva. Later the hemorrhage may spread and become green or yellow, like a bruise. Usually this disappears within 2 weeks.

Although its appearance may be alarming, a subconjunctival hemorrhage is generally a painless and harmless condition; however, it may be associated with high blood pressure, trauma to the eye, or a base of skull fracture if there is no posterior border of the hemorrhage visible.

Causes

- Blood dyscrasia (rare)
- Blood thinners, such as ginger, capsaicin, ginseng, garlic, aspirin, or *Herba* if taken in high doses or combined. These can also make the vessels in the eye more susceptible to the pressure causes listed above.
- Diving accidents - Mask Squeeze (volume inside in mask creates increased pressure with increased depth)
- Severe hypertension
- LASIK
- Leptospirosis
- Minor eye trauma
- Spontaneously with increased venous pressure
    - Choking
    - Coughing
    - Pulling extreme g-forces
    - Sneezing
    - Straining
    - Strenuous Exercising
    - Touching/widening eyes
    - Vomiting
- Prolonged stress
- Severe thoracic trauma, leading to increased pressure in the extremities, including around the eyes.

Subconjunctival hemorrhages in infants may be associated with scurvy (a vitamin C deficiency), abuse or traumatic asphyxia syndrome.

Treatment and management

A subconjunctival hemorrhage is typically a self-limiting condition that requires no treatment in the absence of infection or significant trauma. The elective use of aspirin and NSAIDs is typically discouraged.

A common symptom of a subconjunctival hemorrhage, itchy eyes, is often treated by applying eye drops or artificial tears to the affected eye(s), however, this is discouraged, as it may slow down the healing process.

Chapter 7

Keratitis and Corneal Ulcer

Keratitis

Keratitis

An eye with non-ulcerative sterile keratitis.

ICD-10	H16.
ICD-9	370
DiseasesDB	7150
MeSH	D007634

Keratitis is a condition in which the eye's cornea, the front part of the eye, becomes inflamed. The condition is often marked by moderate to intense pain and usually involves impaired eyesight.

Types

Superficial keratitis involves the superficial layers of the cornea. After healing, this form of keratitis does not generally leave a scar.

Deep keratitis involves deeper layers of the cornea, and the natural course leaves a scar upon healing that impairs vision if on or near the visual axis. This can be reduced or avoided with the use of topical corticosteroid eyedrops.

Causes

Keratitis has multiple causes, one of which is an infection of a present or previous herpes simplex virus secondary to an upper respiratory infection, involving cold sores.

Pathogens

- Amoebic keratitis. Amoebic infection of the cornea is the most serious corneal infection, usually affecting contact lens wearers. It is usually caused by Acanthamoeba. On May 25, 2007, the CDC issued a health advisory due to increased risk of Acanthamoeba keratitis (AK) associated with use of Advanced Medical Optics (AMO) Complete Moisture Plus Multi-Purpose eye solution.
- Bacterial keratitis. Bacterial infection of the cornea can follow from an injury or from wearing contact lenses. The bacteriums usually involved are Staphylococcus aureus and for contact lens wearers Pseudomonas aeruginosa.
- Fungal keratitis (cf. Fusarium, causing recent incidences of keratitis through the possible vector of Bausch & Lomb ReNu with MoistureLoc contact lens solution)
- Viral keratitis
    - Herpes simplex keratitis. Viral infection of the cornea is often caused by the herpes simplex virus which frequently leaves what is called a 'dendritic ulcer'.
    - Herpes zoster keratitis
- Onchocercal keratitis, which follows O. volvulus infection by infected blackfly bite. These blackfly usually dwell near fast-flowing African streams, so the disease is also called "river blindness".

Other

- Exposure keratitis — due to dryness of the cornea caused by incomplete or inadequate eye-lid closure.
- Photokeratitis — keratitis due to intense ultraviolet radiation exposure (e.g. snow blindness or welder's arc eye.)
- Ulcerative keratitis
- Contact lens acute red eye (CLARE) — a non-ulcerative sterile keratitis associated with colonization of Gram-negative bacteria on contact lenses.
- Severe allergic response may lead to corneal inflammation and ulceration (i.e. vernal keratoconjunctivitis).
- Feline eosinophilic keratitis — affecting cats and horses; possibly initiated by feline herpesvirus 1 or other viral infection.

Diagnosis

Effective diagnosis is important in detecting this condition and subsequent treatment as keratitis is sometimes mistaken for an allergic conjunctivitis.

Treatment

Treatment depends on the cause of the keratitis. Infectious keratitis generally requires antibacterial, antifungal, or antiviral therapy to treat the infection. This treatment can involve prescription eye drops, pills, or even intravenous therapy. Over-the-counter eye drops are typically not helpful in treating infections. In addition, contact lens wearers are typically advised to discontinue contact lens wear and discard contaminated contact lenses and contact lens cases.

Antibacterial solutions include Quixin (levofloxacin), Zymar (gatifloxacin), Vigamox (moxifloxacin), Ocuflox (ofloxacin — available generically). Steroid containing medications should not be used for bacterial infections, as they may exacerbate the disease and lead to severe corneal ulceration and corneal perforation. These include Maxitrol (neomycin+polymyxin+dexamethasone — available generically), as well as other steroid medications.. One should consult an ophthalmologist or optometrist for treatment of an eye condition.

Some infections may scar the cornea to limit vision. Others may result in perforation of the cornea, (an infection inside the eye), or even loss of the eye. With proper medical attention, infections can usually be successfully treated without long-term visual loss.

Corneal ulcer

Corneal ulcer	
ICD-10	H16.0
ICD-9	370.00
MedlinePlus	001032 *001017*
eMedicine	oph/249

A **corneal ulcer**, or **ulcerative keratitis**, or **eyesore** is an inflammatory or more seriously, infective condition of the cornea involving disruption of its epithelial layer with involvement of the corneal stroma. It is a common condition in humans particularly in the tropics and the agrarian societies. In developing countries, Children afflicted by Vitamin A deficiency are at high risk for corneal ulcer and may become blind in both eyes, which may persist lifelong.

Corneal healing

An ulcer of the cornea heals by two methods: migration of surrounding epithelial cells followed by mitosis (dividing) of the cells, and introduction of blood vessels from the conjunctiva. Superficial small ulcers heal rapidly by the first method. However, larger or deeper ulcers often require the presence of blood vessels to supply inflammatory cells. White blood cells and fibroblasts produce granulation tissue and then scar tissue, effectively healing the cornea. The ulcer heals by the fourth day.

Superficial and deep corneal ulcers

Corneal ulcers are a common human eye disease. They are caused by trauma, particularly with vegetable matter, as also chemical injury, contact lenses and infections. Other eye conditions can cause corneal ulcers, such as entropion, distichiae, corneal dystrophy, and keratoconjunctivitis sicca (dry eye).

Many micro-organisms cause infective corneal ulcer. Among them are bacteria, fungi, viruses, protozoa, and chlamydia:

- Bacterial keratitis is caused by Staphylococcus aureus, Streptococcus viridans, Escherichia coli, Enterococci, Pseudomonas, Nocardia and many other bacteria.

- Fungal keratitis causes deep and severe corneal ulcer. It is caused by Aspergillus sp., Fusarium sp., Candida sp., as also Rhizopus, Mucor, and other fungi. The typical feature of fungal keratitis is slow onset and gradual progression, where signs are much more than the symptoms. Small satellite lesions around the ulcer are a common feature of fungal keratitis and hypopyon is usually seen.

- Viral keratitis causes corneal ulceration. It is caused most commonly by Herpes simplex, Herpes Zoster and Adenoviruses. Also it can be caused by coronaviruses & many other viruses. Herpes virus cause a dendritic ulcer, which can recur and relapse over the lifetime of an individual.

- Protozoa infection like Acanthamoeba keratitis is characterized by severe pain and is associated with contact lens users swimming in pools.

- Chlamydia trachomatis can also contribute to development of corneal ulcer.

Superficial ulcers involve a loss of part of the epithelium. Deep ulcers extend into or through the stroma and can result in severe scarring and corneal perforation. **Descemetoceles** occur when the ulcer extends through the stroma. This type of ulcer is especially dangerous and can rapidly result in corneal perforation, if not treated in time.

The location of the ulcer depends somewhat on the cause. Central ulcers are typically caused by trauma, dry eye, or exposure from facial nerve paralysis or exophthalmos. Entropion, severe dry eye and trichiasis (inturning of eyelashes) may cause ulceration of

the peripheral cornea. Immune-mediated eye disease can cause ulcers at the border of the cornea and sclera. These include Rheumatoid arthritis, rosacea, systemic sclerosis which lead to a special type of corneal ulcer called **Mooren's ulcer**. It has a circumferential crater like depression of the cornea, just inside the limbus, usually with an overhanging edge.

Symptoms

Corneal ulcers are extremely painful due to nerve exposure, and can cause tearing, squinting, and vision loss of the eye. There may also be signs of anterior uveitis, such as miosis (small pupil), aqueous flare (protein in the aqueous humour), and redness of the eye. An axon reflex may be responsible for uveitis formation — stimulation of pain receptors in the cornea results in release inflammatory mediators such as prostaglandins, histamine, and acetylcholine.

Diagnosis

Diagnosis is done by direct observation under magnified view of slit lamp revealing the ulcer on the cornea. The use of fluorescein stain, which is taken up by exposed corneal stroma and appears green, helps in defining the margins of the corneal ulcer, and can reveal additional details of the surrounding epithelium. Herpes simplex ulcers show a typical dendritic pattern of staining. Rose-Bengal dye is also used for supra-vital staining purposes, but it may be very irritating to the eyes. In descemetoceles, the Descemet's membrane will bulge forward and after staining will appear as a dark circle with a green boundary, because it does not absorb the stain. Doing a corneal scraping and examining under the microscope with stains like Gram's and KOH preparation may reveal the bacteria and fungi respectively. Microbiological culture tests may be necessary to isolate the causative organisms for some cases. Other tests that may be necessary include a Schirmer's test for keratoconjunctivitis sicca and an analysis of facial nerve function for facial nerve paralysis.

Treatment

Proper diagnosis is essential for optimal treatment. Bacterial corneal ulcer require intensive fortified antibiotic therapy to treat the infection. Fungal corneal ulcers require intensive application of topical anti-fungal agents. Viral corneal ulceration caused by herpes virus may respond to antivirals like topical acyclovir ointment instilled at least five times a day. Alongside, supportive therapy like pain medications are given, including topical cycloplegics like atropine or homatropine to dilate the pupil and thereby stop spasms of the ciliary muscle. Superficial ulcers may heal in less than a week. Deep ulcers and descemetoceles may require conjunctival grafts or conjunctival flaps, soft contact lenses, or corneal transplant. Proper nutrition, including protein intake and Vitamin C are usually advised. In cases of Keratomalacia, where the corneal ulceration is due to a deficiency of Vitamin A, supplementation of the Vitamin A by oral or intramuscular route is given. Drugs that are usually contraindicated in corneal ulcer are topical corticosteroids and anesthetics - these should not be used on any type of corneal ulcer

because they prevent healing, may lead to superinfection with fungi and other bacteria and will often make the condition much worse.

Refractory corneal ulcers

Refractory corneal ulcers are superficial ulcers that heal poorly and tend to recur. They are also known as **indolent ulcers** or **Boxer ulcers**. They are believed to be caused by a defect in the basement membrane and a lack of hemidesmosomal attachments. They are recognized by undermined epithelium that surrounds the ulcer and easily peels back. Refractory corneal ulcers are most commonly seen in diabetics and often occur in the other eye later. They are similar to Cogan's cystic dystrophy.

Treatment

Topical fortified antibiotics are used at hourly intervals to treat infectious corneal ulcers. Cycloplegic eye drops are applied to give rest to the eye.Pain medications are given as needed. Loose epithelium and ulcer base can be scraped off and sent for culture sensitivity studies to find out the pathogenic organism. This helps in choosing appropriate antibiotics. Complete healing takes anywhere from about a few weeks to several months.

Refractory corneal ulcers can take a long time to heal, sometimes months. In case of progressive or non-healing ulcers, surgical intervention by an ophthalmologist with corneal transplantation may be required to save the eye. In all corneal ulcers it is important to rule out predisposing factors like diabetes mellitus and immunodeficiency.

Melting ulcers

Melting ulcers are a type of corneal ulcer involving progressive loss of stroma in a dissolving fashion. This is most commonly seen in *Pseudomonas* infection, but it can be caused by other types of bacteria or fungi. These infectious agents produce proteases and collagenases which break down the corneal stroma. Complete loss of the stroma can occur within 24 hours. Treatment includes antibiotics and collagenase inhibitors such as acetylcysteine. Surgery in the form of corneal transplantation (penetrating keratoplasty) is usually necessary to save the eye.

Chapter 8

Photokeratitis and Thygeson's Superficial Punctate Keratopathy

Photokeratitis

Photokeratitis	
ICD-10	H16.1
ICD-9	370.24
DiseasesDB	31147
eMedicine	emerg/759

Photokeratitis or **ultraviolet keratitis** is a painful eye condition, caused by exposure of insufficiently protected eyes to the ultraviolet (UV) rays from either natural or artificial sources. Photokeratitis is akin to a sunburn of the cornea and conjunctiva, and is not usually noticed until several hours after exposure. Symptoms include increased tears and a feeling of gritty pain in the eyes.

The injury may be prevented by wearing eye protection that blocks most of the ultraviolet radiation, such as welding goggles with the proper filters, a welder's helmet, sunglasses rated for sufficient UV protection, or appropriate snow goggles. The condition is usually managed by removal from the source of ultraviolet, covering the corneas, and administration of pain relief.

Photokeratitis is known by a number of different terms including: **snow blindness**, **arc eye**, **welder's flash**, **bake eyes**, **corneal flash burns**, **flash burns**, **niphablepsia**, or **keratoconjunctivitis photoelectrica**.

Signs and symptoms

Common symptoms include pain (described as like having sand poured into the eyes), intense tears, eyelid twitching, discomfort from bright light, and constricted pupils.

Symptoms tend to occur a number of hours after exposure, and typically resolve spontaneously within 36 hours, although constriction of the pupils may last as long as 96 to 128 hours in some cases.

Cause

Any intense exposure to UV light can lead to photokeratitis. Common causes include welders who have failed to use adequate eye protection such as an appropriate welding helmet or welding goggles. This is termed **arc eye**, while photokeratitis caused by exposure to sunlight reflected from ice and snow, particularly at elevation, is commonly called **snow blindness**. It can also occur due to using tanning beds without proper eyewear. Natural sources include bright sunlight reflected from snow or ice or, less commonly, from sea or sand. Fresh snow reflects about 80% of the UV radiation compared to a dry, sandy beach (15%) or sea foam (25%). This is especially a problem in polar regions and at high altitudes, as with every thousand feet (approximately 305 meters) of elevation (above sea level), the intensity of UV rays increases by four percent.

Diagnosis

Fluorescein dye staining will reveal punctate areas of uptake under blue light.

Prevention

Photokeratitis can be prevented by using sunglasses or eye protection that transmits 5–10 % of visible light and absorbs almost all UV rays. Additionally, these glasses should have large lenses and side shields to avoid incidental light exposure. Sunglasses should always be worn, even when the sky is overcast, as UV rays can pass through clouds.

In the event of lost or damaged sunglasses, emergency goggles can be made by cutting slits in dark fabric or tape folded back onto itself. The SAS Survival Guide and Ancient Egyptians recommend blackening the skin underneath the eyes with charcoal to avoid any further reflection.

Treatment

Treatment consists of easing the pain temporarily with anesthetic eye drops. This may be done only initially, however, as continued anesthesia of the eye interferes with corneal healing, and may lead to corneal ulceration. Cool, wet compresses over the eyes and artificial tears may help local symptoms when the feeling returns. Nonsteroidal anti-inflammatory drug (NSAID) eyedrops are widely used to lessen inflammation and eye pain, but have not been proven in rigorous trials. Systemic (oral) pain medication is given if discomfort is severe. Healing is usually rapid (24–72 hours) if the injury source is removed. Further injury should be avoided by isolation in a dark room, removing contact lenses, not rubbing the eyes, and wearing sunglasses until the symptoms improve.

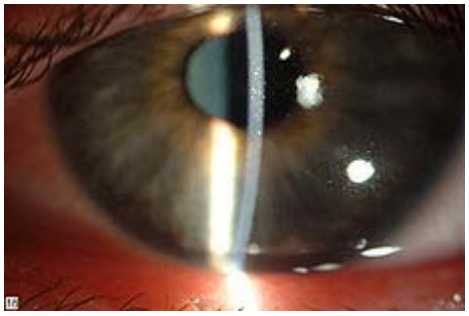
Inuit response

Traditional Inuit goggles used to combat snow blindness

The Inuit carved snow goggles from caribou antlers to help prevent snow blindness. The goggles were curved to fit the user's face, and had a large groove cut in the back to allow for the nose. A long thin slit was cut through the goggles to allow in a small amount of light, diminishing the amount of UV rays that get through. The goggles were held to the head by a cord made of caribou sinew.

Thygeson's superficial punctate keratopathy

Thygeson's superficial punctate keratopathy



Thygeson's keratitis. Note the slightly elevated, grayish-white subepithelial opacities.

ICD-10 H16.1

ICD-9	370.21
DiseasesDB	31288
eMedicine	article/1197335

Thygeson's superficial punctate keratopathy (**TSPK**; also *Thygeson Superficial Punctate Keratitis*) is a disease of the eyes. The causes of TSPK are not currently known, but details of the disease were first published in the Journal of the American Medical Association in 1950 by Phillips Thygeson - after whom it is named.

Symptoms

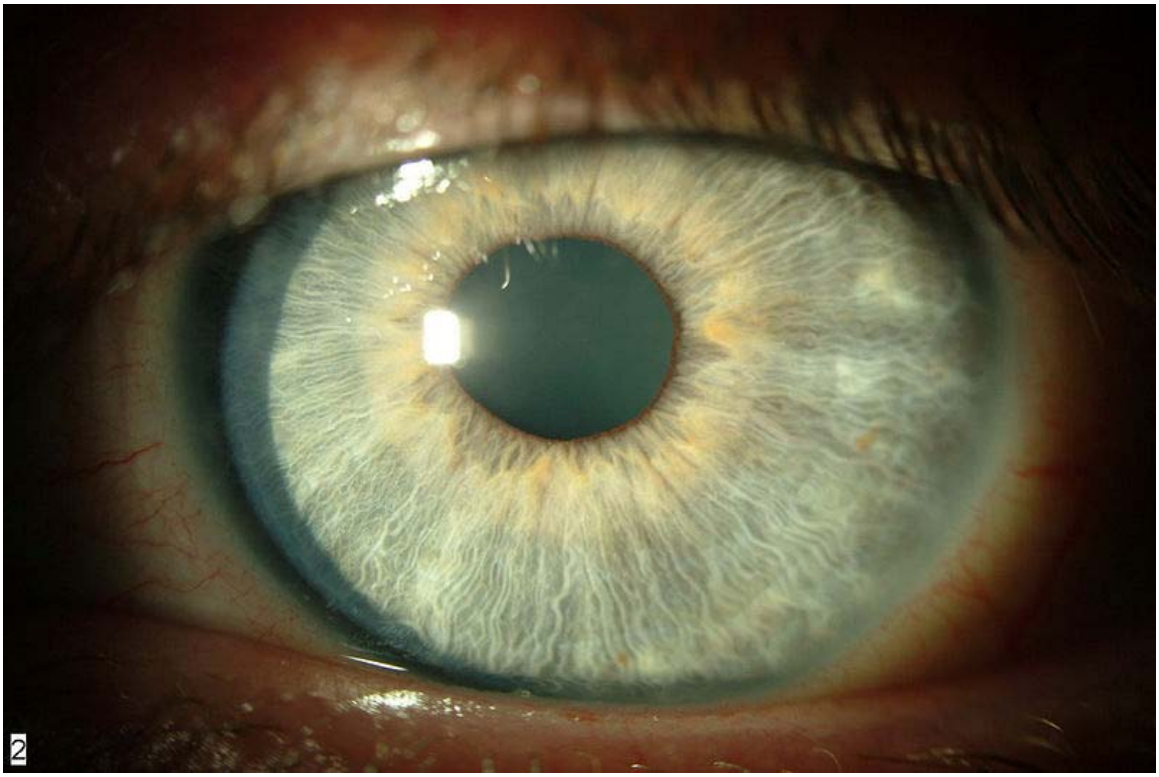
A patient with TSPK may complain of blurred vision, dry eyes, a sensation of having a foreign body stuck in the eye, photophobia (sensitivity to bright light), burning sensations and watery eyes. On inspection with a slit lamp, tiny lumps can be found on the cornea of the eye. These lumps can be more easily seen after applying fluorescein or rose bengal dye eye-drops. The lumps appear to be randomly positioned on the cornea and they may appear and disappear over a period of time (with or without treatment).

TSPK may affect one or both eyes. When both eyes are affected, the tiny lumps found on the cornea may differ in number between eyes. The severity of the symptoms often vary during the course of the disease. The disease may appear to go into remission, only to later reappear after months or years.

Causes

The causes of TSPK are not currently known.

Treatment



Same cornea as in the disease template on the upper right part of the page. Full resolution of opacities. From Hasanreisoglu and Avisar, 2008.

There are a number of different treatments to deal with TSPK. Symptoms may disappear without treatment, but treatment may help increase time to and success of remission.

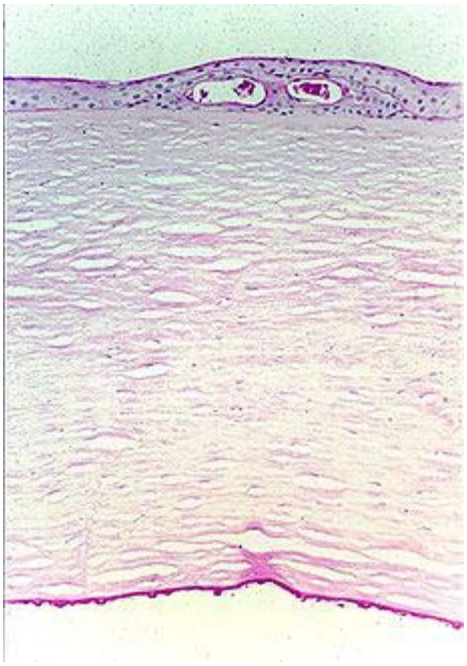
- PRK laser eye surgery may cure this disease (NOTE: A full clinical study has not been done, but a case study of one person was reported in 2002 PRK-pTK as a treatment).
- Artificial tear eye-drops or ointments may be a suitable treatment for mild cases.
- Low-dosage steroidal eye-drops, such as prednisone, fluorometholone, loteprednol (Lotemax 0.5%) or rimexolone. Steroidal drops should be used with caution and the eye pressure should be regularly checked during treatment.
- Soft contact lenses.
- Ciclosporin is an experimental treatment for TSPK. It is usually used during transplants as it reduces the immune system response.
- Laser eye treatment.

Chapter 9

Fuchs' Dystrophy and Iritis

Fuchs' dystrophy

Fuchs' dystrophy



Fuchs corneal dystrophy. Light microscopic appearance of the cornea showing numerous excrescences (guttae) on the posterior surface of Descemet's membrane and the presence of cysts in the corneal epithelium beneath ectopically placed intraepithelial basement membrane. Periodic acid-Schiff stain. From a review by Klintworth, 2009.

ICD-10	H18.5
ICD-9	371.57

OMIM	136800 610158
DiseasesDB	31163
eMedicine	article/1193591
MeSH	D005642

Fuchs' dystrophy, also known as **Fuchs' endothelial dystrophy**, is a slowly progressing corneal disease that usually affects both eyes and is slightly more common in women than in men. Although doctors can often see early signs of Fuchs' dystrophy in people in their 30s and 40s, the disease rarely affects vision until people reach their 50s and 60s.

The condition was first described by Austrian Ernst Fuchs (1851–1930), after whom it is named.

Etiology

Fuchs' endothelial dystrophy (FED) is a degenerative disorder of the corneal endothelium with accumulation of focal excrescences called guttae and thickening of Descemet's membrane, leading to corneal edema and loss of vision. Corneal endothelial cells are the major "pump" cells of the cornea to allow for stromal clarity. In FED, Descemet's membrane is grossly thickened with accumulation of abnormal wide-spaced collagen and numerous guttae. Corneal endothelial cells in end-stage FED are reduced in number and appear attenuated, causing progressive stromal edema. Progressive endothelial cell loss causes relative influx of aqueous humor into the cornea, leading to swelling (corneal stromal edema), which results in distorted vision. Eventually, the epithelium also becomes edematous, resulting in more severe visual impairment. Focal areas or blisters of epithelial edema ("bullae") may be particularly painful.

The inheritance of FED is autosomal dominant with genetic and environmental modifiers such as increased prevalence in the elderly and in females. Endothelial cell loss may be aggravated or accelerated by intraocular trauma or surgery. A common scenario involves excessive corneal swelling or edema following cataract surgery or other types of ocular surgery. Hence, patients with a history of Fuchs' dystrophy may be at a greater risk of corneal edema after ocular surgery as they have fewer functioning endothelial cells.

FED is classified into 4 stages, from early signs of guttae formation to end-stage subepithelial scarring. Diagnosis is made by biomicroscopic examination; other modalities, such as corneal pachymetry, confocal biomicroscopy, and specular microscopy can be used in conjunction.

Exact pathogenesis is unknown but factors include endothelial cell apoptosis, sex hormones, inflammation, and aqueous humor flow and composition. Mutations in collagen VIII, a major component of Descemet's membrane secreted by endothelial cells, have been linked to the early-onset FED.

Genes include:

Type	OMIM	Gene	Locus
FECD1	136800	*COL8A2*	1p34.3-p32.3
FECD4	610206	*SLC4A11*	20p13-p12
FECD6	189909	*ZEB1*	10p11.2

Signs and symptoms

At first, a person with Fuchs' dystrophy will awaken with blurred vision that will gradually clear during the day. This occurs because the cornea is normally thicker in the morning; it retains fluids during sleep that evaporate in the tear film while we are awake. As the disease worsens, this swelling will remain constant and reduce vision throughout the day.

Treatment

Medical management includes topical hypertonic saline, the use of a hairdryer to dehydrate the precorneal tear film, and therapeutic soft contact lenses. In using a hairdryer, the patient is instructed to hold a hairdryer at an arm's length or directed across the face, to dry out the epithelial blisters. This can be done two or three times a day. Definitive treatment, however, (especially with increased corneal edema) is surgical in the form of corneal transplantation, or penetrating keratoplasty (PKP).

Since 1998, new surgical modalities in the treatment of FED have been developed by Melles et al. in The Netherlands. These procedures, called posterior lamellar keratoplasty or endothelial keratoplasty, have been popularized as deep lamellar endothelial keratoplasty (DLEK) and Descemet's stripping with endothelial keratoplasty (DSEK). DLEK and DSEK avoid the surgical complications of PKP such as wound dehiscence and infections and high postoperative astigmatism. Since 2004, DSEK has become the dominant procedure because it is technically much easier for the surgeon compared to DLEK or PKP. Improved surgical instrumentation for DSEK, such as a DSEK graft injector will become available shortly (2008). This could allow faster recovery for patients because of the ability to perform DSEK through very small (3 mm) sutureless incisions.

Recently, endothelial keratoplasty has been further refined to Descemet Membrane Endothelial Keratoplasty (DMEK), in which only a donor Descemet membrane and its endothelium is transplanted. With DMEK, 90% of cases achieve a best spectacle corrected visual acuity 20/40 or better, and 60% of cases 20/25 or better within 1–3 months.

More speculative future directions in the treatment of FED include in vitro expansion of human corneal endothelial cells for transplantation, artificial corneas and genetic modification.

Iritis

Iritis	
ICD-10	H20.0
ICD-9	364.0

Iritis is a form of anterior uveitis and refers to the inflammation of the iris of the eye.

Types

A case of *Iritis* of the right eye

There are two main types of iritis: acute and chronic. They differ in numerous ways. Acute iritis is a type of iritis that can heal independently within a few weeks. If treatment is provided, acute iritis improves quickly. Chronic iritis can exist for months or years before recovery occurs. Chronic iritis does not respond to treatment as well as acute iritis does. Chronic iritis is also accompanied by a higher risk of serious visual impairment.

Signs and symptoms

- Ocular and periorbital pain
- Photophobia
- Consensual photophobia (pain in affected eye when light is shone in unaffected eye)
- Blurred or cloudy vision
- Reddened eye, especially adjacent to the iris

- White blood cells (leukocytes) (seen as tiny white dots, clinically termed cells) and protein (resulting in a grey or near-white haze, clinically termed flare) leak into the anterior chamber.
- Synechia (adhesion of iris to lens or cornea)
- Motion sickness

Causes and comorbidities

- Physical eye trauma

Inflammatory and Autoimmune Disorders:

- Ankylosing Spondylitis and other HLA-B27 related disorders
- Iridocyclitis, and other forms of uveal tract inflammation.
- Rheumatoid arthritis
- Behcet's disease
- Crohn's disease
- Graves disease
- Lupus
- Reactive arthritis
- Chronic psoriasis
- Psoriatic arthritis
- Sarcoidosis
- Scleroderma
- Ulcerative colitis
- Gout

Infections:

- Tuberculosis
- Lyme Disease
- Syphilis
- Toxoplasmosis
- Toxocaridae
- Herpes Simplex
- Herpes Zoster Virus

Cancers:

- Leukemia
- Lymphoma
- Malignant melanoma

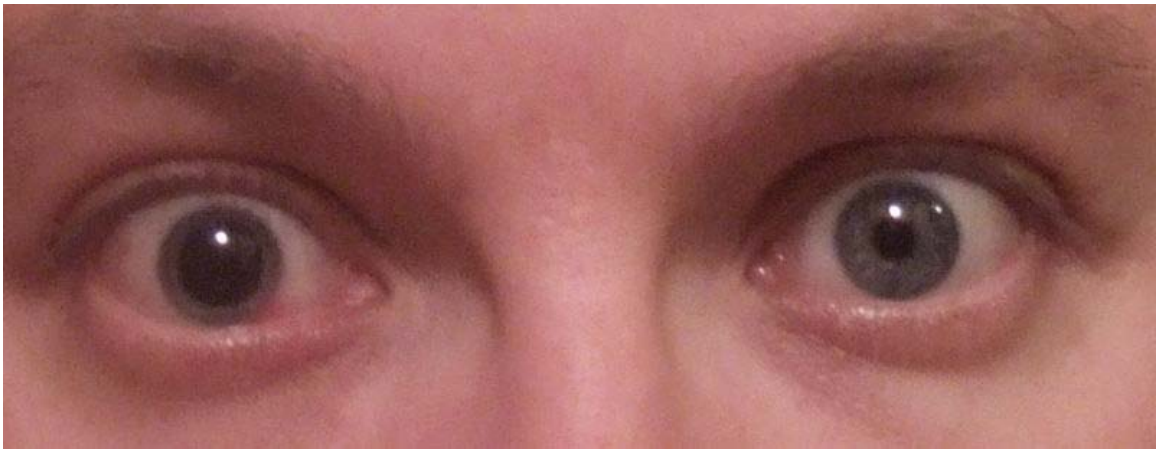
Iritis is usually secondary to some other systemic condition, but can be the only apparent somatic symptom.

Complications

Complications of iritis may include the following:

- Cataract
- glaucoma
- corneal calcification
- posterior uveitis
- blindness
- band keratopathy
- cystoid macular oedema.

Treatment



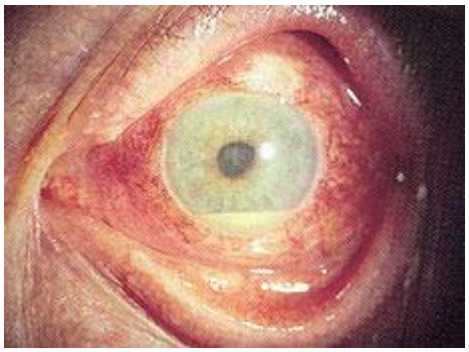
Eye treated with dilating eye drops (Atropine)

- Steroid anti-inflammatory eye drops (such as prednisolone acetate)
- Dilating eye drops (to help prevent synechia and reduce photophobia)
- Pressure-reducing eye drops (such as brimonidine tartrate)
- Oral steroids (such as prednisone)
- Subconjunctival steroid injections
- Steroid-sparing agents such as methotrexate (for prolonged, chronic iritis)

Chapter 10

Uveitis

Uveitis



Hypopyon in anterior uveitis, seen as yellowish exudate in lower part of anterior chamber of eye

ICD-10	H20.
ICD-9	364
DiseasesDB	13676
eMedicine	oph/580 emerg/284
MeSH	D014605

Uveitis specifically refers to inflammation of the middle layer of the eye, termed the "uvea" but in common usage may refer to any inflammatory process involving the interior of the eye.

Uveitis is estimated to be responsible for approximately 10% of the blindness in the United States. Uveitis requires an urgent referral and thorough examination by an optometrist or ophthalmologist along with urgent treatment to control the inflammation.

Anatomical classification

Uveitis may be classified anatomically into *anterior*, *intermediate*, *posterior* and *panuveitic* forms, based on which part of the eye is primarily affected by the inflammation.

- "Anterior uveitis" (or iridocyclitis) is the inflammation of the iris and anterior chamber. Anywhere from two-thirds to 90% of uveitis cases are anterior in location. This condition can occur as a single episode and subside with proper treatment or may take on a recurrent or chronic nature. Symptoms include red eye, injected conjunctiva, pain and decreased vision. Signs include dilated ciliary vessels, presence of cells and flare in the anterior chamber, and keratic precipitates ("KP") on the posterior surface of the cornea.
- "Intermediate uveitis" (pars planitis) consists of vitritis - inflammatory cells in the vitreous cavity, sometimes with *snowbanking*, or deposition of inflammatory material on the pars plana.
- "Posterior uveitis" (or chorioretinitis) is the inflammation of the retina and choroid.
- "Pan-uveitis" is the inflammation of all the layers of the uvea.

In 2004, a group of international uveitis specialists convened in Baltimore, MD, to standardize the method of reporting data in uveitis clinical trials, including anatomical classification. The results of this meeting were published in the American Journal of Ophthalmology in 2005.

Conditions associated with uveitis and uveitis syndromes

Myriad conditions can be associated with uveitis, including diseases with major extra-ocular involvement, as well as syndromes confined to the eye. In anterior uveitis, no associated condition or syndrome is found in approximately one-half of cases. However, anterior uveitis is often one of the syndromes associated with HLA-B27. Presence this type of HLA allele has a relative risk of evolving this disease by approximately 15%.

Systemic disorders associated with uveitis

Systemic disorders that can be associated with uveitis include:

- Ankylosing spondylitis
- Behçet's disease
- Chronic granulomatous disease
- Enthesitis
- Inflammatory bowel disease
- Juvenile rheumatoid arthritis
- Kawasaki's disease
- Multiple sclerosis
- Polyarteritis nodosa

- Psoriatic arthritis
- Reactive arthritis
- Sarcoidosis
- Systemic lupus erythematosus
- Vogt-Koyanagi-Harada syndrome
- Whipple's disease
- Lyme disease
- A wide range of autoimmune and autoinflammatory disease

Infectious causes

Uveitis may be a (normal) immune response to fight an infection inside the eye. While representing the minority of patients with uveitis, such possible infections include:

- Brucellosis
- Herpes simplex
- Herpes zoster
- Leptospirosis
- Lyme disease
- Presumed ocular histoplasmosis syndrome
- Syphilis
- Toxocariasis
- Toxoplasmosis
- Tuberculosis

Uveitis Syndromes

In many cases, uveitis is not associated with a systemic (i.e. extraocular) condition: the inflammation is confined to the eye. In some of these cases, the presentation in the eye is characteristic of a described syndrome, and include the following diagnoses:

- Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)
- Birdshot retinochoroidopathy
- Fuchs Heterochromic Iridocyclitis
- Multifocal Choroiditis and Panuveitis Syndrome
- Multiple Evanescent White Dot Syndrome (MEWDS)
- Punctate Inner Choroidopathy (PIC)
- Serpiginous Choroiditis

Masquerade syndromes

Masquerade syndromes are ophthalmic disorders that clinically present as either an anterior or posterior uveitis, but are not primarily inflammatory. The following are some of the most common:

- Anterior segment

    - Intraocular foreign body
    - Juvenile xanthogranuloma
    - Leukemia
    - Malignant melanoma
    - Retinal detachment
    - Retinoblastoma

- Posterior segment

    - Lymphoma
    - Malignant melanoma
    - Multiple sclerosis
    - Reticulum cell sarcoma
    - Retinitis pigmentosa
    - Retinoblastoma

Symptoms

- Redness of the eye
- Blurred vision
- Sensitivity to light (photophobia)
- Dark, floating spots along the visual field
- Eye pain

Treatment

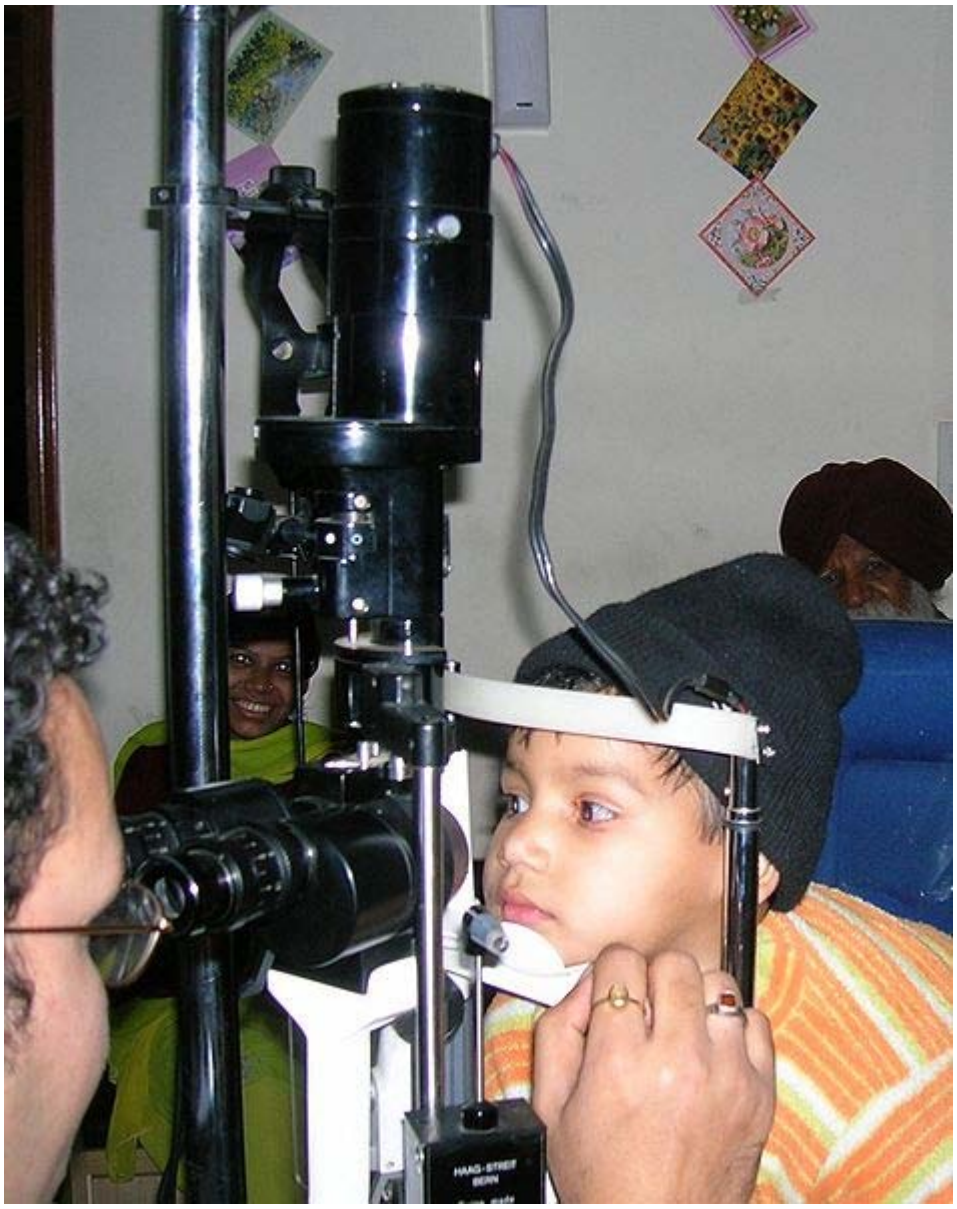
The prognosis is generally good for those who receive prompt diagnosis and treatment, but serious complication (including cataracts, glaucoma, band keratopathy, retinal edema and permanent vision loss) may result if left untreated. The type of uveitis, as well as its severity, duration, and responsiveness to treatment or any associated illnesses, all factor in to the outlook.

Uveitis is typically treated with glucocorticoid steroids, either as topical eye drops (prednisolone acetate) or oral therapy with corticosteroids. But before administration of corticosteroids, corneal ulcers are ruled out, typically by a Florescence Dye test. In addition to corticosteroids, topical cycloplegics, such as atropine or homatropine, may be used. In some cases an injection of PSTTA (posterior subtenon triamcinolone acetate) may also be given to reduce the swelling of the eye.

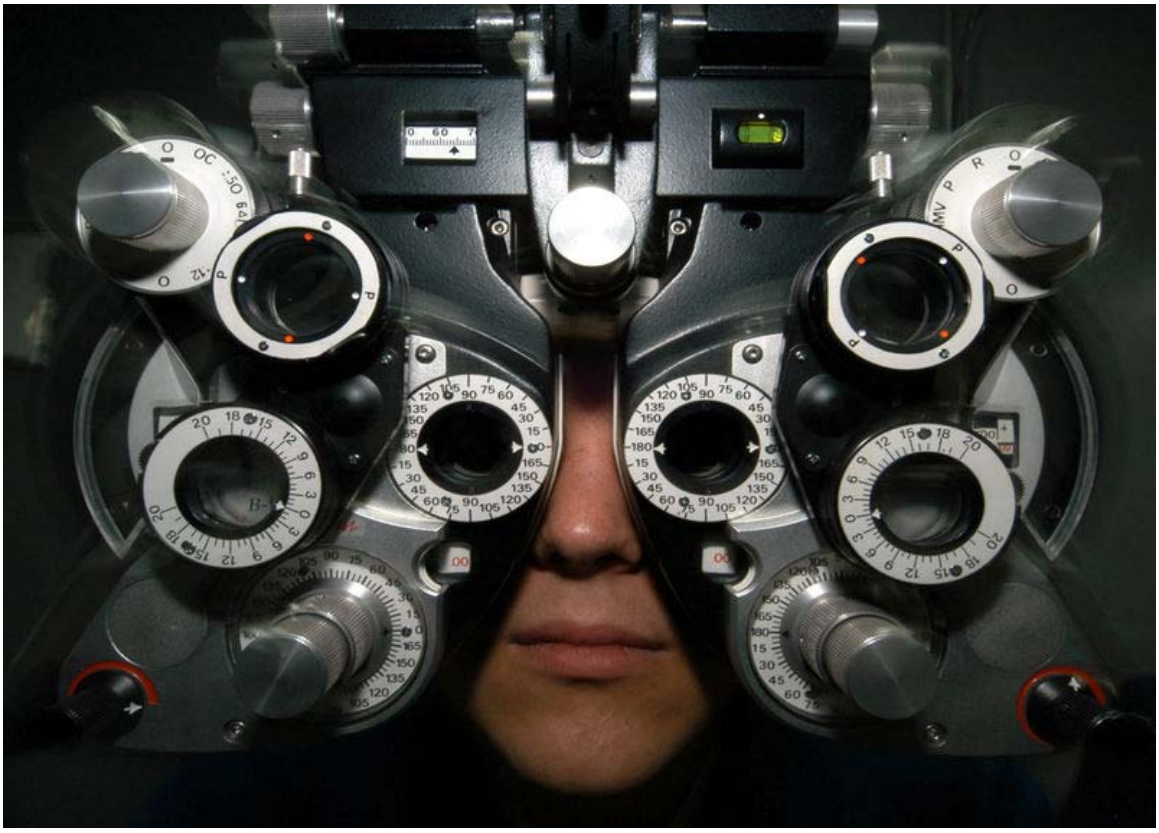
Antimetabolite medications, such as methotrexate are often used for recalcitrant or more aggressive cases of uveitis. Experimental treatments with Infliximab or other anti-TNFs' infusions may prove helpful.

Chapter 11

Ophthalmology



Slit lamp examination of eyes in an Ophthalmology Clinic



A phoropter in use

Ophthalmology is the branch of medicine which deals with the anatomy, physiology and diseases of the eye. The term **ophthalmologist** refers to a specialist in medical and surgical eye problems. Since ophthalmologists perform operations on eyes, they are considered to be both surgical and medical specialists.

The word *ophthalmology* comes from the Greek roots *ophthalmos* meaning *eye* and *logos* meaning *word*, *thought,* or *discourse*; ophthalmology literally means "the science of eyes". "Opthomology" is a common mis-hearing or mis-remembering of the term. As a discipline, it applies to animal eyes also, since the differences from human practice are surprisingly minor and are related mainly to differences in anatomy or prevalence, not differences in disease processes. However, veterinary medicine is regulated separately in many countries and states/provinces resulting in few ophthalmologists treating both humans and animals.

Early Developments

Sushruta

Sushruta wrote *Sushruta Samhita* in Sanskrit in about 800 BC He described 76 ocular diseases (of these 51 surgical) as well as several ophthalmological surgical instruments and techniques. His description of cataract surgery was more akin to extracapsular lens

extraction than to couching. The Indian surgeon Sushruta has been described as the first cataract surgeon.

Pre-Hippocrates

The pre-Hippocratics largely based their anatomical conceptions of the eye on speculation, rather than empiricism. They recognized the sclera and transparent cornea running flushly as the outer coating of the eye, with an inner layer with pupil, and a fluid at the centre. It was believed, by Alcamaeon and others, that this fluid was the medium of vision and flowed from the eye to the brain via a tube. Aristotle advanced such ideas with empiricism. He dissected the eyes of animals, and discovering three layers (not two), found that the fluid was of a constant consistency with the lens forming (or congealing) after death, and the surrounding layers were seen to be juxtaposed. He, and his contemporaries, further put forth the existence of three tubes leading from the eye, not one. One tube from each eye met within the skull.

Rufus

Rufus recognised a more modern eye, with conjunctiva, extending as a fourth epithelial layer over the eye. Rufus was the first to recognise a two chambered eye; with one chamber from cornea to lens (filled with water), the other from lens to retina (filled with an egg-white-like substance). Galen remedied some mistakes including the curvature of the cornea and lens, the nature of the optic nerve, and the existence of a posterior chamber. Though this model was roughly a correct but simplistic modern model of the eye, it contained errors. Yet it was not advanced upon again until after Vesalius. A ciliary body was then discovered and the sclera, retina, choroid and cornea were seen to meet at the same point. The two chambers were seen to hold the same fluid as well as the lens being attached to the choroid. Galen continued the notion of a central canal, though he dissected the optic nerve, and saw it was solid, He mistakenly counted seven optical muscles, one too many. He also knew of the tear ducts.

Middle Eastern ophthalmology

Medieval Islamic physicians are considered founders of ophthalmology as an independent discipline. One of the pioneers of ophthalmology was the Persian physician Rhazes. Innovations from this period include “injection syringe”, invented by the Iraqi physician Ammar ibn Ali of Mosul, which was used for the extraction by suction of soft cataracts. In cataract surgery, Ammar ibn Ali attempted the earliest extraction of cataracts using suction. He introduced a hollow metallic syringe hypodermic needle through the sclera and successfully extracted the cataracts through suction.

Ibn al-Haytham (Alhazen) wrote extensively on optics and the anatomy of the eye in his *Book of Optics* (1021). He was the first to hint at the retina being involved in the process of image formation.

Ibn al-Nafis, in *The Polished Book on Experimental Ophthalmology*, discovered that the muscle behind the eyeball does not support the ophthalmic nerve, and that the optic nerves transect but do not get in touch with each other. He also discovered new treatments for glaucoma and the weakness of vision in one eye when the other eye is affected by disease. Salah–ud-din bin Youssef al-Kalal bi Hama (i.e. the eye doctor of Hama) was a Syrian oculist who flourished in Hama in 1296. He wrote an elaborate treatise of ophthalmology entitled Nur al-Uyun wa Jami al-Funun (light of the eyes and collection of rules).

Seventeenth and eighteenth century

The seventeenth and eighteenth century saw the use of hand lenses (by Malpighi), microscopes (van Leeuwenhoek), preparations for fixing the eye for study (Ruysch) and later the freezing of the eye (Petit). This allowed for detailed study of the eye and an advanced model. Some mistakes persisted such as: why the pupil changed size (seen to be vessels of the iris filling with blood), the existence of the posterior chamber, and of course the nature of the retina. In 1722 Leeuwenhoek noted the existence of rods and cones though they were not properly discovered until Gottfried Reinhold Treviranus in 1834 by use of a microscope.

Ophthalmic surgery in Great Britain

The first ophthalmic surgeon in Great Britain was John Freke, appointed to the position by the Governors of St Bartholomew's Hospital in 1727, but the establishment of the first dedicated ophthalmic hospital in 1805; now called Moorfields Eye Hospital in London, England was a transforming event in modern ophthalmology. Clinical developments at Moorfields and the founding of the Institute of Ophthalmology (now part of the University College London) by Sir Stewart Duke Elder established the site as the largest eye hospital in the world and a nexus for ophthalmic research.

Professional requirements

Ophthalmologists are medical doctors (MD/MBBS or D.O., not OD or BOptom) who have completed a college degree, medical school, and residency in ophthalmology. In many countries, ophthalmologists also undergo additional specialized training in one of the many subspecialities. Ophthalmology was the first branch of medicine to offer board certification, now a standard practice among all specialties.

Australia and New Zealand

In Australia and New Zealand, the FRACO/FRANZCO is the equivalent postgraduate specialist qualification. It is a very competitive speciality to enter training and has a closely monitored and structured training system in place over the five years of postgraduate training. Overseas-trained Ophthalmologists are assessed using the pathway published on the RANZCO website. Those who have completed their formal training in the UK and have the CCST/CCT are usually deemed to be comparable.

Canada

In Canada, an Ophthalmology residency after medical school is undertaken. The residency lasts a minimum of five years after the MD degree although subspecialty training is undertaken by about 30% of fellows (FRCSC). There are about 30 vacancies per year for ophthalmology training in all of Canada.

Finland

In Finland, physicians willing to become Ophthalmologists must undergo a five year specialization which includes practical training and theoretical studies.

Germany

In Germany, physicians willing to become Ophthalmologists must undergo a five year specialization of practical training.

India

In India, after completing MBBS degree, post-graduation in Ophthalmology is required. The degrees are Doctor of Medicine (MD), Master of Surgery (MS), Diploma in Ophthalmic Medicine and Surgery (DOMS) or Diplomate of National Board (DNB). The concurrent training and work experience is in the form of a Junior Residency at a Medical College, Eye Hospital or Institution under the supervision of experienced faculty. Further work experience in form of fellowship, registrar or senior resident refines the skills of these eye surgeons. All India Ophthalmological Society (AIOS) and various state level Ophthalmological Societies (like DOS) hold regular conferences and actively promote continuing medical education. Royal colleges of the united kingdom, mainly Royal college of surgeons of Edinburgh (RCSEd), Royal College of ophthalmologists (RCOphth) and Royal college of physicians and Surgeons of Glasgow (RCPSG) have conducted their fellowship and membership examinations since the mid-1990s and awarding fellowships and memberships to the successful candidates.

Pakistan

In Pakistan, after MBBS, a 4 year full time residency programme leads to FCPS in Ophthalmology. Moreover, a two and a half years residency programme leads to MCPS while 2 years training of DOMS is also being offered. M.S.(Ophthalmology) is also one of the specialty programmes. In addition to programmes for Doctors, various diplomas and degrees for Opticians are also being offered to produce competent Optic technicians in this field. These programmes are being offered notably by Punjab Institute of Preventive Ophthalmology (PIPO) Lahore,Pakistan. Sub-specialty Fellowships are also being offered in the field of Pediatric Ophthalmology and Vitreo-Retinal Ophthalmology.

Philippines

Ophthalmology is a considered a medical specialty that uses medicine and surgery to treat diseases of the eye. To become a general ophthalmologist, a candidate must have completed a Doctor of Medicine degree or its equivalent (e.g. MBBS), have passed the physician licensure exam, completed an internship in medicine, and completed residency at any Philippine Academy of Ophthalmology (PAO) accredited program. Attainment of board certification in ophthalmology from PBO is optional, but is preferred and required to gain privileges in most major health institutions. Graduates of residency programs can receive further training in subspecialties of ophthalmology such as neuro-ophthalmology, etc. by completing a fellowship program which varies in length depending on each program's requirements. The leading professional organization in the country is the Philippine Academy of Ophthalmology which also regulates ophthalmology residency programs and board certification through its accrediting agency, the Philippine Board of Ophthalmology.

United Kingdom and Republic of Ireland

In the United Kingdom, there are three colleges that grant postgraduate degrees in ophthalmology. The Royal College of Ophthalmologists grants MRCOphth and FRCOphth (postgraduate exams), the Royal College of Edinburgh grants MRCSEd, the Royal College of Glasgow grants FRCS. In Ireland the Royal College of Ireland grants FRCSI. Work experience as a specialist registrar and one of these degrees is required for specialisation in eye diseases. There are only 2.3 ophthalmologists per 100,000 population in the UK -fewer pro rata than in any other nation in the European Union.

United States

In the United States, four years of residency training after medical school are required, with the first year being an internship in surgery, internal medicine, pediatrics, or a general transition year. Optional fellowships in advanced topics may be pursued for several years after residency. Most currently practicing ophthalmologists train in medical residency programs accredited by the Accreditation Council for Graduate Medical Education (ACGME) and are board certified by the American Board of Ophthalmology. Some physicians that train in osteopathic medical schools may hold a Doctor of Osteopathy ("DO") degree rather than an MD. The same residency and certification requirements for ophthalmology training must be fulfilled by osteopathic physicians. Completing the requirements of continuing medical education is mandatory for continuing licensure and re-certification. Professional bodies like the AAO and ASCRS organize conferences and help members through continuing medical education programs to maintain certification, in addition to political advocacy and peer support.

Sub-specialities

Ophthalmology includes sub-specialities which deal either with certain diseases or diseases of certain parts of the eye. Some of them are:

- Anterior segment surgery
- Cataract — not usually considered a subspecialty *per se*, since most general ophthalmologists perform cataract surgery
- Cornea, ocular surface, and external disease
- Glaucoma
- Medical retina, deals with treatment of retinal problems through non-surgical means.
- Neuro-ophthalmology
- Ocular oncology
- Oculoplastics & Orbit surgery
- Ophthalmic pathology
- Pediatric ophthalmology/Strabismus (mis-alignment of the eyes)
- Refractive surgery
- Uveitis/Immunology
- Veterinary Formal specialty training programs in veterinary ophthalmology now exist in some countries.
- Vitreo-retinal surgery, deals with surgical management of retinal and posterior segment diseases and disorders. Medical retina and vitreo-retinal surgery sometimes together called posterior segment subspecialisation.

Notable ophthalmologists

18th-19th century

- Sir William Adams (UK) Founder of Exeter's West of England Eye Infirmary.
- Carl Ferdinand von Arlt (1812–1887), the elder (Austrian) proved that myopia is largely due to an excessive axial length, published influential textbooks on eye disease, and ran annual eye clinics in needy areas long before the concept of volunteer eye camps became popular. His name is still attached to some disease signs, e.g., von Arlt's line in trachoma. His son Ferdinand Ritter von Arlt, the younger, was also an ophthalmologist.
- Jacques Daviel (France) claimed to be the 'father' of modern cataract surgery in that he performed extracapsular extraction instead of needling the cataract or pushing it back into the vitreous. It is said that he carried out the technique on 206 patients in 1752-3, out of which 182 were reported to be successful. These figures are not very credible, given the total lack of both anaesthesia and aseptic technique at that time.
- Frans Cornelis Donders (1818–1889) (Dutch) published pioneering analyses of ocular biomechanics, intraocular pressure, glaucoma, and physiological optics. Made possible the prescribing of combinations of spherical and cylindrical lenses to treat astigmatism.
- Albrecht von Graefe (1828–1870) (Germany) Along with Helmholtz and Donders, one of the 'founding fathers' of ophthalmology as a specialty. A brilliant clinician and charismatic teacher who had an international influence on the development of ophthalmology. A pioneer in mapping visual field defects and diagnosis and treatment of glaucoma. Introduced a cataract extraction technique

that remained the standard for over 100 years, and many other important surgical techniques such as iridectomy. Rationalised the use of many ophthalmically important drugs, including mydriatics & miotics. The founder of the one of the earliest ophthalmic societies (German Ophthalmological Society, 1857) and one of the earliest ophthalmic journals (Graefe's Archives of Ophthalmology). The most important ophthalmologist of the nineteenth century.

- Allvar Gullstrand (Sweden), Nobel Prize winner in 1911 for his research on the eye as a light-refracting apparatus. Described the *schematic eye* a mathematical model of the human eye based on his measurements known as the *optical constants* of the eye. His measurements are still used today.
- Hermann von Helmholtz, great German polymath, invented the ophthalmoscope (1851) and published important work on physiological optics, including colour vision (1850s).
- Hermann Snellen (Netherlands) introduced the Snellen chart to study visual acuity.
- Sir Arthur Conan Doyle (United Kingdom). English writer, primarily of the Sherlock Holmes stories. Trained in but apparently never practiced Ophthalmology.
- Jose Rizal (Philippines). The Philippines National Hero was an Ophtalmologist, One of his works was operation of his mother's eye for twice from cataract.

20th-21st century

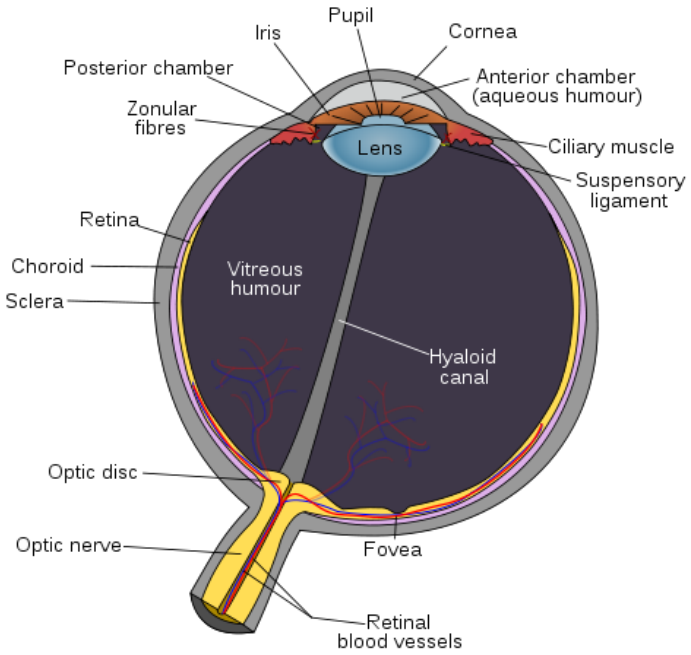
- William Horatio Bates (1860–1931) (United States) Creator of the unorthodox Bates Method, credited for being the founder of the Natural Vision Improvement movement.
- Vladimir Petrovich Filatov (1875–1956) (Ukraine) His contributions to the medical world include the tube flap grafting method, corneal transplantation and preservation of grafts from cadaver eyes and tissue therapy. He founded The Filatov Institute of Eye Diseases & Tissue Therapy, Odessa, one of the leading eye care institutes in the world.
- Ignacio Barraquer (1884–1965) (Spain) In 1917, invented the first motorized vacuum instrument (erisophake) for intracapsular cataract extraction. Founded of the Barraquer Clinic in 1941 and the Barraquer Institute in 1947 in Barcelona, Spain.
- Tsutomu Sato (Japan) Pioneer in incisional refractive surgery, including techniques for astigmatism and the invention of radial keratotomy for myopia.
- Jules Gonin (1870–1935) (Switzerland) "Father of retinal detachment surgery".
- Sir Harold Ridley (United Kingdom) In 1949, may have been the first to successfully implant an artificial intraocular lens after observing that plastic fragments in the eyes of wartime pilots were well tolerated. He fought for decades against strong reactionary opinions to have the concept accepted as feasible and useful.
- Charles Schepens (Belgium) "Father of modern retinal surgery". Developer of the Schepens indirect binocular ophthalmoscope whilst at Moorfields Eye Hospital. Founder of the Schepens Eye Research Institute in Boston, Massachusetts. This

premier research institute is associated with Harvard Medical School and Massachusetts Eye & Ear Infirmary.
- Marshall M. Parks "Father of pediatric ophthalmology".
- José Ignacio Barraquer (1916–1998) (Spain) "Father of modern refractive surgery". In the 1960s, developed lamellar techniques including keratomileusis and keratophakia, as well as the first microkeratome and corneal microlathe.
- Tadeusz Krwawicz (Poland) In 1961, developed the first cryoprobe for intracapsular cataract extraction.
- Svyatoslav Fyodorov (Russia) Popularizer of radial keratotomy.
- Charles Kelman (United States) Developed the ultrasound and mechanized irrigation and aspiration system for phacoemulsification, first allowing cataract extraction through a small incision.
- Ioannis Pallikaris (Greece) Performed the first laser-assisted intrastromal keratomileusis or LASIK surgery.
- Fred Hollows (New Zealand/Australia) Pioneered programs in Nepal, Eritrea, and Vietnam, and among Australian aborigines, including the establishment of cheap laboratory production of intraocular lenses in Nepal and Eritrea.
- Ian Constable (Australia) Founded the Lions Eye Institute in Perth, Western Australia, the largest eye research institute in the southern hemisphere and home to ten ophthalmologists.
- Rand Paul (United States) is a current member of The United States Senate from Kentucky. His father is U.S. Representative Ron Paul.
- L. L. Zamenhof (Poland) Creator of the Esperanto language.
- Bashar al-Assad (Syria) The President of Syria. He did his ophthalmology residency in a hospital in London.
- Syed Modasser Ali (Bangladesh) An ophthalmic surgeon who used to be the Director-General of Health Services for the government of Bangladesh. He wrote the first book on community ophthalmology (public eye health).
- David Taylor Pediatric ophthalmologist and author.
- Wallace Foulds (United Kingdom) Founder president of the Royal College of Opthamologists, founder of the Scottish Ophthalmic Oncology Service, and provided support for the founding of the Singapore Eye Research Institute.

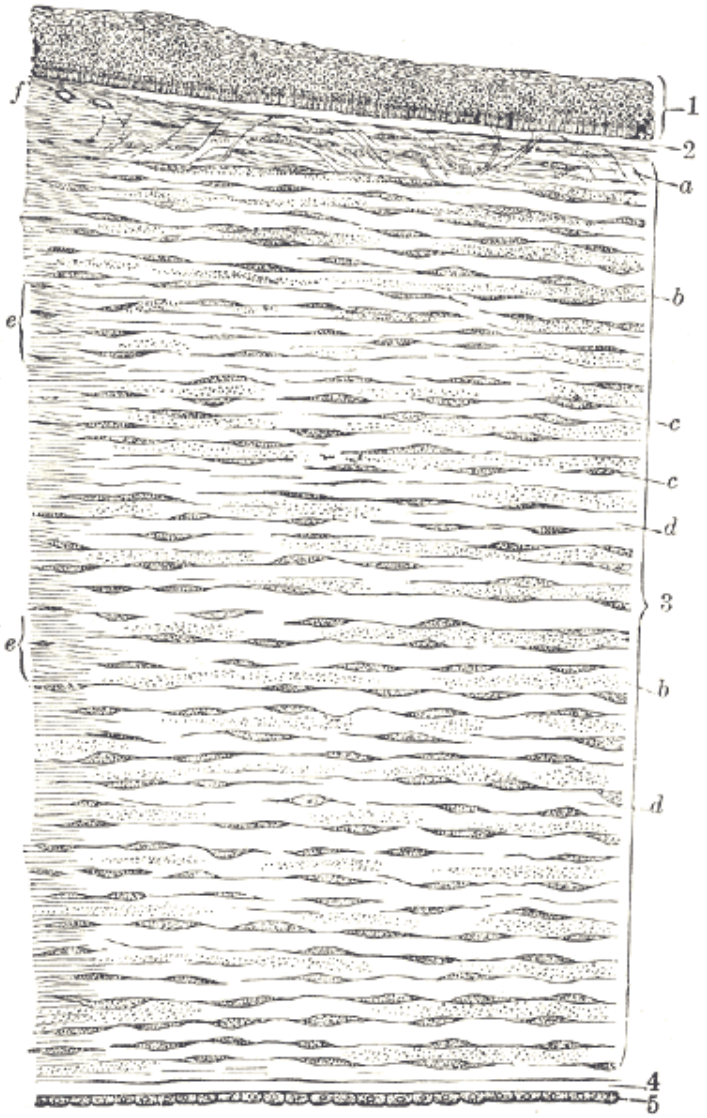
Chapter 12

Cornea

Cornea



Schematic diagram of the human eye. (Cornea labeled at center top.)



Vertical section of human cornea from near the margin.

(Waldeyer.) Magnified.

1. Epithelium.

2. Anterior elastic lamina.

3. substantia propria.

4. Posterior elastic lamina.

5. Endothelium of the anterior chamber.

a. Oblique fibers in the anterior layer of the substantia propria.

b. Lamellæ the fibers of which are cut across, producing a dotted appearance.

c. Corneal corpuscles appearing fusiform in section.

d. Lamellæ the fibers of which are cut longitudinally.

e. Transition to the sclera, with more distinct fibrillation, and surmounted by a thicker epithelium.

f. Small bloodvessels cut across near the margin of the cornea.

The **cornea** is the transparent front part of the eye that covers the iris, pupil, and anterior chamber. Together with the lens, the cornea refracts light, with the cornea accounting for approximately two-thirds of the eye's total optical power. In humans, the refractive power of the cornea is approximately 43 dioptres. While the cornea contributes most of the eye's focusing power, its focus is fixed. The curvature of the lens, on the other hand, can be adjusted to "tune" the focus depending upon the object's distance. Medical terms related to the cornea often start with the prefix "*kerat-*" from the Greek word κέρας, *horn*.

Structure

The cornea has unmyelinated nerve endings sensitive to touch, temperature and chemicals; a touch of the cornea causes an involuntary reflex to close the eyelid. Because transparency is of prime importance the cornea does not have blood vessels; it receives nutrients via diffusion from the tear fluid at the outside and the aqueous humour at the inside and also from neurotrophins supplied by nerve fibres that innervate it. In humans, the cornea has a diameter of about 11.5 mm and a thickness of 0.5–0.6 mm in the center and 0.6–0.8 mm at the periphery. Transparency, avascularity, the presence of immature resident immune cells, and immunologic privilege makes the cornea a very special tissue. The cornea has no blood supply; it gets oxygen directly through the air. Oxygen first dissolves in the tears and then diffuses throughout the cornea to keep it healthy.

It borders with the sclera by the corneal limbus.

The most abundant soluble protein in mammalian cornea is albumin.

In lampreys, the cornea is solely an extension of the sclera, and is separate from the skin lying above it, but in more advanced vertebrates it is always fused with the skin to form a single structure, albeit one composed of multiple layers. In fish, and aquatic vertebrates in general, the cornea plays no role in focusing light, since it has virtually the same refractive index as water.

Layers

The human cornea, like those of other primates, has five layers; the corneas of cats, dogs, wolves, and other carnivores only have four. From the anterior to posterior the five layers of the human cornea are:

1. **Corneal epithelium**: a thin epithelial multicellular tissue layer (non-keratinized stratified squamous epithelium) of fast-growing and easily-regenerated cells, kept moist with tears. Irregularity or edema of the corneal epithelium disrupts the smoothness of the air-tear film interface, the most significant component of the

total refractive power of the eye, thereby reducing visual acuity. It is continuous with the conjunctival epithelium is composed of about 6 layers of cells which are shed constantly on the exposed layer and are regenerated by multiplication in the basal layer.

2. **Bowman's layer** (also erroneously known as the *anterior limiting membrane*, when in fact it is not a membrane but a condensed layer of collagen): a tough layer that protects the corneal stroma, consisting of a similar irregularly-arranged collagen fibers, essentially a type of stroma. It is eight to 14 micrometres thick. This layer is absent in carnivores.
3. **Corneal stroma** (also *substantia propria*): a thick, transparent middle layer, consisting of regularly-arranged collagen fibers along with sparsely distributed interconnected keratocytes, which are the cells for general repair and maintenance. They are parallel and are superimposed like book pages The corneal stroma consists of approximately 200 layers of type I collagen fibrils. Each layer is 1.5 to 2.5 micrometres. Up to 90% of the corneal thickness is composed of stroma. There are 2 theories of how transparency in the cornea comes about:
    1. The lattice arrangements of the collagen fibrils in the stroma. The light scatter by individual fibrils is cancelled by destructive interference from the scattered light from other individual fibrils.(Maurice)
    2. The spacing of the neighbouring collagen fibrils in the stroma must be < 200 nm for there to be transparency. (Goldman and Benedek)
4. **Descemet's membrane** (also *posterior limiting membrane*): a thin acellular layer that serves as the modified basement membrane of the corneal endothelium, from which the cells are derived (but in a different collagen structure. It is 5-10 micrometres thick
5. **Corneal endothelium**: a simple squamous or low cuboidal monolayer of mitochondria-rich cells responsible for regulating fluid and solute transport between the aqueous and corneal stromal compartments. (The term *endothelium* is a misnomer here. The corneal endothelium is bathed by aqueous humour, not by blood or lymph, and has a very different origin, function, and appearance from vascular endothelia.) Unlike the corneal epithelium the cells of the endothelium do not regenerate. Instead, they stretch to compensate for dead cells which reduces the overall cell density of the endothelium and has an impact on fluid regulation. If the endothelium can no longer maintain a proper fluid balance, stromal swelling due to excess fluids and subsequent loss of transparency will occur.

The mnemonic "**EBSDE**in", read as "Ebstein" can be used to remember the layers in sequence.

Keeping the cornea transparent

Upon death or removal of an eye the cornea absorbs the aqueous humor, thickens, and becomes hazy. Transparency can be restored by putting it in a warm, well-ventilated chamber at 31 °C (88 °F, the normal temperature), allowing the fluid to leave the cornea and become transparent. The cornea takes in fluid from the aqueous humor and the small

blood vessels of the limbus, but a pump ejects the fluid immediately upon entry. When energy is deficient the pump may fail, or works too slowly to compensate, causing swelling. This could arise at death, but a dead eye can be placed in a warm chamber and the reservoirs of sugar and glycogen can keep the cornea transparent for at least 24 hours. The endothelium controls this pumping action, and as discussed above, damage thereof is more serious, and is a cause of opaqueness and swelling. When damage to the cornea occurs, such as in a viral infection, the collagen used to repair the process is not regularly arranged, leading to an opaque patch (leukoma). When a cornea is needed for transplant, as from an eye bank, the best procedure is to remove the cornea from the eyeball, preventing the cornea from absorbing the aqueous humor.

Innervation

The cornea is one of the most sensitive tissues of the body, as it is densely innervated with sensory nerve fibres via the ophthalmic division of the trigeminal nerve by way of 70–80 long ciliary nerves and short ciliary nerves. The ciliary nerves run under the endothelium and exit the eye through holes in the sclera apart from the optic nerve (which transmits only optic signals).

The nerves enter the cornea via three levels; *scleral, episcleral and conjunctival*. Most of the bundles give rise by subdivision to a network in the stroma, from which fibres supply the different regions. The three networks are *midstromal, subepithelial/Bowman's layer, and epithelium*. The receptive fields of each nerve ending are very large, and may overlap.

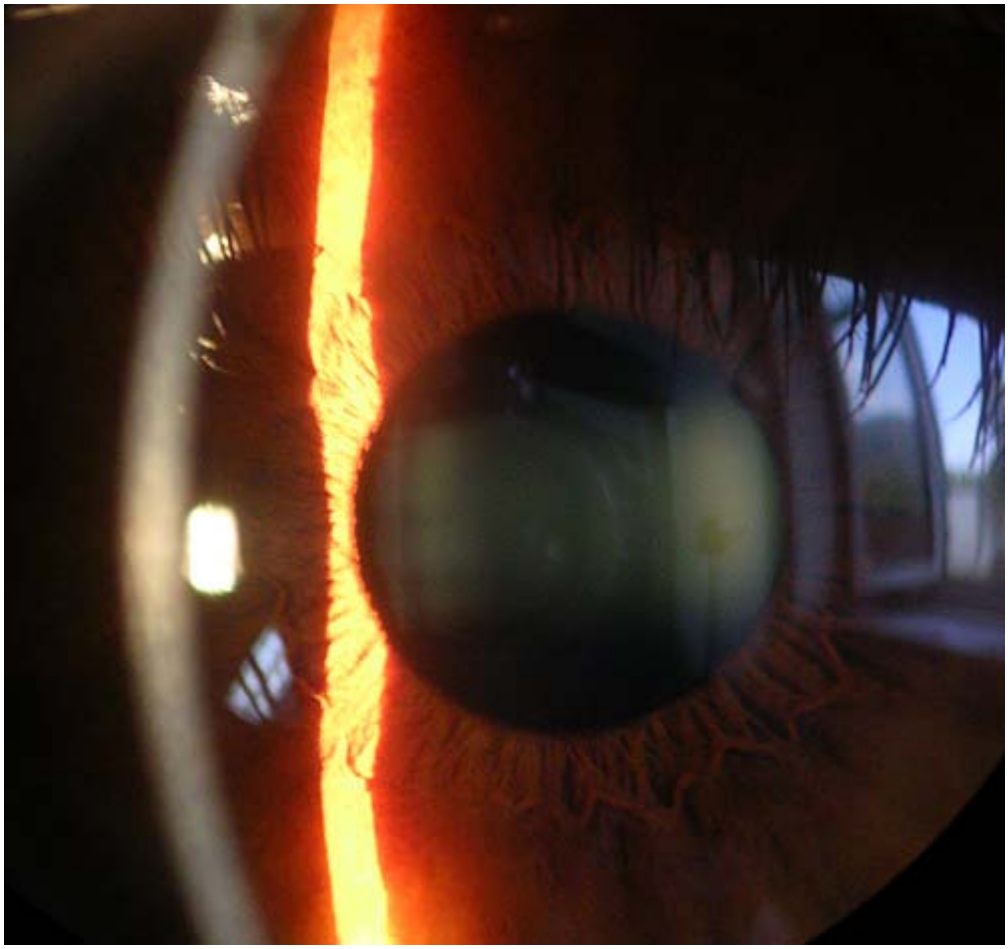
Corneal nerves of the subepithelial layer terminate near superficial epithelial layer of the cornea in a logarithmic spiral pattern.

Refractive nature

The optical component is concerned with producing a reduced inverted image on the retina. The eye's optical system consists of not only two but four surfaces—two on the cornea, two on the lens. Rays are refracted toward the midline. Distant rays, due to their parallel nature, converge to a point on the retina. The cornea admits light at the greatest angle. The aqueous and vitreous humors both have a refractive index of 1.336, whereas the cornea has a refractive index of 1.376. Because the change in refractive index between cornea and aqueous humor is relatively small compared to the change at the air–cornea interface, it has a negligible refractive effect, typically -6 diopters.

Diseases and disorders

Treatment and management



Slit lamp image of the cornea, iris and lens

Surgical procedures

Various refractive eye surgery techniques change the shape of the cornea in order to reduce the need for corrective lenses or otherwise improve the refractive state of the eye. In many of the techniques used today, reshaping of the cornea is performed by photoablation using the excimer laser.

If the corneal stroma develops visually significant opacity, irregularity, or edema, a cornea of a deceased donor can be transplanted. Because there are no blood vessels in the cornea, there are also few problems with rejection of the new cornea.

There are also synthetic corneas (keratoprostheses) in development. Most are merely plastic inserts, but there are also those composed of biocompatible synthetic materials

that encourage tissue ingrowth into the synthetic cornea, thereby promoting biointegration.

Non-surgical procedures

Orthokeratology is a method using specialized hard or rigid gas-permeable contact lenses to transiently reshape the cornea in order to improve the refractive state of the eye or reduce the need for eyeglasses and contact lenses.

In 2009, researchers at the University of Pittsburgh Medical center demonstrated that stem cells collected from human corneas can restore transparency without provoking a rejection response in mice with corneal damage.

Chapter 13

Pediatric Ophthalmology and Orthoptics

Pediatric ophthalmology

Pediatric ophthalmology is a sub-speciality of ophthalmology concerned with eye diseases, visual development, and vision care in children.

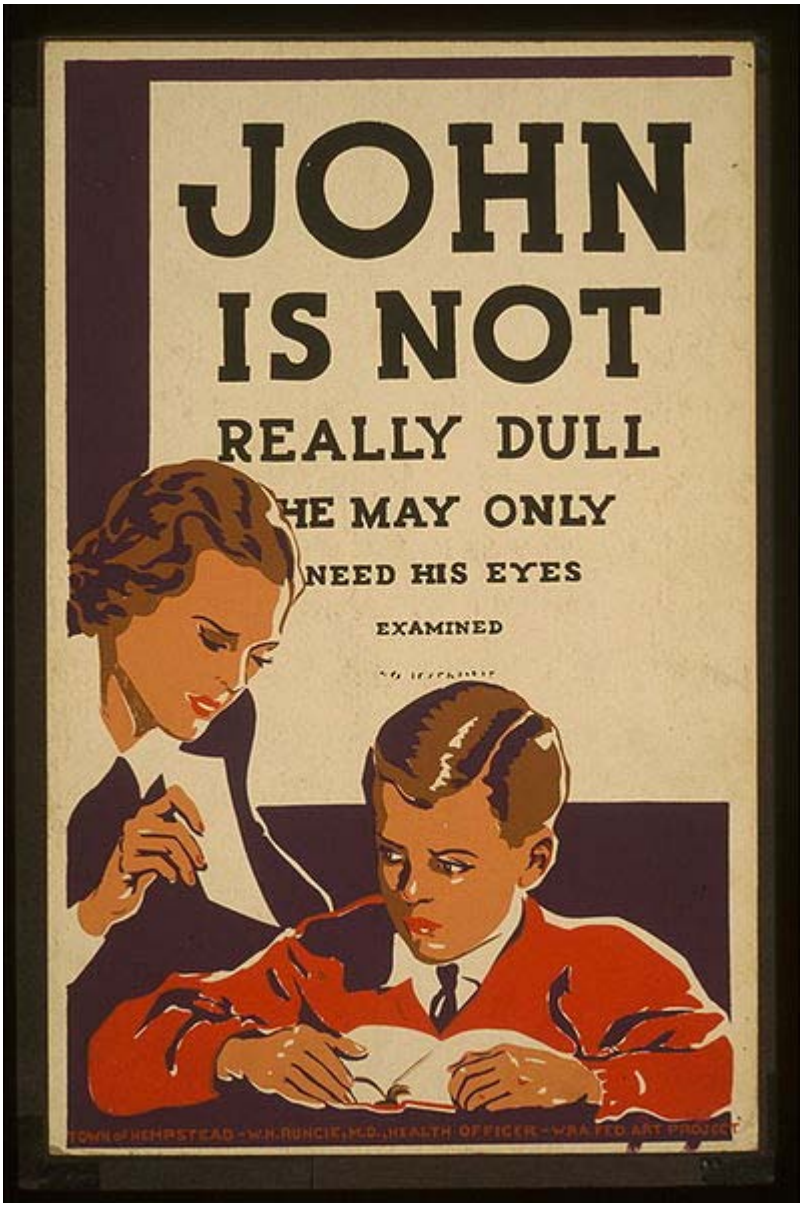
Training

In the United States, pediatric ophthalmologists are physicians who have completed medical school, a 1-year internship, 3-year residency in ophthalmology, and a 1-2 year fellowship in pediatric ophthalmology and strabismus. Pediatric ophthalmology fellowships in the United States are accredited by the American Association for Pediatric Ophthalmology and Strabismus.

Clinical expertise

Pediatric ophthalmologists focus on the development of the visual system and the various diseases that disrupt visual development in children. Pediatric ophthalmologists also have expertise in managing the various ocular diseases that affect children. Pediatric ophthalmologists are qualified to perform complex eye surgery as well as to manage children's eye problems using glasses and medications. Many ophthalmologists and other physicians refer pediatric patients to a pediatric ophthalmologist for examination and management of ocular problems due to children's unique needs. In addition to children with obvious vision problems, children with head turns, head tilts, squinting of the eyes, or preferred head postures (torticollis) are typically referred to a pediatric ophthalmologist for evaluation. Pediatric ophthalmologists typically also manage adults with eye movement disorders (strabismus) due to their familiarity with strabismus conditions.

Eye problems in children



Public education poster urging eye exams for children (Works Progress Administration, circa 1937)

Children experience a variety of eye problems, many quite distinct from adult eye diseases. Pediatric ophthalmologists are specially trained to manage the following disorders:

- Infections (conjunctivitis).
- Strabismus is a misalignment of the eyes that affects 2-4% of the population; it is often associated with amblyopia. The inward turning gaze commonly referred to as "crossed-eyes" is an example of strabismus. The term strabismus applies to

other types of misalignments, including an upward, downward, or outward turning eye.
- Amblyopia (aka lazy eye) occurs when the vision of one eye is significantly better than the other eye, and the brain begins to rely on the better eye and ignore the weaker one. Amblyopia affects 4% of the population and is clinically diagnosed when the refractive error of one eye is more than 1.5 diopters different than the other eye. The management of amblyopia involves correcting of significant refractive errors and using techniques that encourage the brain to pay attention to the weaker eye such as patching the stronger eye.
- Blocked tear ducts.
- Ptosis
- Retinopathy of prematurity
- Visual inattention
- Pediatric cataracts
- Pediatric glaucoma
- Abnormal vision development
- Genetic disorders often cause eye problems for affected children. Since approximately 30% of genetic syndromes affect the eyes, examination by a pediatric ophthalmologist can help with the diagnosis of genetic conditions. Many pediatric ophthalmologists participate with multi-disciplinary medical teams that treat children with genetic syndromes.
- Congenital malformations affecting vision or the tear drainage duct system can be evaluated and possibly surgically corrected by a pediatric ophthalmologist.
- Orbital tumours
- Refractive errors such as myopia (near-sightedness) and astigmatism can often be corrected with prescriptions for glasses or contacts.
- Accommodative insufficiency
- Convergence insufficiency and asthenopia
- Evaluation of visual issues in education, including dyslexia and attention deficit disorder.

Pediatric ophthalmologists often work in conjunction with orthoptists in the treatment of strabismus.

History

Frank D. Costenbader was an American physician frequently credited as the world's first pediatric ophthalmologist. Costenbader and Marshall M. Parks (his mentee who would later be known to many as "the father of pediatric ophthalmology") began the first ophthalmology fellowship trained program of any subspecialty at the Children's Hospital in Washington, D.C., now known as the Children's National Medical Center. Parks trained many pediatric ophthalmologists during his career and was instrumental in the establishment of the American Association for Pediatric Ophthalmology and Strabismus, a national organization dedicated to improving the quality and management of pediatric ocular disease. Over time, over 30 programs were developed for the training of pediatric ophthalmologists throughout the United States. The American Academy of Pediatric

Ophthalmology and Strabismus works with the American Academy of Pediatrics on issues related to pediatric eye disease and vision screening guidelines.

Orthoptics

Orthoptics (from the Greek words *ortho* meaning "straight", and *optikas* meaning "vision" is a discipline dealing with the diagnosis and treatment of defective eye movement and coordination (such as nystagmus), binocular vision, and amblyopia by eye care professionals. There are five areas of treatment for orthoptic problems:

- corrective lenses (spherical, cylindrical lens, prismatic and Fresnel lenses)
- strabismic-related orthoptics as an "eye exercise" is limited to the treatment of eye coordination problems by increasing the range of binocular fusion.
- eyepatching
- pharmaceuticals, such as cycloplegics
- surgery

However the term *orthoptics* is sometimes used to refer simply to eye exercises which are a component of strabismic-related vision therapy.

Orthoptists

Orthoptists are Eye care professionals who specialise in the diagnosis and management of binocular vision problems alongside Ophthalmologists. Orthoptists are represented worldwide by the International Orthoptic Association.

Orthoptics is usually studied as a primary or master's degree, or as a 2 to 4 years post graduate training course. Orthoptists usually work in close cooperation with Ophthalmologists, pediatricians, and sometimes neurologists. Continuing professional development and registration is required in most countries.

History

Orthoptists and ophthalmologists introduced a wide variety of techniques for the improvement of binocular function in the 1930s. The first pioneer was Mary Maddox, the daughter of an English ophthalmologist.

The orthoptic health care profession evolved and specialised as scientific development increased in the diagnosis, management and pre/post-surgical care of patients with strabismus, binocular vision abnormalities and specific pediatric disorders. Because of their lower prevalence and variational presentation, these were beyond the realm of a primary eyecare consultation at a spectacle shop (where most Optometrists work) and

beyond the Ophthalmologists' demanding surgical workload and practice. Hence, Orthoptists began to specialize in hospitals with these problems throughout more than 20 countries.

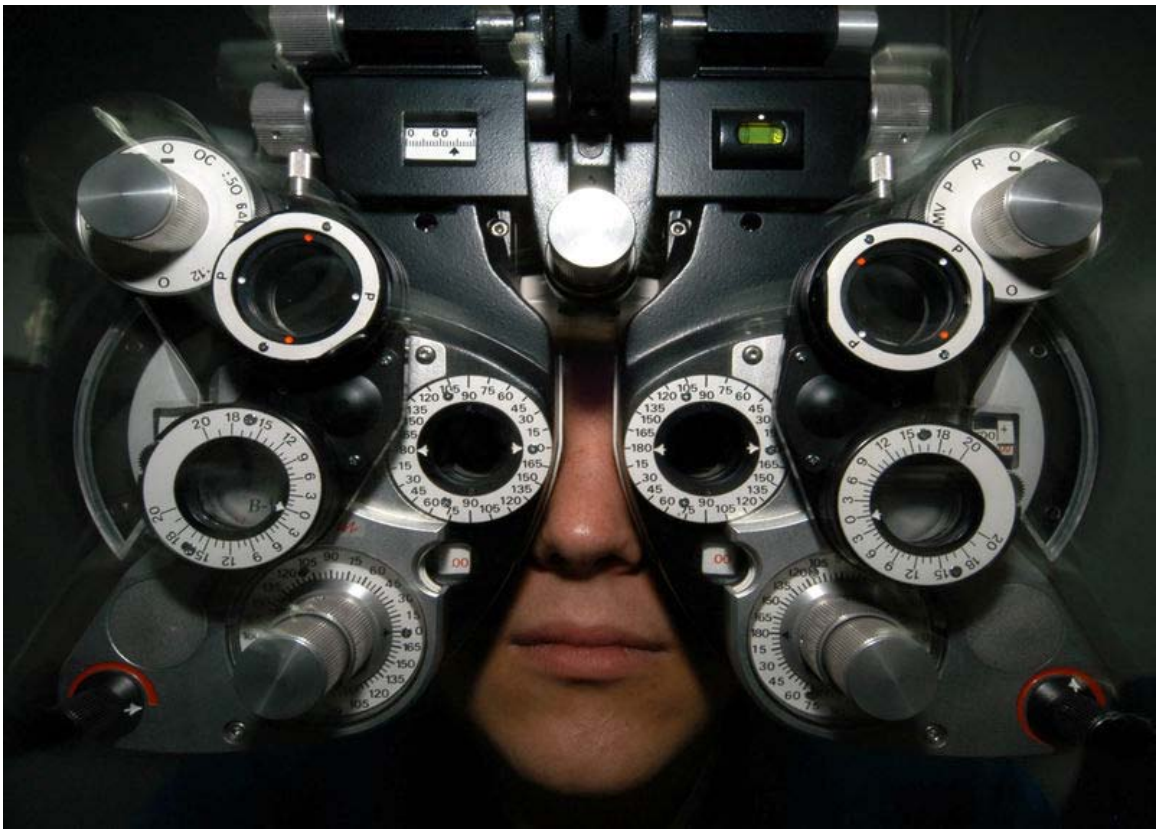
Current orthoptic practice

Orthoptists are mainly involved with diagnosing and managing patients with binocular vision disorders which relate to amblyopia, extraocular muscle balance such as with version, refractive errors, vergence, accommodation imbalances, (positive relative accommodation, negative relative accommodation) and pathological causes. They work closely with ophthalmologists to ensure that patients with eye muscle disorders are offered a full range of treatment options. According to the International Orthoptic Association, professional orthoptic practice involves the following:

- **Primary activities**
  - Ocular motility diagnosis & co-management
  - Vision screening
  - Assessment of special needs
  - Assessment and rehabilitation in neurological disorders
- **Secondary activities**
  - Low Vision assessment and management
  - Glaucoma assessment & stable glaucoma management
  - Biometry (includes sonography work)
  - Fundus photography & screening
  - Visual electrodiagnosis
  - Retinoscopy and refraction, such as using a phoropter to assess refractive errors
- **Further activities**
  - Specific outpatient waiting list initiatives to reduce the delay for children referred to the eye clinic (filter screening)
  - Joint multidisciplinary children’s vision screening clinics (orthoptics/optometry)
  - Organisation/prioritisation of the strabismus surgical admissions list according to agreed criteria
  - Assistance with surgical procedures

Chapter 14

Optometry



An optical refractor (phoropter) in use

Optometry is a health care profession concerned with eyes and related structures, as well as vision, visual systems, and vision information processing in humans. Optometrists are qualified to diagnose and treat eye diseases such as infections and glaucoma.

Like most professions, optometry education, certification, and practice is regulated in most countries. Optometrists and optometry-related organizations interact with governmental agencies, other health care professionals, and the community to deliver eye and vision care. Optometrists are one of three eye care professionals, the others being ophthalmologists (medical doctors), and opticians.

Background

The term "optometry" comes from the Greek words ὄψις (opsis), meaning *sight*, and μέτρον (metron), meaning *measurement.*

The eye, including its structure and mechanism, has fascinated scientists and the public in general since ancient times. Many of the expressions in the English language that mean to understand are equivalent vision terms. "I see," to mean I understand.

Many patients will be more concerned about diseases that affect vision than other, more lethal diseases when told that they may have an eye problem. Being deprived of sight can have a devastating effect on the psyche, as well as economic and social effects. Many blind individuals require significant assistance with activities of daily living and are often unable to continue gainful employment that might have previously been held while they could see. It is also well-known that serious diseases such as myasthenia gravis, diabetes, and atherosclerosis can show their first signs during an eye examination, well before a patient experiences any symptoms.

The maintenance of ocular health and correction of eye problems that decrease vision contribute greatly to the ability to appreciate the longer lifespan that all of medicine continues to allow. Given the importance of vision to quality of life, many optometrists consider their job to be rewarding, as they are often able to restore or improve a patient's sight.

Behavioral optometry is a related area of non-strabismus vision therapy that some optometrists practice. Generally ophthalmologists and orthoptists do not practice this. It generally involves intense therapy that requires at least a weekly visit with eye exercises at home. In some cases it can improve vision beyond that which eyeglasses alone can do.

In the United States, optometry is currently governed by state boards that determine their scope of practice. The scope of practice can vary dramatically from state to state. Optometrists have been successful in getting the right to use some types of medication, including pills, eye drops, and injections. In Oklahoma, optometrists are allowed by the state legislature to perform laser surgery.

History

Optometric history is tied to the development of

- vision science (related areas of medicine, microbiology, neurology, physiology, psychology, etc)
- optics, optical aids
- optical instruments, imaging techniques
- other eye care professions

The history of optometry can be traced back to the early studies on optics and image formation by the eye.

The origins of optometric science date back a few thousand years BC as evidence of the existence of lenses for decoration has been found. It is unknown when the first spectacles were made. According to research by David A. Goss, O.D., Ph.D., they originated in the late 13th century in Italy as stated in a manuscript from 1305 AD where a monk from Pisa named Rivalto stated “It is not yet 20 years since there was discovered the art of making eyeglasses”. Spectacles were manufactured in Italy, Germany, and the Netherlands by 1300 AD.

Benito Daza de Valdes published the third book on optometry in 1623, where he mentioned the use and fitting of eyeglasses. The term *optometrist* was coined by Edmund Landolt in 1886, referring to the "fitter of glasses". Prior to this, there was a distinction between "dispensing" and "refracting" opticians in the 19th century. The latter were later called optometrists.

In 1692, William Molyneux wrote a book on optics and lenses where he stated his ideas on myopia and problems related to close-up vision.

The scientists Claudius Ptolemy and Johannes Kepler also contributed to the creation of optometry. Kepler discovered how the retina in the eye creates vision.

From 1773 until around 1829, Thomas Young discovered the disability of astigmatism and it was George Biddell Airy who designed glasses to correct that problem that included spherocylindrical lens.

A pilgrim named Peter Brown is believed to be the first person to wear a pair of glasses in the US, however, eyeglasses were only made in Europe for a long period of time which made them both expensive and difficult to find. The first man to buy a pair of eyeglasses in the US was John McAllister Sr., from Philadelphia Pennsylvania, in 1783. McAllister, together with his son, John McAllister Jr. started making the first eyeglasses in the US in 1811. Their business continued until the 20th century. The family also taught refraction, and one of their students, James W. Queen also began his own business in 1853.

Benjamin Pike and James Prentice were two other early optometrists who studied in England and came to the US in 1847. They trained their sons, and James's son, Charles Prentice, had an important role in the development of optometry in the US.

The American Optometric Association was then formed on January 11, 1922 after Morris Steinfeld held a meeting with seven optometrists to discuss whether optometry should be a business or a profession. At the end of this meeting, they formed the American Academy of Optometry with the vision to transform the entire body optometric to a profession with a scientific base. The American Optometric Society was formed in August 2009. Doctors were concerned that policy decisions by the AOA leadership did

not represent the desires of the majority of the profession and were considered to not be in the best interest of the profession.

The first schools of optometry were established in 1850–1900 (in USA), and contact lenses were first used in 1940s

The first schools of optometry in the US began in the late 19th century, with the Illinois College of Optometry in 1872, and the New England College of Optometry in 1894. In 1914, a program in optometry began at The Ohio State University after Professor Charles Sheard gave a presentation to the Ohio State Optical Association who assisted him financially to open the program. It started as a two-year course that later became a four-year degree-granting program. Until 1937 the program was known as Applied Optics, when it then became known as Optometry.

Nowadays, there are many community and local resources to help those with financial difficulties to secure free or reduced cost eye care. Contact can be made to charities or non-profit organizations in the are to receive such help.

Licensing

Most countries have regulations concerning optometry education and practice. Optometrists like many other health care professionals are required to participate in ongoing continuing education courses to stay current on the latest standards of care.

Optometry is officially recognized:

- in North America (Canada and US)
- in Latin America and some Caribbean countries
- in most English speaking countries including UK, Republic of Ireland, Australia, New Zealand and South Africa
- in Europe including Spain, Germany and the English speaking European countries
- in Asia including China, Hong Kong, Malaysia, Philippines, Singapore, Taiwan and Thailand
- in the Middle East including Saudi Arabia, Iran and Israel

Argentina

In Argentina optometrists are required to register with the local Ministry of Public Information, but licensing is not required. Anyone holding a Bachelor's degree may register as an optometrist after completing a written exam. Fees for the exam are set by the provincial government and vary from province to province.

Australia

Australia currently has three recognised courses in Optometry. These are offered through the University of New South Wales: Bachelor of Optometry Bachelor of Science

(BOptom BSc), a 5 year course; Queensland Universtiy of Technology: Bachelor of Vision Science and Masters of Optometry, a 5 year course; and Melbourne University which is transitioning to a Doctor of Optometry course a 4 year postgraduate course. These courses are developments of prior course offerings at these institutions that have been expanded along with the increased scope of practice for Optometrists in Australia, specifically the ability to prescribe certain therapeautic agents.

New courses are being developed at Flinders University in South Australia, which accepted students in a science degree in 2010 and will begin the post graduate component of the course in 2013. A second new course is expected to be offered at Deakin University in Geelong, Vic at the beginning of 2012.

Canada

In Canada optometrists hold a Doctorate of Optometry degree and are licensed by the boards in the provinces they wish to practice. There are two schools of optometry, one at the University of Waterloo and the other at Universite de Montreal.

Colombia

In Colombia optometry education has been accredited by the Ministry of Health. The last official revision to the laws regarding health care standards in the country was issued in 1992 through the Law 30. Currently there are eight official universities that are entitled by ICFES to grant the Optometrist certification. The first optometrists arrived in the country from North America and Europe circa 1914. These professionals specialized in optics and refraction. In 1933, under Decrees 449 and 1291, the Colombian Government officially set the rules for the formation of professionals in the field of optometry. In 1966 La Salle University opened its first Faculty of Optometry after recommendation from a group of professionals. At the present time optometrists are encouraged to keep up with new technologies through congresses and scholarships granted by the government or the private sector (such as Bausch & Lomb).

Europe

Currently, optometry education and licensing varies throughout Europe. For example, in Germany, optometric tasks are performed by ophthalmologists and professionally trained and certified opticians. In France, there is no regulatory framework and optometrists are sometimes trained by completing an apprenticeship at an ophthalmologists' private office.

Since the formation of the European Union, "there exists a strong movement, headed by the Association of European Schools and Colleges of Optometry (AESCO), to unify the profession by creating a European-wide examination for optometry" and presumably also standardized practice and education guidelines within EU countries. The first examinations of the new European Diploma in Optometry were held in 1998 and this was a landmark event for optometry in continental Europe.

Ireland

The profession of Optometry has been represented for over a century by the Association of Optometrists, Ireland [AOI]. In Ireland an optometrist must first complete a four year degree in optometry at D.I.T. Kevin Street. Following successful completion of the a degree, an optometrist must then complete Professional Qualifying Examinations in order to be entered into the register of the Opticians Board [Bord na Radharcmhaistoiri]. Optometrists must be registered with the Board in order to practice in the Republic of Ireland.

The A.O.I. runs a comprehensive continuing education and professional development program on behalf of Irish optometrists. The legislation governing Optometry was drafted in 1956. Some feel that the legislation restricts optometrists from using their full range of skills, training and equipment for the benefit of the Irish public. The amendment to the Act in 2003 addressed one of the most significant restrictions - the use of cycloplegic drugs to examine children.

United Kingdom

In the United Kingdom, optometrists have to complete a 3 or 4 (Scotland) year undergraduate honours degree followed by a minimum of a one-year "pre-registration period" where they complete supervised practice under the supervision of an experienced qualified practitioner. During this year the pre-registration candidate is given a number of quarterly assessments and on successfully passing all of these assessments, a final one-day set of examinations (Examination details correct for candidates from 2006 onwards). Following successful completion of these assessments and having completed one year's supervised practice, the candidate is eligible to register as an optometrist with the General Optical Council (GOC) and, should they wish, are entitled to membership of The College of Optometrists. Registration with the GOC is mandatory to practice in the UK. Members of the College of Optometrists (incorporated by a Royal Charter) may use the suffix MCOptom. There are 9 universities which offer optometry in the UK.

Philippines

Optometry is regulated by the Professional Regulation Commission of the Philippines. To be eligible for licensing, each candidate must have satisfactorily completed a Doctor of Optometry course at an accredited institution and demonstrate good moral character with no previous record of professional misconduct. Professional organizations of optometry in the Philippines include Optometric Association of the Philippines and Integrated Philippine Association of Optometrists, Inc. (IPAO)

Russia

In Russia optometry education has been accredited by the Federal Agency of Health and Social Development. There are only two educational institutions that teach optometry in Russia: Saint Petersburg Medical Technical College, formerly known as St. Petersburg

College of Medical Electronics and Optics, and The Helmholz Research Institute for Eye Diseases. They both belong and are regulated by the Ministry of Health. The Optometry program is a 4 year program. It includes 1–2 science foundation years, 1 year focused on clinical and proficiency skills, and 1 year of clinical rotations in hospitals. Graduates take college/state examinations and then receive a specialist diploma. This diploma is valid for only 5 years and must be renewed every 5 years after receiving additional training at state accredited programs.

United States

The American Optometric Association (AOA) and the American Optometric Society (AOS) represent optometrists nationally in the USA. Prior to admittance into optometry school, optometrists typically complete four years of undergraduate study, culminating in a bachelor's degree. Required undergraduate coursework for pre-optometry students covers a variety of health, science and mathematics courses. These courses include: 4 semesters of chemistry to include organic and biochemistry, 2 semesters of physics and biology, as well as 1 semester of calculus, statistics, physiology, anatomy, microbiology, and psychology. Additional requirements are imposed by specific institutions. Once completing these courses in order to be admitted to an optometry doctorate program one must score well on the O.A.T., Optometry Admission Tests. There are currently 20 optometry schools in the United States, and admission into these schools are considered to be extremely competitive.

Optometrists are required to complete a four-year postgraduate degree program to earn their Doctor of Optometry (O.D.) titles. The four-year program includes classroom and clinical training in geometric, physical, physiological and ophthalmic optics, ocular anatomy, ocular disease, ocular pharmacology, neuroanatomy and neurophysiology of the vision system, binocular vision, color, form, space, movement and vision perception, design and modification of the visual environment, and vision performance and vision screening. In addition, an optometric education also includes a thorough study of human anatomy, general pharmacology, general pathology, sensory and perceptual psychology, biochemistry, statistics and epidemiology. There are three new colleges of optometry (Midwestern University Arizona College of Optometry, University of the Incarnate World School of Optometry, Western University of Health Sciences College of Optometry) that have received the pre-accreditation status of preliminary approval from the Accreditation Council on Optometric Education (ACOE). Programs with "Preliminary Approval" have shown that they are developing within the ACOE's standards. The programs have approval to begin recruiting and admitting students, and to begin offering the program.

Upon completion of an accredited program in optometry, graduates hold the Doctor of Optometry (O.D. - Oculis Doctor) degree. Optometrists must then pass a national examination administered by the National Board of Examiners in Optometry (NBEO). The three-part exam includes basic science, clinical science and patient care. (The structure and format of the NBEO exams are subject to change beginning in 2008.) Some optometrists go on to complete 1–2 year residencies with training in a specific sub-

specialty such as pediatric eyecare, geriatric eyecare, specialty contact lens, ocular disease or neuro-optometry. All optometrists are required to fulfill continuing education requirements to stay current regarding the latest standards of care.

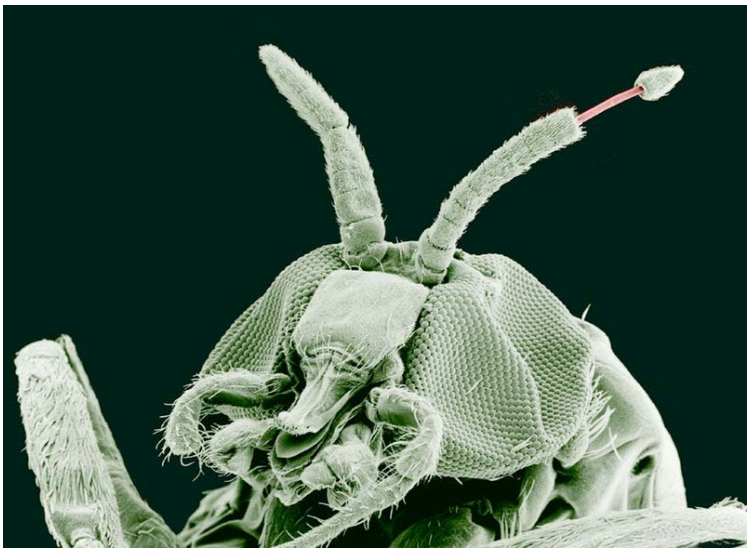
Within the healthcare system, optometrists function as primary eye care providers that are especially experienced in fitting contact lenses and glasses prescriptions. Although they are not technically physicians, in that they do not hold an MD or DO degree, some states, such as Florida, refer to optometrists as "Optometric Physicians" in a legal sense. Optometrists also have the ability to treat a wide variety of eye diseases though the administration of topical, oral and injectable medicines (depending on the state) although their scope is limited to the eye. Optometrists may be trained in some surgical techniques, including those for foreign body removal, corneal injury, eyelid & lacrimal disease, and others. In Oklahoma, the state optometry board also allows state-certified optometrists to perform laser surgeries limited to the anterior segment of the eye. Additionally, Department of Veterans Affairs (VA) facilities permit optometrists who have been trained in these laser techniques and hold licensure in Oklahoma to perform laser surgeries in any VA facility in the country, not just Oklahoma VA hospitals.

Ophthalmologists differ from optometrists in that ophthalmologists are physicians who are licensed to perform all eye surgeries, can treat systemic diseases, and who have completed 4 years of medical school and a residency in ophthalmology. In many cases optometrists and ophthalmologists work together in the treatment and management of patients with various eye conditions. Opticians generally dispense corrective eye wear, and in some cases will also construct the corrective eye wear. The scope of practice in optometry varies as it is regulated by each state.

Chapter 15

Onchocerciasis

Onchocerciasis



Adult Black Fly (*Simulium yahense*) with parasite (*Onchocerca volvulus*) emerging from the insect's antenna. Magnified 100x.

ICD-10	B73.
ICD-9	125.3
DiseasesDB	9218
eMedicine	med/1667 oph/709
MeSH	D009855

Onchocerciasis also known as **river blindness** and **Robles' Disease**, is a parasitic disease caused by infection by *Onchocerca volvulus*, a nematode (roundworm). Onchocerciasis is the world's second-leading infectious cause of blindness. It is not the nematode but its endosymbiont, *Wolbachia pipientis*, that causes the severe inflammatory response that leaves many blind. The parasite is transmitted to humans through the bite of a blackfly of the genus *Simulium*. The larval nematodes spread throughout the body.

When the worms die their Wolbachia symbionts are released, triggering a host immune system response that causes intense itching and can destroy nearby tissue, such as the eye.

The vast majority of infections occur in sub-Saharan Africa, although cases have also been reported in Yemen and isolated areas of Central and South America. An estimated 18 million people suffer from onchocerciasis, with approximately 270,000 cases of blindness related to the infection.

In 1915, Dr. Rodolfo Robles Valverde's study on patients with river blindness in Guatemala led to the discovery that the disease is caused by filaria volvulus and sheds light on the life cycle and transmission of the parasite. Using case studies of coffee plantation workers in Guatemala, Robles hypothesized that the vector of the disease is a day-biting insect, and more specifically two anthrophilic species of Simulium flies found in the endemic areas. He publishes his findings on a “new disease” from Guatemala associated with subcutaneous nodules, anterior ocular lesions, dermatitis, and microfilariae in 1917.

Treatment may involve the use of the drug ivermectin. For best effect, entire communities are treated at the same time. A single dose may kill first-stage larvae (microfilariae) in infected people and prevents transmission for many months in the remaining population. Other drugs are also available including the tetracycline-class antibiotic doxycycline, which kills the Wolbachia and renders the female nematodes sterile. The removal of the palpable nodules is popular in Guatemala, Ecuador, and Mexico.

Classification

Onchocerciasis may be divided into the following phases or types:

Erisipela de la costa
: An acute phase characterized by swelling of the face with erythema and itching. Onchocerciasis causes different kinds of skin changes and these changes vary in different geographic regions. This skin change, erisípela de la costa, of acute onchocerciasis is most commonly seen among victims in Central and South America.

Mal morando
: A cutaneous condition characterized by inflammation that is accompanied by hyperpigmentation.

Sowda
: A cutaneous condition, a localized type of onchocerciasis.

Additionally, the various skin changes associated with onchocerciasis may be described as follows:

Leopard skin
: A term referring to the spotted depigmentation of the skin that may occur with onchocerciasis.

Elephant skin
: A term used to describe the thickening of human skin that may be associated with onchocerciasis.

Lizard skin
: A term used to describe the thickened, wrinkled skin changes that may result with onchocerciasis.

Life cycle of onchocerca volvulus

The life of the O. Volvulus parasite can be traced through the black fly and the human hosts in the following steps:

1. A Simulium female black fly takes a blood meal on an infected human host ingesting microfilaria.
2. The microfilaria enter the gut and thoracic flight muscles of the black fly progressing into the first larval stage (J1.).
3. The larvae mature into the second larval stage (J2.) and moves to the proboscis and into the saliva in its third larval stage (J3.). Mature in about 7 days.
4. The black fly takes another blood meal passing the larvae into the next human host's blood.
5. The larvae migrate to the subcutaneous tissue and undergo two more molts. They form nodules as they mature into adult worms over six to twelve months.
6. After maturing, adult male worms mate with female worms in the subcutaneous tissue to produce between 700 and 1,500 microfilaria per day.
7. The microfilaria migrate to the skin during the day and the black flies only feed in the day, so the parasite is in a prime position for the female fly to ingest it. Black flies take blood meals to ingest these microfilaria to restart the cycle.

Signs and symptoms

Adult worms remain in subcutaneous nodules, limiting access to the host's immune system. Microfilariae, in contrast, are able to induce intense inflammatory responses, especially upon their death. Dying microfilariae have been recently discovered to release *Wolbachia Surface Protein* that activates TLR2 and TLR4, triggering innate immune responses and producing the inflammation and its associated morbidity. Wolbachia species have been found to be endosymbionts of *O. volvulus* adults and microfilariae, and are thought to be the driving force behind most of *O. volvulus* morbidity. The severity of illness is directly proportional to the number of infected microfilariae and the power of the resultant inflammatory response.

Skin involvement typically consists of intense itching, swelling, and inflammation. A grading system has developed to categorize the degree of skin involvement:

- Acute papular onchodermatitis - scattered pruritic papules;
- Chronic papular onchodermatitis - larger papule, resulting in hyperpigmentation;
- Lichenified onchodermatitis - hyperpigmented papules and plaques, with edema, lymphadenopathy, pruritus and common secondary bacterial infections;
- Skin atrophy - loss of elasticity, skin resembles tissue paper, 'lizard skin' appearance;
- Depigmentation - 'leopard skin' appearance, usually on anterior lower leg.

Ocular involvement provides the common name associated with onchocerciasis, river blindness and may involve any part of the eye from conjunctiva and cornea to uvea and posterior segment including retina and optic nerve. The microfilariae migrate to the surface of the cornea. Punctate keratitis occurs in the infected area. This clears up as the inflammation subsides. However, if the infection is chronic, sclerosing keratitis can occur, making the affected area become opaque. Over time the entire cornea may become opaque, thus leading to blindness. There is some evidence to suggest that the effect on the cornea is caused by an immune response to bacteria present in the worms.

The Mazzotti reaction, first described in 1948, is a symptom complex seen in patients after undergoing treatment of onchocerciasis with the medication diethylcarbamazine (DEC). Mazzotti reactions can be life-threatening and are characterized by fever, urticaria, swollen and tender lymph nodes, tachycardia, hypotension, arthralgias, oedema, and abdominal pain that occur within seven days of treatment of microfilariasis. The phenomenon is so common when DEC is used for the treatment of onchocerciasis that this drug is the basis of a skin patch test used to confirm that diagnosis. The drug patch is placed on the skin, and if the patient is infected with the microfilaria of Onchocerca volvulus, localized pruritus and urticaria are seen at the application site.

Prevention

There are various control programs that aim to stop onchocerciasis from being a public health problem. The first was the Onchocerciasis Control Programme (OCP), which was launched in 1974 and at its peak covered 30 million people in eleven countries. Through the use of larvicide spraying of fast flowing rivers to control black fly populations and, from 1988 onwards, the use of ivermectin to treat infected people, the OCP eliminated onchocerciasis as a public health problem. The OCP, a joint effort of the World Health Organisation, the World Bank, the United Nations Development Programme and the UN Food and Agriculture Organization, was considered to be a success and came to an end in 2002. Continued monitoring ensures that onchocerciasis cannot reinvade the area of the OCP.

In 1992 the Onchocerciasis Elimination Programme for the Americas (OEPA) was launched. The OEPA also relies on ivermectin.

In 1995 the African Programme for Onchocerciasis Control (APOC) began covering another nineteen countries and mainly relying upon the use of ivermectin. Its goal is to

set up a community-directed supply of ivermectin for those who are infected. In these ways, transmission has declined.

Treatment

The burden of onchocerciasis: children leading blind adults in Africa.

In mass drug administration (MDA) programmes, the treatment for onchocerciasis is ivermectin (trade name: Mectizan); infected people can be treated with two doses of ivermectin, six months apart, repeated every three years. The drug paralyses and kills the microfilariae, causing fever, itching, and possibly oedema, arthritis and lymphadenopathy. Intense skin itching is eventually relieved and progression towards blindness halted. In addition, while the drug does not kill the adult worm, it does prevent them from producing additional offspring. The drug therefore prevents both morbidity and transmission.

Ivermectin treatment is particularly effective because it only needs to be taken once or twice a year, needs no refrigeration, and has a wide margin of safety, with the result that it has been widely given by minimally trained community health workers.

Antibiotics

For the treatment of individuals, doxycycline is used to kill the Wolbachia bacteria that lives in adult worms. This adjunct therapy has been shown to significantly lower microfilarial loads in the host, and may have activity against the adult worms, due to the symbiotic relationship between Wolbachia and the worm. In four separate trials over ten years with various dosing regimes of doxycycline for individualized treatment, doxycycline was found to be effective in sterilizing the female worms and reducing their numbers over a period of four to six weeks. Research on other antibiotics such as rifampicin has shown it to been effective in animal models at reducing Wolbachia both as an alternative and as an adjunct to doxycycline. However, doxycycline treatment requires daily dosing for at least four to six weeks, making it more difficult to administer in the affected areas.

Ivermectin

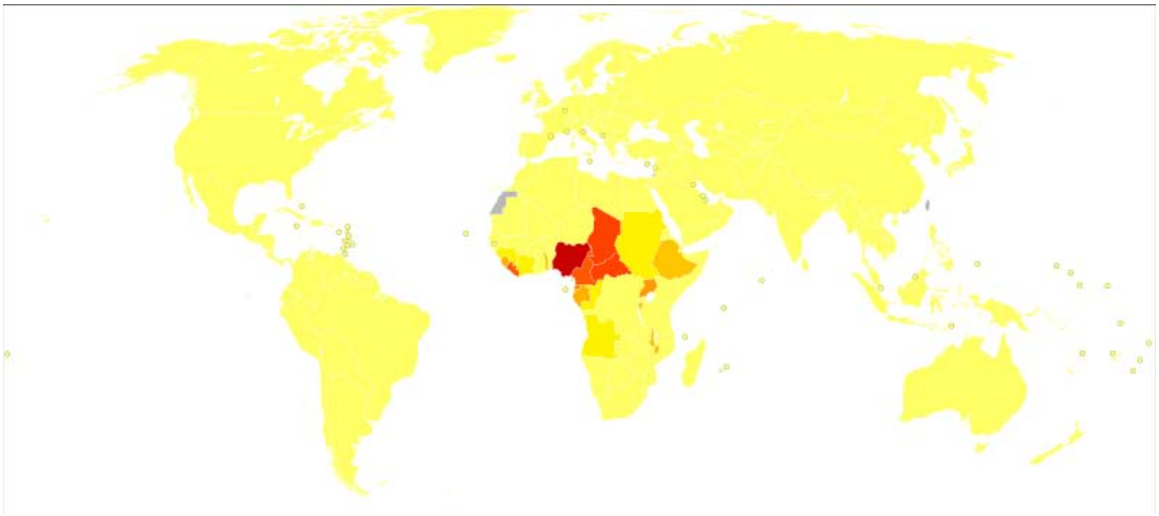
Ivermectin kills the parasite by interfering with the nervous system and muscle function, in particular, by enhancing inhibitory neurotransmission. The drug binds to and activates glutamate-gated chloride channels (GluCls). These channels, present in neurons and myocytes, are not invertebrate-specific, but are protected in vertebrates from the action of ivermectin by the blood-brain barrier. Ivermectin is thought to irreversibly activate these channel receptors in the worm, eventually causing an inhibitory postsynaptic potential (IPSP). The chance of a future action potential occurring in synapses between neurons decreases and the nematodes experience flaccid paralysis followed by death.

Ivermectin is directly effective against the larval stage microfilariae *Onchocerca volvulus*, which it paralyzes so that they can be killed by eosinophils and macrophages. Ivermectin does not kill adult females (macrofilariae) but causes them to cease releasing microfilariae, perhaps by paralyzing the reproductive tract.

Since 1988, ivermectin has been provided free of charge for use in humans by Merck through the Mectizan donation program (MDP). The MDP works together with ministries of health and non-governmental development organisations such as the World Health Organization to provide free ivermectin to those who need it in endemic areas.

A study of 2501 people in Ghana showed that the prevalence rate doubled between 2000 and 2005 despite treatment, suggesting that the parasite is developing resistance to the drug. A clinical trial of another antiparasitic agent, moxidectin (manufactured by Wyeth), began on July 1, 2009 (NCT00790998).

Epidemiology



Disability-adjusted life year for onchocerciasis per 100,000 inhabitants.
- no data
- less than 10
- 10-50
- 50-60
- 60-70
- 70-80
- 80-90
- 90-100
- 100-150
- 150-200
- 200-300
- 300-400
- more than 400

99% of onchocerciasis cases occur in Africa. As of 2008 about 18 million people were infected with this parasite; approximately 300,000 had been permanently blinded. Onchocerciasis is currently endemic in 30 African countries, Yemen, and isolated regions of South America. Travelers who do not stay long in those areas have little risk of developing the disease as it requires prolonged exposure to the fly bites and parasite introduction.

Onchocerciasis is endemic in 36 countries across Africa, Latin America and Yemen. Over 85 million people live in endemic areas and half of these reside in Nigeria. Another 120 million people are at risk for contracting the disease. Due to the vector's breeding habitat, the disease is more severe along the major rivers in the northern and central areas of the continent, and severity declines in villages farther from rivers.

According to a 2002 WHO report, Onchocerciasis has not caused a single death, but its global burden is 987,000 disability adjusted life years (DALYs). The severe pruritis alone

accounts for 60% of the DALYs. Infection reduces the host's immunity and resistance to other diseases. This results in an estimated reduction in life expectancy of 13 years.

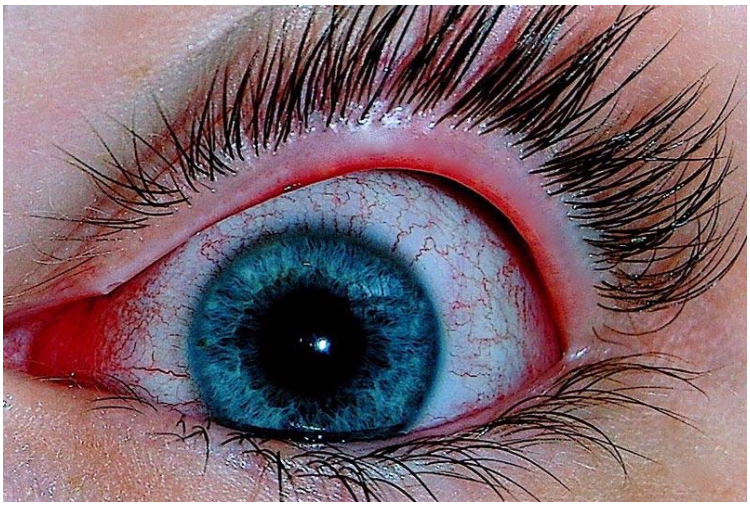
Research

Animal models for the disease are somewhat limited, as the parasite only lives in primates, but there are close parallels. *Litomosoides sigmodontis*, which will naturally infect cotton rats, has been found to fully develop in BALB/c mice. *Onchocerca ochengi*, the closest relative of *O. volvulus*, lives in intradermal cavities in cattle and is also spread by blackflies. Both systems are useful but not exact animal models.

Chapter 16

Conjunctivitis

Conjunctivitis



An eye with viral conjunctivitis.

ICD-10	H10.
ICD-9	372.0
DiseasesDB	3067
MedlinePlus	001010
eMedicine	emerg/110
MeSH	D003231

Conjunctivitis (also called **pink eye** or **madras eye**) refers to inflammation of the conjunctiva (the outermost layer of the eye and the inner surface of the eyelids). It is most commonly due to an infection (usually viral, but sometimes bacterial) or an allergic reaction.

Classification

Classification can be either by cause or by extent of the inflamed area.

By cause

- Allergic conjunctivitis
- Bacterial conjunctivitis
- Viral conjunctivitis
- Chemical conjunctivitis
- Neonatal conjunctivitis is often defined separately due to different organisms

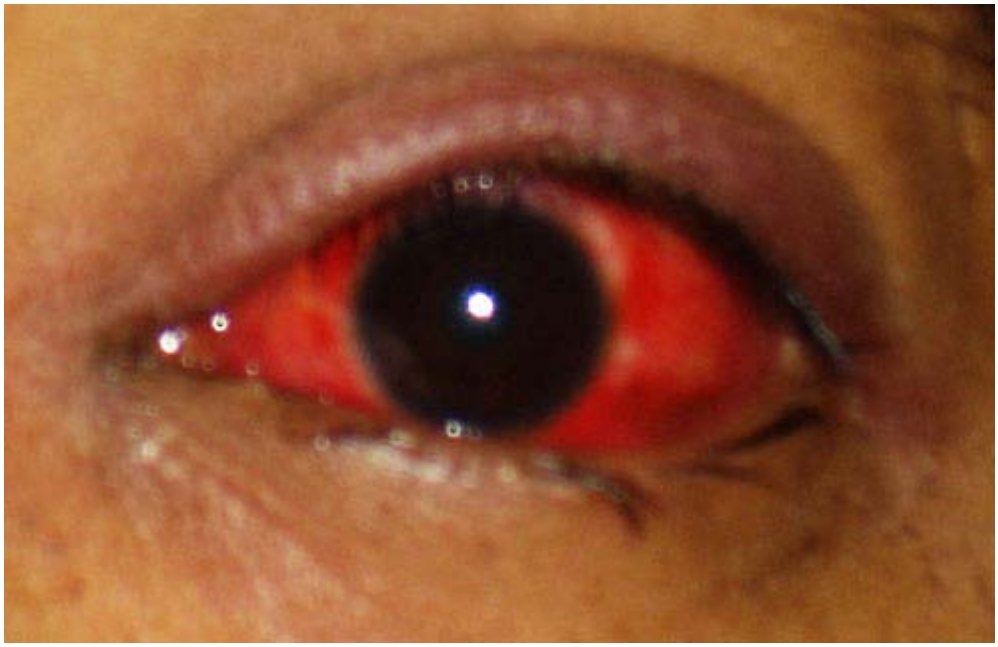
By extent of involvement

Blepharoconjunctivitis is the dual combination of conjunctivitis with blepharitis (inflammation of the eyelids).

Keratoconjunctivitis is the combination of conjunctivitis and keratitis (corneal inflammation).

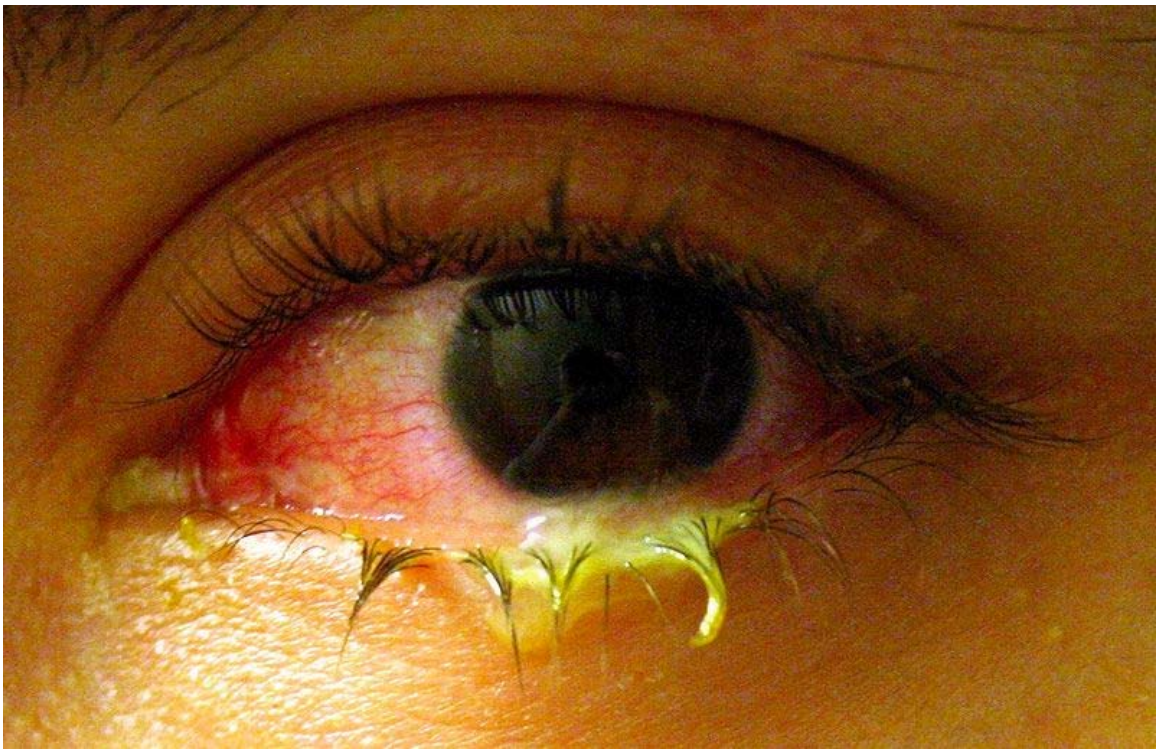
Episcleritis is an inflammatory condition that produces a similar appearance to conjunctivitis, but without discharge or tearing.

Signs and symptoms



An eye, red due to acute conjunctivitis

Eyes with conjunctivitis



An eye with bacterial conjunctivitis

Red eye (hyperaemia), irritation (chemosis) and watering (epiphora) of the eyes are symptoms common to all forms of conjunctivitis. However the pupils should be normally reactive and the visual acuity normal.

Viral

Viral conjunctivitis is often associated with an infection of the upper respiratory tract, a common cold, and/or a sore throat. Its symptoms include watery discharge and variable itch. The infection usually begins with one eye, but may spread easily to the other.

Viral conjunctivitis, commonly known as "pink eye" shows a fine diffuse pinkness of the conjunctiva which is easily mistaken for the 'ciliary injection' of iritis, but there are

usually corroborative signs on microscopy, particularly numerous lymphoid follicles on the tarsal conjunctiva, and sometimes a punctate keratitis.

Bacterial

Bacterial conjunctivitis due to the common pyogenic (pus-producing) bacteria causes marked grittiness/irritation and a stringy, opaque, greyish or yellowish mucopurulent discharge that may cause the lids to stick together, especially after sleep. Another symptom that could be caused by bacterial conjunctivitis is severe crusting of the infected eye and the surrounding skin. However discharge is not essential to the diagnosis, contrary to popular belief. Bacteria such as *Chlamydia trachomatis* or *Moraxella* can cause a non-exudative but persistent conjunctivitis without much redness. The gritty and/or scratchy feeling is sometimes localized enough for patients to insist they must have a foreign body in the eye. The more acute pyogenic infections can be painful. Like viral conjunctivitis, it usually affects only one eye but may spread easily to the other eye. However, it is dormant in the eye for three days before the patient shows signs of symptoms.

Chemical

Chemical eye injury is due to either an acidic or alkali substance getting in the eye. Alkalis are typically worse than acidic burns. Mild burns will produce conjunctivitis while more severe burns may cause the cornea to turn white. Litmus paper is an easy way to rule out the diagnosis by verifying that the pH is within the normal range of 7.0—7.2. Large volumes of irrigation is the treatment of choice and should continue until the pH is 6—8. Local anaesthetic eye drops can be used to decrease the pain.

Irritant or toxic conjunctivitis show primarily marked redness. If due to splash injury, it is often present only in the lower conjunctival sac. With some chemicals, above all, with caustic alkalis such as sodium hydroxide—there may be necrosis of the conjunctiva with a deceptively white eye due to vascular closure, followed by sloughing of the dead epithelium. This is likely to be associated with slit-lamp evidence of anterior uveitis.

Other

Inclusion conjunctivitis of the newborn (ICN) is a conjunctivitis that may be caused by the bacteria *Chlamydia trachomatis*, and may lead to acute, purulent conjunctivitis. However, it is usually self-healing.

Conjunctivitis is identified by irritation and redness of the conjunctiva. Except in obvious pyogenic or toxic/chemical conjunctivitis, a slit lamp (biomicroscope) is needed to have any confidence in the diagnosis. Examination of the tarsal conjunctiva is usually more diagnostic than the bulbar conjunctiva.

Causes

Conjunctivitis is most commonly caused by viral infection, but bacterial infections, allergies, other irritants and dryness are also common etiologies for its occurrence. Both bacterial and viral infections are contagious. Commonly, conjunctival infections are passed from person-to-person, but can also spread through contaminated objects or water.

The most common cause of viral conjunctivitis is adenoviruses. Herpetic keratoconjunctivitis (caused by herpes simplex viruses) can be serious and requires treatment with acyclovir. Acute hemorrhagic conjunctivitis is a highly contagious disease caused by one of two enteroviruses, Enterovirus 70 and Coxsackievirus A24. These were first identified in an outbreak in Ghana in 1969 and have spread worldwide since then, causing several epidemics.

Differential diagnosis

Conjunctivitis is a relatively non-specific symptom. Even after bio microscopy, laboratory tests are often necessary if proof of etiology is needed.

A purulent discharge (a whitish-yellow, yellow or yellow-brown substance more commonly known as pus) strongly suggests a cause from fecal matter, unless there is known exposure to toxins. It can also be caused by bacteria from feces, pet hair, or by smoke or other fumes. Infection with *Neisseria gonorrhoeae* should be suspected if the discharge is particularly thick and copious.

Itching (rubbing eyes) is the hallmark symptom of allergic conjunctivitis. Other symptoms include past history of eczema, or asthma.

A diffuse, less "injected" conjunctivitis (looking pink rather than red) suggests a viral cause, especially if numerous follicles are present on the lower tarsal conjunctiva on bio microscopy.

Scarring of the tarsal conjunctiva suggests trachoma, especially if seen in endemic areas, if the scarring is linear (von Arlt's line), or if there is also corneal vascularisation.

Clinical tests for lagophthalmos, dry eye (Schirmer test) and unstable tear film may help distinguish the various types of conjunctivitis.

Other symptoms including pain, blurring of vision and photophobia should not be prominent in conjunctivitis. Fluctuating blurring is common, due to tearing and mucoid discharge. Mild photophobia is common. However, if any of these symptoms are prominent, it is important to exclude other diseases such as glaucoma, uveitis, keratitis and even meningitis or caroticocavernous fistula.

Many people who have conjunctivitis have trouble opening their eyes in the morning because of the dried mucus on their eyelids. There is often excess mucus over the eye after sleeping for an extended period.

Diagnosis

These are done infrequently because most cases of conjunctivitis are treated empirically and (eventually) successfully, but often only after running the gamut of the common possibilities.

Swabs for bacterial culture are necessary if the history and signs suggest bacterial conjunctivitis, but there is no response to topical antibiotics. Research studies indicate that many bacteria implicated in low-grade conjunctivitis are not detected by the usual culture methods of medical microbiology labs, so false negative results are common. Viral culture may be appropriate in epidemic case clusters. Conjunctival scrapes for cytology can be useful in detecting chlamydial and fungal infections, allergy and dysplasia, but are rarely done because of the cost and the general lack of laboratory staff experienced in handling ocular specimens. Conjunctival incisional biopsy is occasionally done when granulomatous diseases (e.g., sarcoidosis) or dysplasia are suspected.

Management

Conjunctivitis resolves in 65% of cases without treatment, within 2 – 5 days. The prescribing of antibiotics to most cases is not necessary.

Allergic

For the allergic type, cool water poured over the face with the head inclined downward constricts capillaries, and artificial tears sometimes relieve discomfort in mild cases. In more severe cases, non-steroidal anti-inflammatory medications and antihistamines may be prescribed. Persistent allergic conjunctivitis may also require topical steroid drops.

Bacterial

Bacterial conjunctivitis usually resolves without treatment. Antibiotics, eye drops, or ointment are thus only needed if no improvement is observed after 3 days. In patients receiving no antibiotics recovery was in 4.8 days, immediate antibiotics 3.3 days, delayed antibiotics 3.9 days. No serious effects were noted either with or without treatment.

Viral

Although there is no specific treatment for viral conjunctivitis, symptomatic relief may be achieved with cold compresses and artificial tears. People are often advised to avoid touching their eyes or sharing towels and washcloths.

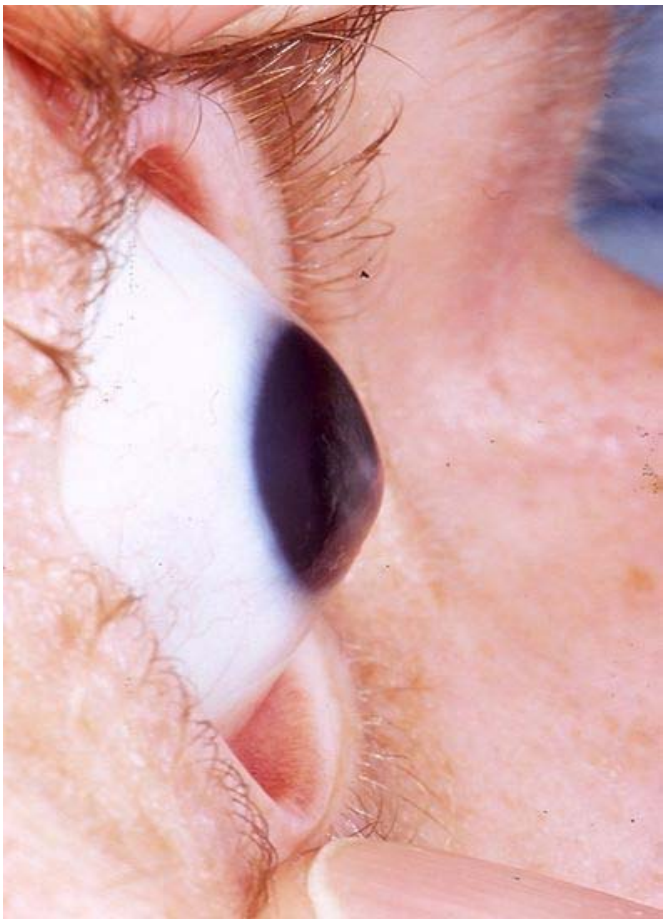
Chemical

Conjunctivitis due to chemicals is treated via irrigation with Ringer's lactate or saline solution. Chemical injuries (particularly alkali burns) are medical emergencies as they can lead to severe scarring, and intraocular damage.Do not touch your eyes. Even if you washed your hands still no touching. This may cause it to spread on to another eye.

Chapter 17

Keratoconus

Keratoconus



The "conical cornea" that is characteristic of keratoconus

ICD-10	H18.6
ICD-9	371.6
OMIM	148300
DiseasesDB	7158

MedlinePlus	001013
eMedicine	oph/104

Keratoconus (from Greek: *kerato-* horn, cornea; and *konos* cone), is a degenerative disorder of the eye in which structural changes within the cornea cause it to thin and change to a more conical shape than its normal gradual curve.

Keratoconus can cause substantial distortion of vision, with multiple images, streaking and sensitivity to light all often reported by the patient. It is typically diagnosed in the patient's adolescent years and attains its most severe state in the twenties and thirties. If afflicting both eyes, the deterioration in vision can affect the patient's ability to drive a car or read normal print. In most cases, corrective lenses are effective enough to allow the patient to continue to drive legally and likewise function normally. Further progression of the disease may require surgery including intrastromal corneal ring segments, corneal collagen cross-linking, or corneal transplantation. However, despite the disease's unpredictable course, keratoconus can often be successfully managed with little or no impairment to the patient's quality of life.

Keratoconus affects around one person in a thousand. It seems to occur in populations throughout the world, although it occurs more frequently in certain ethnic groups such as South Asians. The exact cause of keratoconus is uncertain, but has been associated with detrimental enzyme activity within the cornea. A genetic link seems likely, as the incidence rate is greater if a family member has been diagnosed. The progression of keratoconus is rapid in patients having Down syndrome.

Signs and symptoms

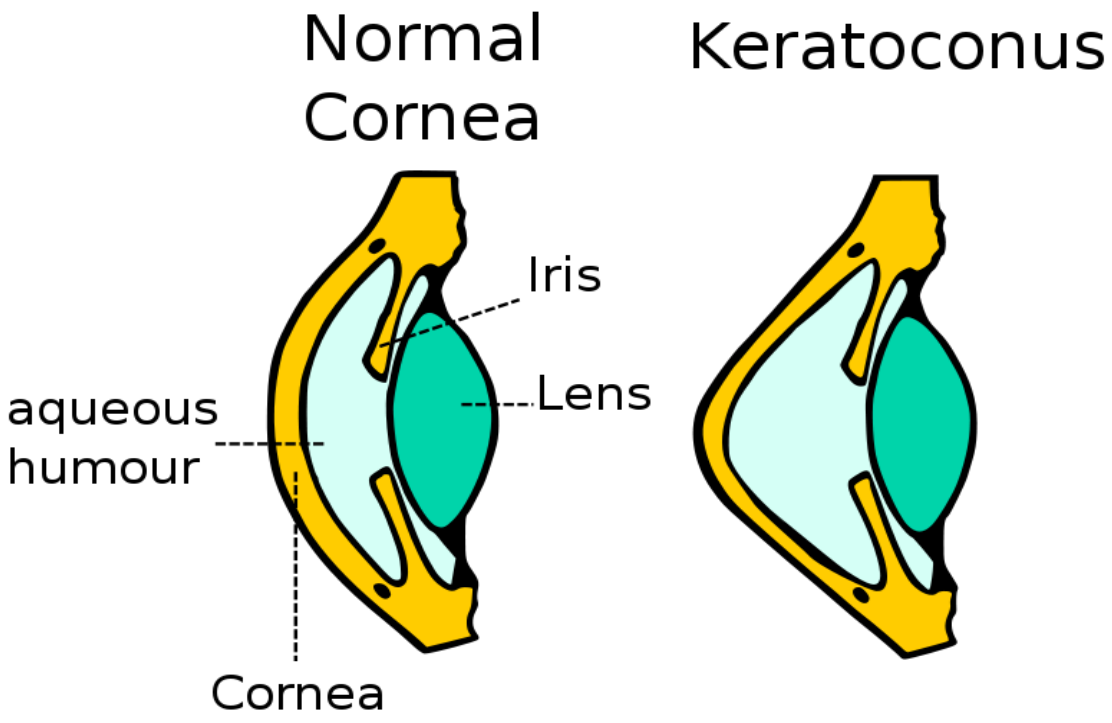
multiple images seen by a person with keratoconus.
"... a candle, when looked at, appears like a number of lights, confusedly running into one another" — Nottingham

People with early keratoconus typically notice a minor blurring of their vision and come to their clinician seeking corrective lenses for reading or driving. At early stages, the symptoms of keratoconus may be no different from those of any other refractive defect of the eye. As the disease progresses, vision deteriorates, sometimes rapidly. Visual acuity becomes impaired at all distances, and night vision is often quite poor. Some individuals have vision in one eye that is markedly worse than that in the other eye. The disease is often bilateral, though asymmetrical in many patients. Some develop photophobia (sensitivity to bright light), eye strain from squinting in order to read, or itching in the eye, but there is normally little or no sensation of pain.

The classic symptom of keratoconus is the perception of multiple 'ghost' images, known as monocular polyopia. This effect is most clearly seen with a high contrast field, such as a point of light on a dark background. Instead of seeing just one point, a person with keratoconus sees many images of the point, spread out in a chaotic pattern. This pattern does not typically change from day to day, but over time it often takes on new forms. Patients also commonly notice streaking and flaring distortion around light sources. Some even notice the images moving relative to one another in time with their heart beat.

The predominant optical aberration of the eye as an optical system in keratoconus is the so-called coma.

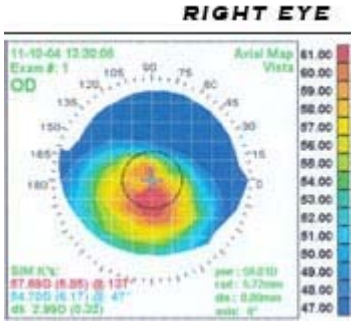
Diagnosis



A schematic digram showing change in cornea

Prior to any physical examination, the diagnosis of keratoconus frequently begins with an ophthalmologist's or optometrist's assessment of the patient's medical history, particularly the chief complaint and other visual symptoms, the presence of any history of ocular disease or injury which might affect vision, and the presence of any family history of ocular disease. An eye chart, such as a standard Snellen chart of progressively smaller letters, is then used to determine the patient's visual acuity. The eye examination may proceed to measurement of the localised curvature of the cornea with a manual keratometer, with detection of irregular astigmatism suggesting a possibility of keratoconus. Severe cases can exceed the instrument's measuring ability. A further indication can be provided by retinoscopy, in which a light beam is focused on the patient's retina and the reflection, or *reflex*, observed as the examiner tilts the light source back and forth. Keratoconus is amongst the ophthalmic conditions that exhibit a *scissor reflex* action of two bands moving toward and away from each other like the blades of a pair of scissors.

If keratoconus is suspected, the ophthalmologist or optometrist will search for other characteristic findings of the disease by means of slit lamp examination of the cornea. An advanced case is usually readily apparent to the examiner, and can provide for an unambiguous diagnosis prior to more specialised testing. Under close examination, a ring of yellow-brown to olive-green pigmentation known as a Fleischer ring can be observed in around half of keratoconic eyes. The Fleischer ring, caused by deposition of the iron oxide hemosiderin within the corneal epithelium, is subtle and may not be readily detectable in all cases, but becomes more evident when viewed under a cobalt blue filter. Similarly, around 50% of subjects exhibit Vogt's striae, fine stress lines within the cornea caused by stretching and thinning. The striae temporarily disappear while slight pressure is applied to the eyeball. A highly pronounced cone can create a V-shaped indentation in the lower eyelid when the patient's gaze is directed downwards, known as Munson's sign. Other clinical signs of keratoconus will normally have presented themselves long before Munson's sign becomes apparent, and so this finding, though a classic sign of the disease, tends not to be of primary diagnostic importance.



Corneal topogram of a keratoconic eye

A handheld keratoscope, sometimes known as *Placido's disk*, can provide a simple non-invasive visualization of the surface of the cornea by projecting a series of concentric rings of light onto the cornea. A more definitive diagnosis can be obtained using corneal

topography, in which an automated instrument projects the illuminated pattern onto the cornea and determines its topology from analysis of the digital image. The topographical map indicates any distortions or scarring in the cornea, with keratoconus revealed by a characteristic steepening of curvature which is usually below the centreline of the eye. The technique can record a snapshot of the degree and extent of the deformation as a benchmark for assessing its rate of progression. It is of particular value in detecting the disorder in its early stages when other signs have not yet presented.

Once keratoconus has been diagnosed, its degree may be classified by several metrics:

- The steepness of greatest curvature from *mild* (< 45 D), *advanced* (up to 52 D) or *severe* (> 52 D);
- The morphology of the cone: *nipple* (small: 5 mm and near-central), *oval* (larger, below-center and often sagging), or *globus* (more than 75% of cornea affected);
- The corneal thickness from mild (> 506 μm) to advanced (< 446 μm).

Increasing use of corneal topography has led to a decline in use of these terms.

Cause

A specimen of keratoconic cornea taken out six years after diagnosis: thin stroma, wrinkled posterior surface.

Despite considerable research, the etiology of keratoconus remains somewhat of a mystery. A number of sources suggest that keratoconus likely arises from a number of different factors: genetic, environmental or cellular, any of which may form the trigger for the onset of the disease.

A genetic predisposition to keratoconus has been observed, with the disease running in certain families, and incidences reported of concordance in identical twins. The frequency of occurrence in close family members is not clearly defined, though it is known to be considerably higher than that in the general population, and studies have obtained estimates ranging between 6% and 19%. A responsible gene has not been identified: two studies involving isolated, largely homogenetic communities have contrarily mapped putative gene locations to chromosomes 16q and 20q. However, most genetic studies agree on an autosomal dominant model of inheritance. Keratoconus is also diagnosed more often in people with Down syndrome, though the reasons for this link have not yet been determined. Keratoconus has been associated with atopic diseases, which include asthma, allergies, and eczema, and it is not uncommon for several or all of these diseases to affect one person. A number of studies suggest that vigorous eye rubbing contributes to the progression of keratoconus, and that patients should be discouraged from the practice.

Iatrogenic keratoconus has also been observed following LASIK surgery, caused by removal of excessive stromal bed tissue.

Pathophysiology

Once initiated, the disease normally develops by progressive dissolution of Bowman's layer, which lies between the corneal epithelium and stroma. As the two come into contact, cellular and structural changes in the cornea adversely affect its integrity and lead to the bulging and scarring that are characteristic of the disorder. Within any individual keratoconic cornea, there may be found regions of degenerative thinning coexisting with regions undergoing wound healing.

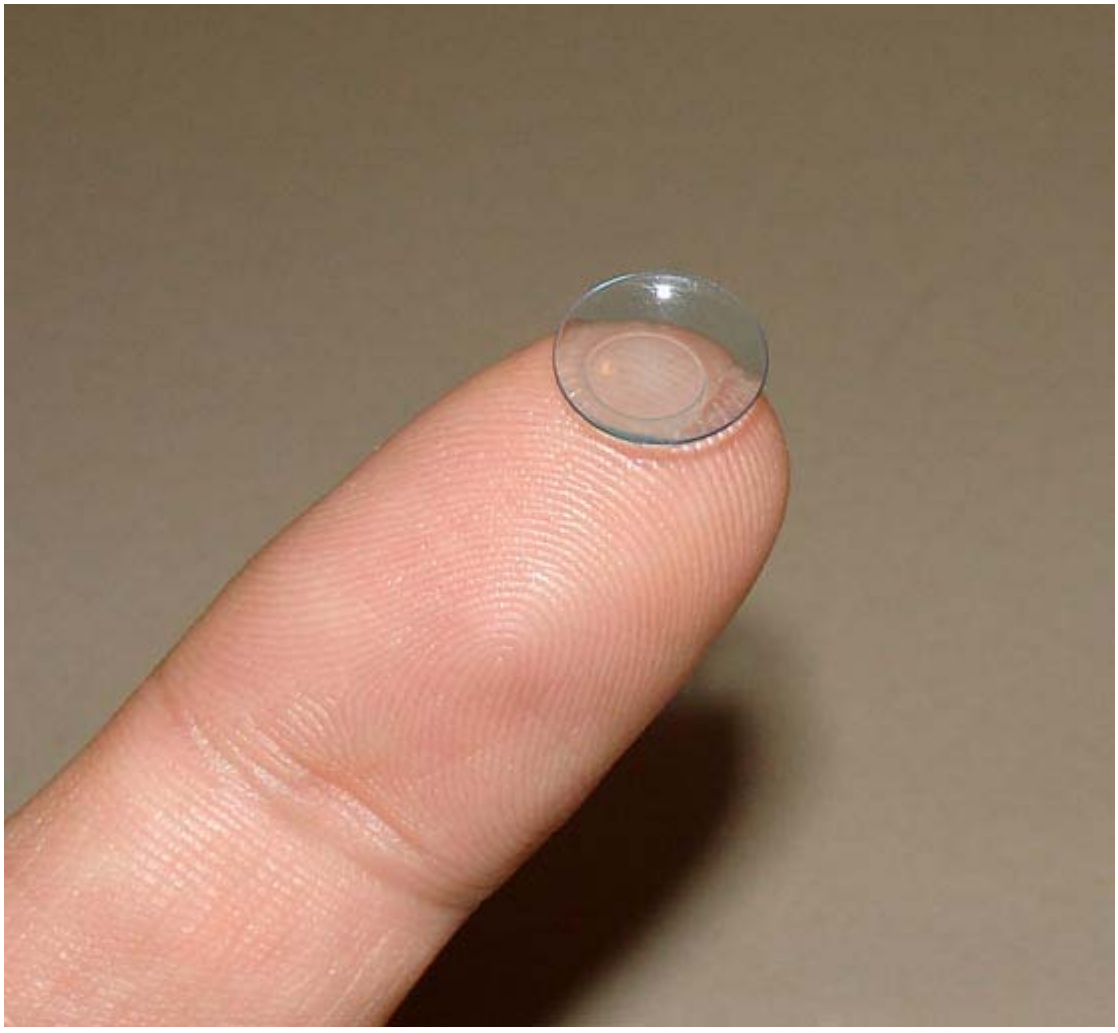
The visual distortion experienced by the patient comes from two sources, one being the irregular deformation of the surface of the cornea; the other being scarring that occurs on its exposed highpoints. These factors act to form regions on the cornea that map an image to different locations on the retina and give rise to the symptom of monocular polyopia. The effect can worsen in low light conditions as the dark-adapted pupil dilates to expose more of the irregular surface of the cornea. Scarring appears to be an aspect of the corneal degradation; however, a recent, large, multi-center study suggests that abrasion by contact lenses may increase the likelihood of this finding by a factor of over two. A number of patients complain of chronic eye rubbing and also think it as a possible cause to the disease but it is not so; however, it has been observed that keratoconus progresses faster due to eye-rubbing.

A number of studies have indicated that keratoconic corneas show signs of increased activity by proteases, a class of enzymes that break some of the collagen cross-linkages in

the stroma, with a simultaneous reduced expression of protease inhibitors. Other studies have suggested that reduced activity by the enzyme aldehyde dehydrogenase may be responsible for a build-up of free radicals and oxidising species in the cornea. It seems likely that, whatever the pathogenetical process, the damage caused by activity within the cornea results in a reduction in its thickness and biomechanical strength. While keratoconus is considered a non-inflammatory disorder, one study shows that rigid contact lens wear by patients leads to overexpression of pro-inflammatory cytokines, such as IL-6, TNF-alpha, ICAM-1, and VCAM-1 in the tear fluid.

Treatment

Contact lenses



Rigid gas permeable lens for keratoconus

In early stages of keratoconus, spectacles or soft contact lenses can suffice to correct for the mild astigmatism. As the condition progresses, these may no longer provide the patient with a satisfactory degree of visual acuity, and most clinical practitioners will

move to managing the condition with rigid contact lenses, known as rigid gas-permeables, or RGPs. RGP lenses provide a good level of visual correction, but do not arrest progression of the condition.

In keratoconic patients, rigid contact lenses improve vision by means of tear fluid filling the gap between the irregular corneal surface and the smooth regular inner surface of the lens, thereby creating the effect of a smoother cornea. Many specialized types of contact lenses have been developed for keratoconus, and affected people may seek out both doctors specialized in conditions of the cornea, and contact lens fitters who have experience managing patients with keratoconus. The irregular cone presents a challenge and the fitter will endeavour to produce a lens with the optimal contact, stability and steepness. Some trial-and-error fitting may prove necessary.

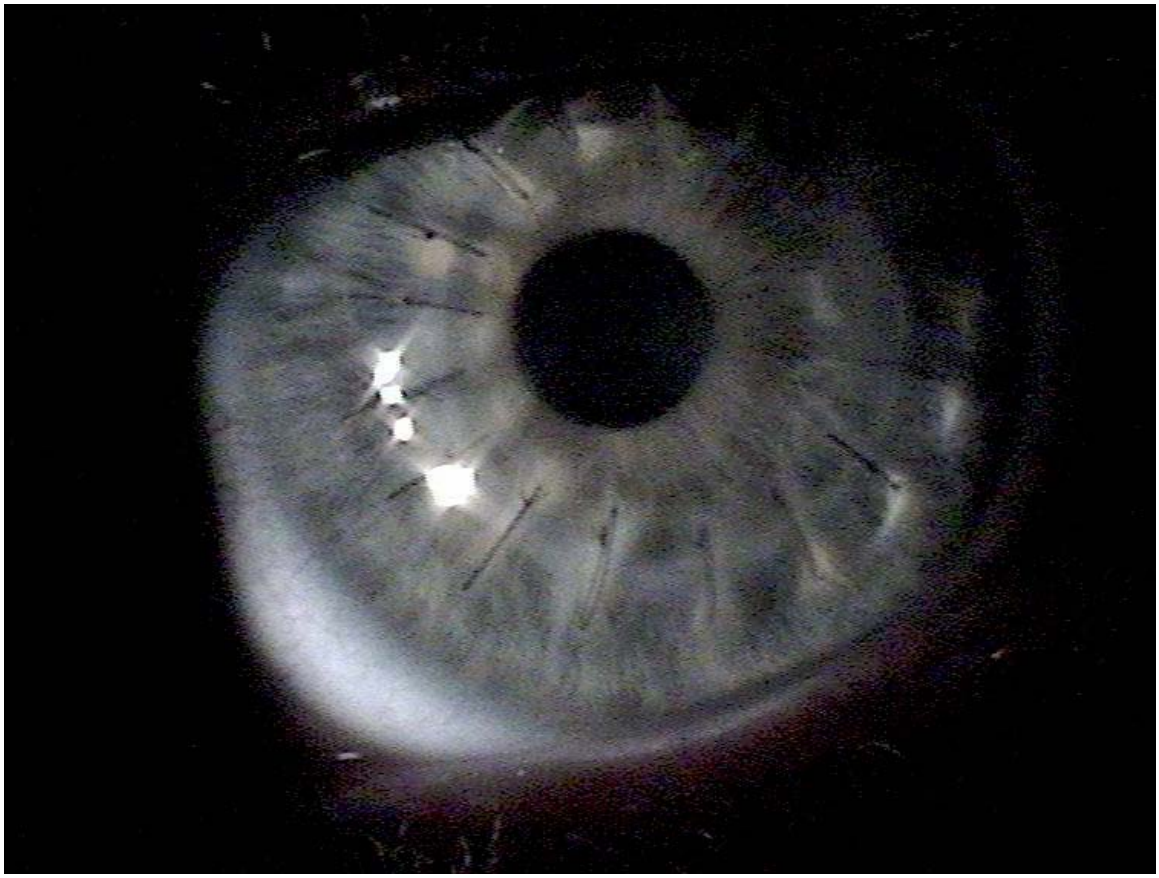
Traditionally, contact lenses for keratoconus have been the 'hard' or rigid gas-permeable variety, although manufacturers have also produced specialized 'soft' or hydrophilic lenses and, most recently, silicone hydrogel lenses. A soft lens has a tendency to conform to the conical shape of the cornea, thus diminishing its effect. To counter this, hybrid lenses have been developed which are hard in the centre and encompassed by a soft skirt. However, soft or earlier generation hybrid lenses did not prove effective for every patient. Early generation lenses like SoftPerm have been discontinued. The fourth generation of hybrid lens technology has improved significantly, giving more patients an option that combines the comfort of a soft lens with the visual acuity of an RGP lens. The new generation of technology fixes the issues that were prevalent in earlier generations and allows contact lenses to be fit for the majority of patients.

Some patients also find good vision correction and comfort with a "piggyback" lens combination, in which gas-permeable rigid lenses are worn over soft lenses, both providing a degree of vision correction. One form of piggyback lens makes use of a soft lens with a countersunk central area to accept the rigid lens. Fitting a piggyback lens combination requires experience on the part of the lens fitter, and tolerance on the part of the keratoconic patient.

Scleral lenses are sometimes prescribed for cases of advanced or very irregular keratoconus; these lenses cover a greater proportion of the surface of the eye and hence can offer improved stability. The larger size of the lenses may make them unappealing or uncomfortable to some; however, their easier handling can find favour with patients with reduced dexterity, such as the elderly.

Surgical options

Corneal transplant



Corneal transplant for keratoconus, approximately one week after surgery. Multiple light reflections indicate folds in the cornea which later resolved.

Between 10% and 25% of cases of keratoconus will progress to a point where vision correction is no longer possible, thinning of the cornea becomes excessive, or scarring as a result of contact lens wear causes problems of its own, and a corneal transplantation or *penetrating keratoplasty* becomes required. Keratoconus is the most common grounds for conducting a penetrating keratoplasty, generally accounting for around a quarter of such procedures. The corneal transplant surgeon trephines a lenticule of corneal tissue and then grafts the donor cornea to the existing eye tissue, usually using a combination of running and individual sutures. The cornea does not have a direct blood supply, and so donor tissue is not required to be blood type matched. Eye banks check the donor corneas for any disease or cellular irregularities.

Spanish-born eye surgeon Ramon Castroviejo successfully performed keratoplasty as early as 1936

The acute recovery period can take four to six weeks and full post-operative vision stabilization often takes a year or more but most transplants are very stable in the long term. The National Keratoconus Foundation reports that penetrating keratoplasty has the most successful outcome of all transplant procedures, and when performed for keratoconus in an otherwise healthy eye, its success rate can be 95% or greater. The sutures used usually dissolve over a period of three to five years but individual sutures can be removed during the healing process if they are causing irritation to the patient.

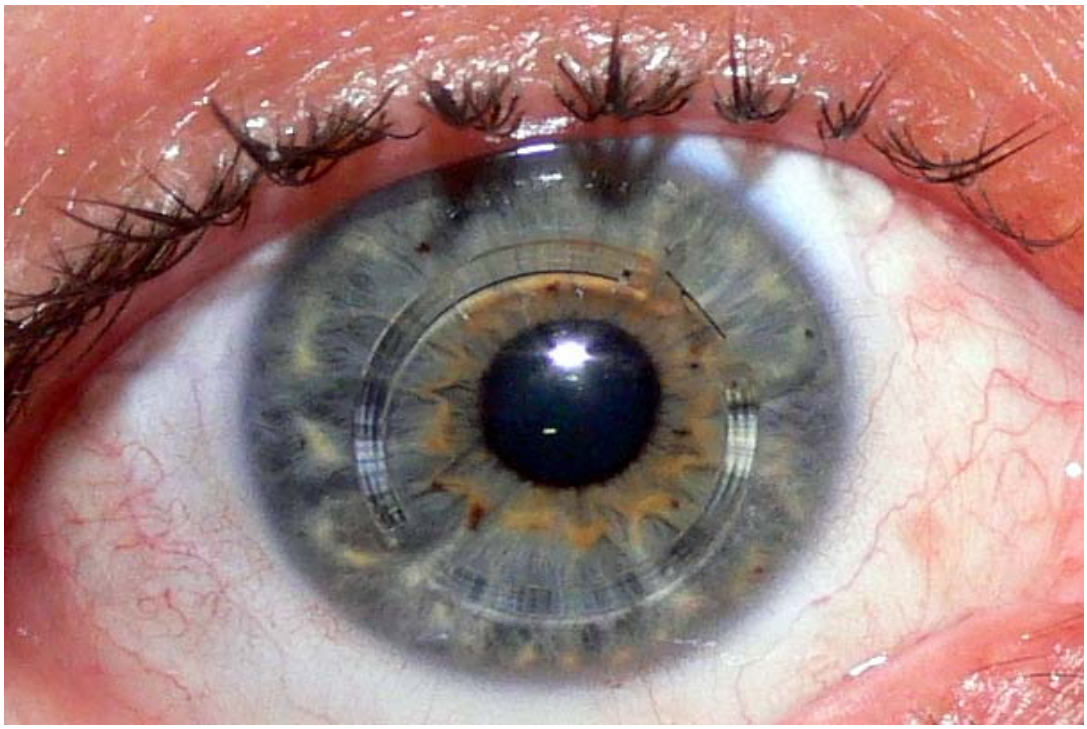
In the USA, corneal transplants (also known as corneal grafts) for keratoconus are usually performed under sedation as outpatient surgery. In other countries, such as Australia and the UK, the operation is commonly performed with the patient undergoing a general anaesthetic. All cases require a careful follow-up with an eye surgeon (ophthalmologist) for a number of years. Frequently, vision is greatly improved after the surgery, but even if the actual visual acuity does not improve, because the cornea is a more normal shape after the healing is completed, patients can more easily be fitted with corrective lenses. Complications of corneal transplants are mostly related to vascularization of the corneal tissue and rejection of the donor cornea. Vision loss is very rare, though difficult-to-correct vision is possible. When rejection is severe, repeat transplants are often attempted, and are frequently successful. Keratoconus will not normally reoccur in the

transplanted cornea; incidences of this have been observed, but are usually attributed to incomplete excision of the original cornea or inadequate screening of the donor tissue. The long-term outlook for corneal transplants performed for keratoconus is usually favorable once the initial healing period is completed and a few years have elapsed without problems.

Corneal ring segment inserts

A recent surgical alternative to corneal transplant is the insertion of intrastromal corneal ring segments. A small incision is made in the periphery of the cornea and two thin arcs of polymethyl methacrylate are slid between the layers of the stroma on either side of the pupil before the incision is closed. The segments push out against the curvature of the cornea, flattening the peak of the cone and returning it to a more natural shape. The procedure, carried out on an outpatient basis under local anaesthesia, offers the benefit of being reversible and even potentially exchangeable as it involves no removal of eye tissue.

The principal intrastromal ring available is known by the trade name of *Intacs*. Internationally, *Ferrara Rings* are also available. Intacs are a patented technology and are placed outside the optical zone versus the smaller prismatic Ferrara rings that are placed just inside the 5 mm optical zone. Intacs are the only corneal implants that have gone through the FDA Phase I, II and III clinical trials and were first approved by the Food and Drug Administration (FDA) in the United States in 1999 for myopia; this was extended to the treatment of keratoconus in July 2004.

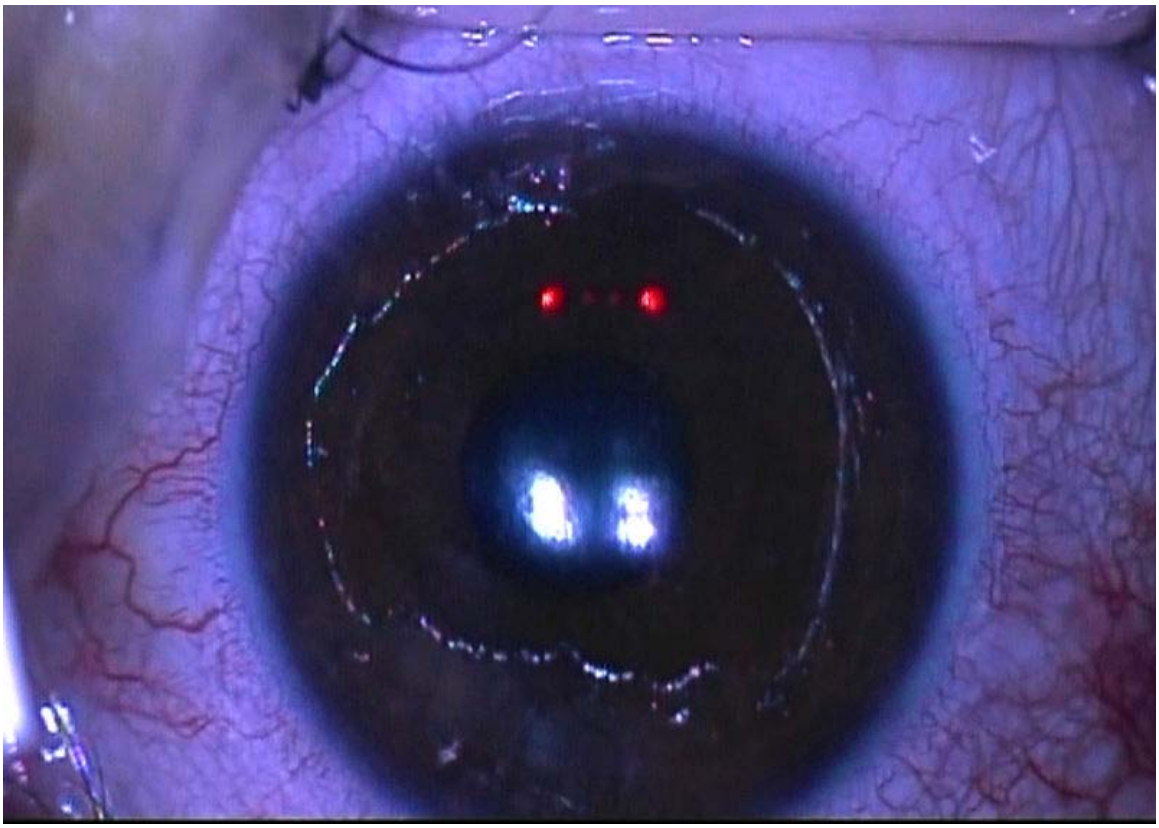


A pair of *Intacs* after insertion into the cornea

Clinical studies on the effectiveness of intrastromal rings on keratoconus are in their early stages, and results have so far been generally encouraging, though they have yet to enter into wide acceptance with the refractive surgery community. In common with penetrating keratoplasty, the requirement for some vision correction in the form of spectacles or hydrophilic contact lenses may remain subsequent to the operation. Potential complications of intrastromal rings include accidental penetration through to the anterior chamber when forming the channel, post-operative infection of the cornea, and migration or extrusion of the segments. The rings offer a good chance of vision improvement even in otherwise hard to manage eyes, but results are not guaranteed and in a few cases may worsen.

Early studies on intrastromal corneal rings involved use of two segments to cause global flattening of the cornea. A later study reported that better results could be obtained for those cones located more to the periphery of the cornea by using a single Intacs segment. This leads to preferential flattening of the cone below, but also to steepening the over-flat upper part of the cornea.

Corneal collagen crosslinking with riboflavin



Removed corneal epithelium during CCR operation on an eye with post-LASIK complication

A treatment developed at the Technische Universität Dresden, and which has shown early success is Corneal Collagen Crosslinking with Riboflavin, also known as CXL, CCR, CCL and KXL. A one-time application of riboflavin solution is administered to the eye and is activated by illumination with UV-A light for approximately 30 minutes. The riboflavin causes new bonds to form across adjacent collagen strands in the stromal layer of the cornea, which recovers and preserves some of the cornea's mechanical strength. The corneal epithelial layer is generally removed in order to increase penetration of the riboflavin into the stroma.

Clinical trials are ongoing, but crosslinking is seeing increasing adoption by the ophthalmological community, and has shown success in treating early cases of the disease. And early results from an Australian study are very promising in reporting stabilization in all treated eyes, and a slight correction in visual acuity in most patients. The procedure, with epithelium removed, is approved for use throughout Europe, and commenced clinical trials in the USA in 2008. Over 300 patients have now been treated in the United States in those trials, which are composed of two randomized, controlled, multi-site clinical trials for the treatment of progressive keratoconus and post LASIK ectasia. Avedro, Inc., the trial's sponsor, is closing the follow-up phase of the study and completing the necessary steps to file the results with the FDA.

In some cases, crosslinking may also be successfully combined with other treatment methods such as corneal ring segment inserts and Keraflex, a new refractive correction procedure, which recently received CE Mark in Europe. Corrective lenses may still be required after these treatments but with more normal prescriptions possible now, and these newer methods may have an important role in limiting deterioration of vision, increasing unaided/uncorrected vision and reducing the case for corneal transplantation.

Radial keratotomy

Radial keratotomy is a refractive surgery procedure where the surgeon makes a spoke-like pattern of incisions into the cornea to modify its shape. This early surgical option for myopia has been largely superseded by LASIK and other similar procedures. LASIK itself is absolutely contraindicated in keratoconus and other corneal thinning conditions as removal of corneal stromal tissue will further damage an already thin and weak cornea.

For similar reasons, radial keratotomy has also generally not been used for keratoconic patients. However, an Italian clinic has reported some success with a modified asymmetric radial keratotomy procedure, in which the incisions are confined to one sector of the eye. The corneal thickness is first measured using a pachymeter, then the surgeon makes cuts to a depth of 70–80% of the measured thickness. The patient may initially experience photophobia and fluctuation of vision, in common with other forms of refractive surgery. This technique has yet to go through the official experimentation and follow-up period which is generally required by the Italian National Health Service to accept a new surgery technique before it can be offered to patients.

DALK transplants

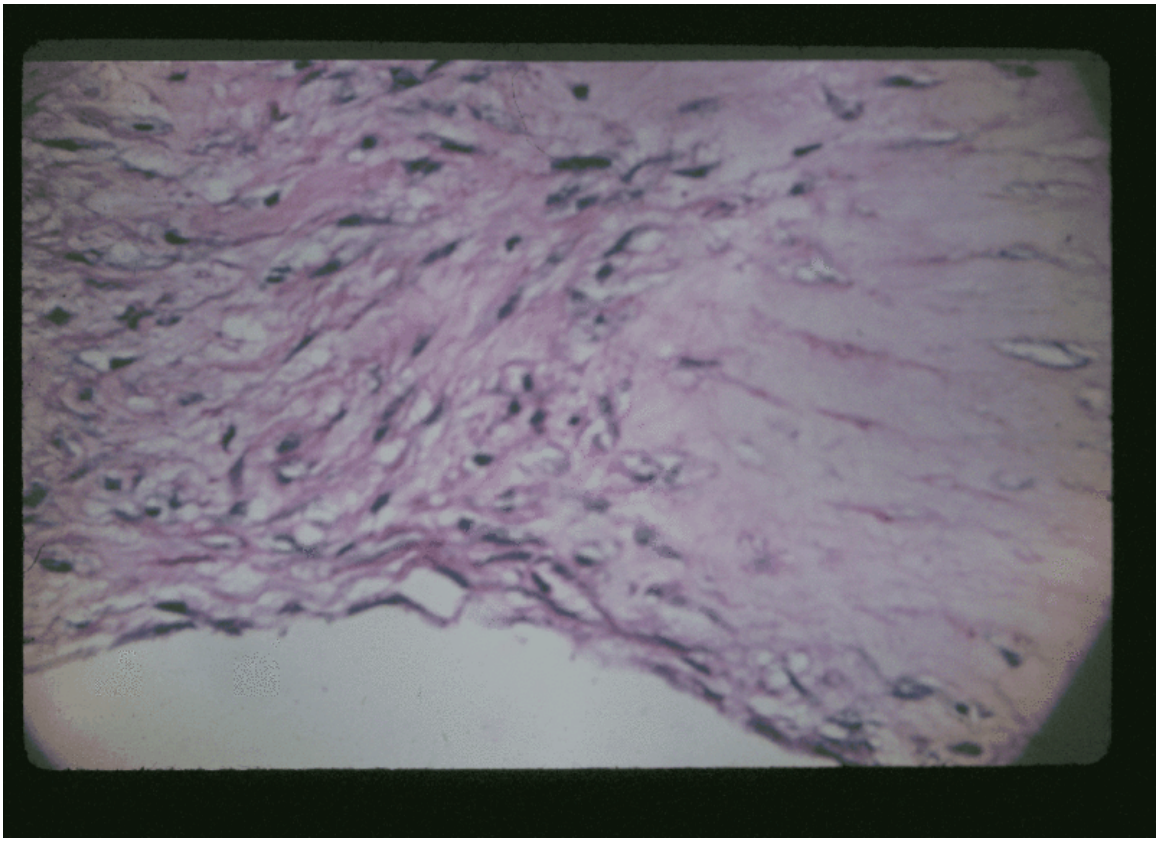
One way of reducing the risk of rejection is to use a newer technique called a *Deep Anterior Lamellar Keratoplasty*, referred to as *DALK*. In a DALK graft, only the outermost epithelium and the main bulk of the cornea, the stroma, are replaced; the patient's rearmost endothelium layer and the Descemet's membrane are left, giving some additional structural integrity to the post-graft cornea. Because a graft rejection usually begins in the endothelium, the chance of a rejection episode is greatly reduced. Furthermore, it is possible to transplant tissue from a donor which has been freeze-dried. The freeze-drying process ensures that this tissue is dead, so there is no chance of a rejection.

Some surgeons prefer to remove the donor epithelium, others leave the donor's cells in place. Removing it can cause a slight improvement in overall vision, but a corresponding increase in visual recovery time.

Epikeratophakia

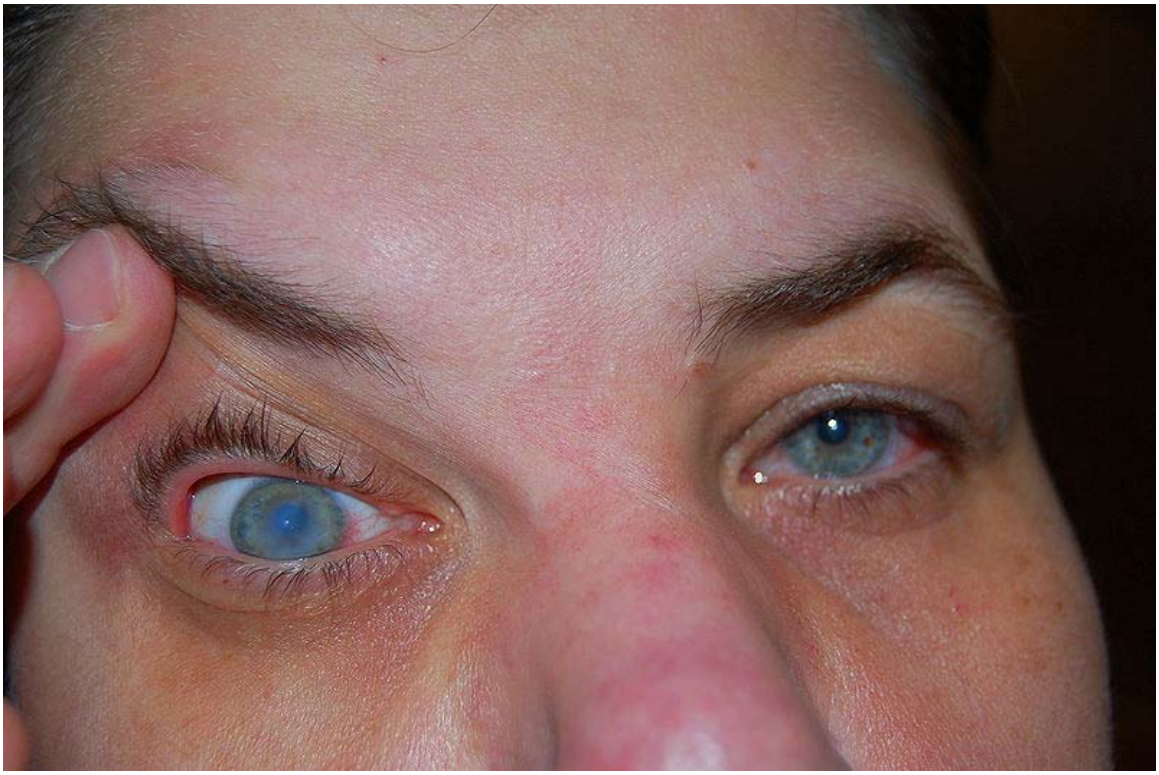
Rarely, a non-penetrating keratoplasty known as an *epikeratophakia* (or *epikeratoplasty*) may be performed in cases of keratoconus. The corneal epithelium is removed and a lenticule of donor cornea grafted on top of it. The procedure requires a greater level of skill on the part of the surgeon, and is less frequently performed than a penetrating keratoplasty as the outcome is generally less favorable. However, it may be seen as an option in a number of cases, particularly for young patients.

Prognosis



A small rupture of Descemet's membrane (magnified view)

Patients with keratoconus typically present initially with mild astigmatism, commonly at the onset of puberty, and are diagnosed as having the disease by the late teenage years or early 20s. In rare cases keratoconus can occur in children or is not present until later adulthood. A diagnosis of the disease at an early age may indicate a greater risk of severity in later life. Patients' vision will seem to fluctuate over a period of months, driving them to change lens prescriptions frequently, but as the condition worsens, contact lenses are required in the majority of cases. The course of the disorder can be quite variable, with some patients remaining stable for years or indefinitely, while others progress rapidly or experience occasional exacerbations over a long and otherwise steady course. Most commonly, keratoconus progresses for a period of ten to twenty years before the course of the disease generally ceases.



leads to *corneal hydrops*

In advanced cases, bulging of the cornea can result in a localized rupture of Descemet's membrane, an inner layer of the cornea. Aqueous humor from the eye's anterior chamber seeps into the cornea before Descemet's membrane reseals. The patient experiences pain and a sudden severe clouding of vision, with the cornea taking on a translucent milky-white appearance known as a *corneal hydrops*. Although disconcerting to the patient, the effect is normally temporary and after a period of six to eight weeks the cornea usually returns to its former transparency. The recovery can be aided non-surgically by bandaging with an osmotic saline solution. Although a hydrops usually causes increased scarring of the cornea, occasionally it will benefit a patient by creating a flatter cone, aiding the fitting of contact lenses. Occasionally, in extreme cases, the cornea thins to the point that a partial rupture occurs, resulting in a small, bead-like swelling on the cornea that has been filled with fluid. When this occurs, a corneal transplant can become urgently necessary to avoid complete rupture and resulting loss of the eye.

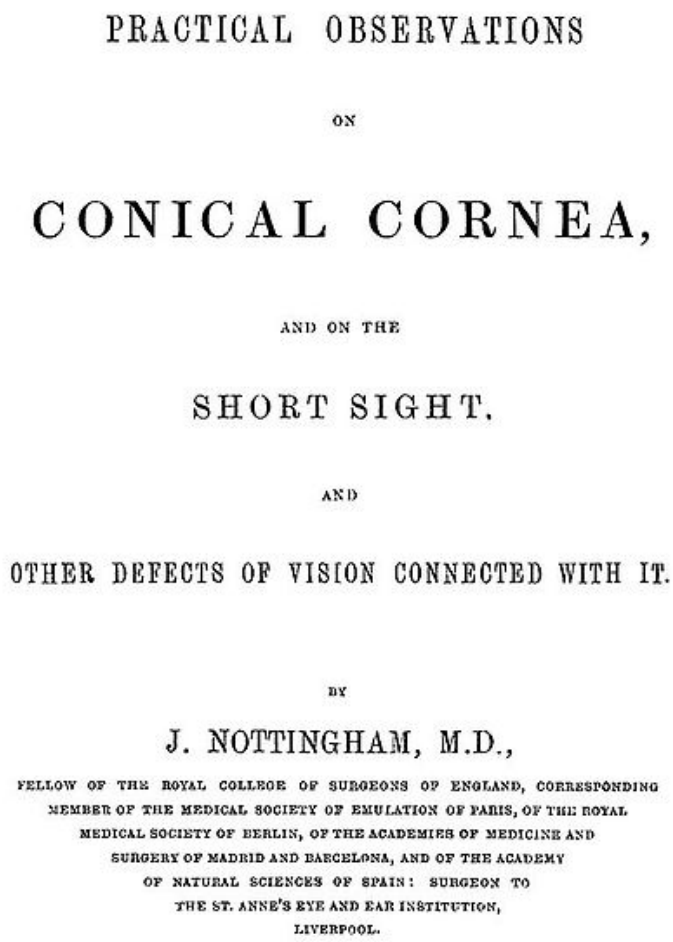
Epidemiology

The National Eye Institute reports that keratoconus is the most common corneal dystrophy in the United States, affecting approximately 1 in 2,000 Americans, but some reports place the figure as high as 1 in 500. The inconsistency may be due to variations in diagnostic criteria, with some cases of severe astigmatism interpreted as those of keratoconus, and vice versa. A long-term study found a mean incidence rate of 2.0 new cases per 100,000 population per year. It is suggested that males and females, and all

ethnicities appear equally susceptible, though some recent studies have cast doubt upon this, suggesting a higher prevalence amongst females; the literature, however, varying as to its extent. Also, a study carried out in the UK suggests that people of a South Asian heritage are 4.4 times as likely to suffer from keratoconus as Caucasians, and are also more likely to be affected with the condition earlier.

Keratoconus is normally bilateral (affecting both eyes) although the distortion is usually asymmetric and is rarely completely identical in both corneas. Unilateral cases tend to be uncommon, and may in fact be very rare if a very mild condition in the better eye is simply below the limit of clinical detection. It is common for keratoconus to be diagnosed first in one eye and not until later in the other. As the condition then progresses in both eyes, the vision in the earlier-diagnosed eye will often persist to be poorer than that in its fellow.

History



PRACTICAL OBSERVATIONS

ON

CONICAL CORNEA,

AND ON THE

SHORT SIGHT,

AND

OTHER DEFECTS OF VISION CONNECTED WITH IT.

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Practical observations on conical cornea, Nottingham's ground-breaking text on keratoconus, 1854

In a 1748 doctoral dissertation, the German oculist Burchard Mauchart provided an early description of a case of keratoconus, which he called *staphyloma diaphanum*. However, it was not until 1854 that British physician John Nottingham clearly described keratoconus and distinguished it from other ectasias of the cornea. Nottingham reported the cases of "conical cornea" that had come to his attention, and described several classic features of the disease, including polyopia, weakness of the cornea, and difficulty matching corrective lenses to the patient's vision. In 1859 British surgeon William Bowman used an ophthalmoscope (recently invented by Hermann von Helmholtz) to diagnose keratoconus, and described how to angle the instrument's mirror so as to best see the conical shape of the cornea. Bowman also attempted to restore the vision by pulling on the iris with a fine hook inserted through the cornea and stretching the pupil into a vertical slit, like that of a cat. He reported that he had had a measure of success with the technique, restoring vision to an 18-year old woman who had previously been unable to count fingers at a distance of 8 inches (20 cm). By 1869, when the pioneering Swiss ophthalmologist Johann Horner wrote a thesis entitled *On the treatment of keratoconus*, the disorder had acquired its current name. The treatment at that time, endorsed by the leading German ophthalmologist Albrecht von Graefe, was an attempt to physically reshape the cornea by chemical cauterization with a silver nitrate solution and application of a miosis-causing agent with a pressure dressing. In 1888 the treatment of keratoconus became one of the first practical applications of the then newly invented contact lens, when the French physician Eugène Kalt manufactured a glass scleral shell which improved vision by compressing the cornea into a more regular shape. Since the start of the twentieth century, research on keratoconus has both improved understanding of the disease and greatly expanded the range of treatment options. The first successful transplantation of cornea to treat keratoconus was done in 1936 by Ramon Castroviejo.

Related disorders

Several other non-inflammatory eye disorders, generally rarer than keratoconus, also cause thinning of the cornea:

Keratoglobus
: Keratoglobus is a very rare condition that causes corneal thinning primarily at the margins, resulting in a spherical, slightly enlarged eye. It may be genetically related to keratoconus.

Pellucid Marginal Degeneration
: Pellucid marginal degeneration causes thinning of a narrow (1–2 mm) band of the cornea, usually along the inferior corneal margin. It causes irregular astigmatism that can often be corrected by spectacles. Differential diagnosis may be made by slit-lamp examination.

Posterior keratoconus
: Keratoconus and posterior keratoconus are distinct disorders, despite their similar names. Posterior keratoconus is a rare abnormality, usually congenital, which causes a non-progressive thinning of the inner surface of the cornea, while the curvature of the anterior surface remains normal. Normally only a single eye is affected.

Chapter 18

Keratoconjunctivitis Sicca

Keratoconjunctivitis sicca	
ICD-10	H19.3, M35.0 (ILDS M35.010)
ICD-9	370.33, 710.2
DiseasesDB	12155
eMedicine	article/1196733 article/1210417
MeSH	D007638

Keratoconjunctivitis sicca (KCS), also called **keratitis sicca**, **xerophthalmia**, **dry eye syndrome** (**DES**), or simply **dry eyes**, is an eye disease caused by decreased tear production or increased tear film evaporation commonly found in humans and some animals. The phrase "keratoconjunctivitis sicca" is Latin, and its literal translation is "dryness of the cornea and conjunctiva".

Symptoms

Typical symptoms of keratoconjunctivitis are dryness, burning and a sandy-gritty eye irritation that gets worse as the day goes on. Symptoms may also be described as itchy, scratchy, stingy or tired eyes. Other symptoms are pain, redness, a pulling sensation, and pressure behind the eye. There may be a feeling that something, such as a speck of dirt, is in the eye. The resultant damage to the eye surface increases discomfort and sensitivity to bright light. Both eyes usually are affected.

There may also be a stringy discharge from the eyes. Although it may seem strange, dry eye can cause the eyes to water. This can happen because the eyes are irritated. One may experience excessive tearing in the same way as one would if something got into the eye. These reflex tears will not necessarily make the eyes feel better. This is because they are the watery type that are produced in response to injury, irritation, or emotion. They do not have the lubricating qualities necessary to prevent dry eye.

Because blinking coats the eye with tears, symptoms are worsened by activities in which the rate of blinking is reduced due to prolonged use of the eyes. These activities include

prolonged reading, computer usage, driving, or watching television. Symptoms increase in windy, dusty or smoky (including cigarette smoke) areas, in dry environments, high altitudes including airplanes, on days with low humidity, and in areas where an air conditioner (especially in a car), fan, heater, or even a hair dryer is being used. Symptoms reduce during cool, rainy, or foggy weather and in humid places, such as in the shower.

Most people who have dry eyes experience mild irritation with no long-term effects. However, if the condition is left untreated or becomes severe, it can produce complications that can cause eye damage, resulting in impaired vision or (rarely) in the loss of vision.

Symptom assessment is a key component of dry eye diagnosis - to the extent that many believe dry eye syndrome to be a symptom-based disease. Several questionnaires have been developed to determine a score that would allow for dry eye diagnosis. The McMonnies & Ho dry eye questionnaire is often used in clinical studies of dry eyes. It has 14 questions, resulting in a score from 0 to 45. Scores above 14.5 are consistent with dry eyes.

Pathophysiology

Having dry eyes for a while can lead to tiny abrasions on the surface of the eyes. In advanced cases, the epithelium undergoes pathologic changes, namely squamous metaplasia and loss of goblet cells. Some severe cases result in thickening of the corneal surface, corneal erosion, punctate keratopathy, epithelial defects, corneal ulceration (sterile and infected), corneal neovascularization, corneal scarring, corneal thinning, and even corneal perforation.

Causes

Any abnormality of any one of the three layers of tears produces an unstable tear film, resulting in symptoms of keratitis sicca.

Deficient tear production

Keratoconjunctivitis sicca is usually due to inadequate tear production. The aqueous tear layer is affected, resulting in **aqueous tear deficiency** (**ATD**) or **lacrimal hyposecretion**. The lacrimal gland does not produce sufficient tears to keep the entire conjunctiva and cornea covered by a complete layer. This usually occurs in people who are otherwise healthy. Increased age is associated with decreased tearing. This is the most common type found in postmenopausal women.

Causes include idiopathic, congenital alacrima, xerophthalmia, lacrimal gland ablation, and sensory denervation. In rare cases, it may be a symptom of collagen vascular diseases, including rheumatoid arthritis, Wegener's granulomatosis, and systemic lupus erythematosus. Sjögren's syndrome and autoimmune diseases associated with Sjögren's syndrome are also conditions associated with aqueous tear deficiency. Drugs such as

isotretinoin, sedatives, diuretics, tricyclic antidepressants, antihypertensives, oral contraceptives, antihistamines, nasal decongestants, beta-blockers, phenothiazines, atropine, and pain relieving opiates such as morphine can cause or worsen this condition. Infiltration of the lacrimal glands by sarcoidosis or tumors, or postradiation fibrosis of the lacrimal glands can also cause this condition.

Abnormal tear composition

Keratoconjunctivitis sicca can also be caused by abnormal tear composition resulting in rapid evaporation or premature destruction of the tears. When caused by rapid evaporation, it is termed **evaporative dry eyes**. In this, although the tear gland produces a sufficient amount of tears, the rate of evaporation of the tears is too rapid. There is a loss of water from the tears that results in tears that are too "salty" or hypertonic. As a result, the entire conjunctiva and cornea cannot be kept covered with a complete layer of tears during certain activities or in certain environments.

Additional causes

Aging is one of the most common causes of dry eyes. This is because tear production decreases with age. It may be caused by thermal or chemical burns, or (in epidemic cases) by adenoviruses. A number of studies have found that diabetics are at increased risk for the disease.

About half of all people who wear contact lenses complain of dry eyes. There are two potential connections between contact usage and dry eye. Traditionally, it was believed that soft contact lenses, which float on the tear film that covers the cornea, absorb the tears in the eyes. However, it is also now known that contact usage damages corneal nerve sensitivity, which subsequently may lead to decreased lacrimal gland tear production and dry eye. The effect of contact on corneal nerve sensitivity is well established for hard contacts as well as soft and rigid gas permeable. The connection between this loss in nerve sensitivity and tear production is the subject of current research.

Dry eyes also occurs or gets worse after LASIK and other refractive surgeries, in which the corneal nerves are cut during the creation of a corneal flap. The corneal nerves stimulate tear secretion. Dry eyes caused by these procedures usually resolves after several months. Persons who are thinking about refractive surgery should consider this.

An eye injury or other problem with the eyes or eyelids, such as bulging eyes or a drooping eyelid can cause keratoconjunctivitis sicca. Disorders of the eyelid can impair the complex blinking motion required to spread tears.

Abnormalities of the lipid tear layer caused by blepharitis and rosacea, and abnormalities of the mucin tear layer caused by vitamin A deficiency, trachoma, diphtheric keratoconjunctivitis, mucocutaneous disorders and certain topical medications are causes of keratoconjunctivitis sicca.

Persons with keratoconjunctivitis sicca have elevated levels of tear nerve growth factor (NGF). It is possible that this ocular surface NGF plays an important role in ocular surface inflammation associated with dry eyes.

Diagnosis

Dry eyes can usually be diagnosed by the symptoms alone. Tests can determine both the quantity and the quality of the tears. A slit lamp examination can be performed to diagnose dry eyes and to document any damage to the eye.

A Schirmer's test can measure the amount of moisture bathing the eye. This test is useful for determining the severity of the condition. A five-minute Schirmer's test with and without anesthesia using a Whatman #41 filter paper 5 mm wide by 35 mm long is performed. For this test, wetting under 5 mm with or without anesthesia is considered diagnostic for dry eyes.

If the results for the Schirmer's test are abnormal, a Schirmer II test can be performed to measure reflex secretion. In this test, the nasal mucosa is irritated with a cotton-tipped applicator, after which tear production is measured with a Whatman #41 filter paper. For this test, wetting under 15 mm after five minutes is considered abnormal.

A tear breakup time (TBUT) test measures the time it takes for tears to break up in the eye. The tear breakup time can be determined after placing a drop of fluorescein in the cul-de-sac.

A tear protein analysis test measures the lysozyme contained within tears. In tears, lysozyme accounts for approximately 20 to 40 percent of total protein content.

A lactoferrin analysis test provides good correlation with other tests.

The presence of the recently described molecule Ap4A, naturally occurring in tears, is abnormally high in different states of ocular dryness. This molecule can be quantified biochemically simply by taking a tear sample with a plain Schirmer test. Utilizing this technique it is possible to determine the concentrations of Ap4A in the tears of patients and in such way diagnose objectively if the samples are indicative of dry eye.

Treatment

A variety of approaches can be taken to treatment. These can be summarised as: avoidance of exacerbating factors, tear stimulation and supplementation, increasing tear retention, and eyelid cleansing and treatment of eye inflammation.

General measures

Dry eyes can be exacerbated by smokey environments, dust and air conditioning and by our natural tendency to reduce our blink rate when concentrating. Purposefully blinking,

especially during computer use and resting tired eyes are basic steps that can be taken to minimise discomfort. Rubbing one's eyes can irritate them further, so should be avoided. Conditions such as blepharitis can often co-exist and paying particular attention to cleaning the eyelids morning and night with mild shampoos and warm compresses can improve both conditions.

Environmental control

Dry, drafty environments and those with smoke and dust should be avoided. This includes avoiding hair dryers, heaters, air conditioners or fans, especially when these devices are directed toward the eyes. Wearing glasses or directing gaze downward, for example, by lowering computer screens can be helpful to protect the eyes when aggravating environmental factors cannot be avoided. Using a humidifier, especially in the winter, can help by adding moisture to the dry indoor air.

Rehydration

For mild and moderate cases, supplemental lubrication is the most important part of treatment.

Artificial tears

Application of artificial tears every few hours can provide temporary relief.

Autologous serum eye drops

None of the commercially available artificial tear preparations include essential tear components such as epidermal growth factor, hepatocyte growth factor, fibronectin, neurotrophic growth factor, and vitamin A-all of which have been shown to play important roles in the maintenance of a healthy ocular surface epithelial milieu. Autologous serum eye drops contain these essential factors. However, there is some controversy regarding the efficacy of this treatment. At least one study (PubMed) has demonstrated that this modality is more effective than artificial tears in a randomized control study.

Additional options

Lubricating tear ointments can be used during the day, but they generally are used at bedtime due to poor vision after application. They contain white petrolatum, mineral oil, and similar lubricants. They serve as a lubricant and an emollient. Application requires pulling down the eyelid and applying a small amount (0.25 in) inside. Depending on the severity of the condition, it may be applied from every hour to just at bedtime. It should not be used with contact lenses. Specially designed glasses that form a moisture chamber around the eye may be used to create additional humidity.

Medication

Inflammation occurring in response to tears film hypertonicity can be suppressed by mild topical steroids or with topical immunosuppressants such as ciclosporin. Elevated levels of tear NGF can be decreased with 0.1% prednisolone.

Fish consumption and omega-3 fatty acids

Consumption of dark fleshed fish containing dietary omega-3 fatty acids is associated with a decreased incidence of dry eyes syndrome in women. This finding is consistent with postulated biological mechanisms. Early experimental work on omega-3 has shown promising results when used in a topical application or given orally.

Restasis

Topical ciclosporin (topical cyclosporin A, tCSA) 0.05% ophthalmic emulsion is an immunosuppressant, marketed in the United States by Allergan under the trade name Restasis. Approved as a prescription product by the U.S. Food and Drug Administration in 2002, the drug decreases surface inflammation. It is thought to work through inhibition of transcription factors required for cytokine production and T-lymphocyte maturation. In a trial involving 1200 people, Restasis increased tear production in 15% of people, compared to 5% with placebo.

Usually, 1 gtt (drop) of Restasis is instilled in each eye twice a day, 12 hours apart. It should not be used while wearing contact lenses, during eye infections or in people with a history of herpes virus infections. Side effects include burning sensation (common), redness, discharge, watery eyes, eye pain, foreign body sensation, itching, stinging, and blurred vision. Long term use of ciclosporin at high doses is associated with an increased risk of cancer.

Generic alternatives

Cheaper generic alternatives to Restasis are available in some countries. In India, it is marketed as Cyclomune by Sun Pharma.

Conserving tears

There are methods that allow both natural and artificial tears to stay longer.

Blocking tear drainage

In each eye, there are two puncta — little openings that drain tears into the tear ducts. There are methods to partially or completely close the tear ducts. This blocks the flow of tears into the nose, and thus more tears are available to the eyes.

Punctal plugs

Punctal plugs are inserted into the puncta to block tear drainage. For people who have not found dry eye relief with drugs, punctal plugs may help. They are reserved for people with moderate or severe dry eye when other medical treatment has not been adequate.

Cauterization

If punctal plugs are effective, thermal or electric cauterization of puncti can be performed.

In thermal cauterization, a local anesthetic is used, and then a hot wire is applied. This shrinks the drainage area tissues and causes scarring, which closes the tear duct.

Customized contact lenses

Persons with severe dry eyes may benefit from the Boston Scleral Lens which is a customized contact lens. Resting on the sclera, it creates a fluid filled layer over the cornea, thus preventing it from drying.

Surgery

In severe cases of keratoconjunctivitis sicca, tarsorrhaphy may be performed where the eyelids are partially sewn together. This reduces the palpebral fissure (eyelid separation), ideally leading to a reduction in tear evaporation.

Prognosis

Keratoconjunctivitis sicca usually is a chronic problem. Its prognosis shows considerable variance, depending upon the severity of the condition. Most patients have mild-to-moderate cases, and can be treated symptomatically with lubricants. This provides an adequate relief of symptoms.

When dry eyes symptoms are severe, they can interfere with quality of life. People sometimes feel their vision blurs with use, or severe irritation to the point that they have trouble keeping their eyes open or they may not be able to work or drive.

Prevention

There is no way to prevent keratoconjunctivitis sicca. Complications can be prevented by use of wetting and lubricating drops and ointments.

Epidemiology

Keratoconjunctivitis sicca is relatively common within the United States, especially so in older patients. Specifically, the persons most likely to be affected by dry eyes are those aged 40 or older.

While persons with autoimmune diseases have a high likelihood of having dry eyes, most persons with dry eyes do not have an autoimmune disease. Instances of Sjögren syndrome and keratoconjunctivitis sicca associated with it are present much more commonly in women, with a ratio of 9:1. In addition, milder forms of keratoconjunctivitis sicca also are more common in women. This is partly because hormonal changes, such as those that occur in pregnancy, menstruation, and menopause, can decrease tear production.

In areas of the world where malnutrition is common, vitamin A deficiency is a common cause. This is rare in the United States.

Racial predilections do not exist for this disease.

Occurrence in animals

Among animals, keratoconjunctivitis sicca occurs in dogs, cats, and horses.

Dogs

Keratoconjunctivitis sicca is common in dogs. Most cases are caused by a genetic predisposition, but chronic conjunctivitis, canine distemper, and drugs such as sulfasalazine and trimethoprim-sulfonamide also cause the disease. Symptoms include eye redness, a yellow or greenish discharge, ulceration of the cornea, pigmented cornea, and blood vessels on the cornea. Diagnosis is made by measuring tear production with a Schirmer tear test. Less than 15 millimeters of tears produced in a minute is abnormal.

Tear replacers are a mainstay of treatment, preferably containing methylcellulose or carboxymethyl cellulose. Ciclosporin stimulates tear production and acts as a suppressant on the immune-mediated processes that cause the disease. Topical antibiotics and corticosteroids are sometimes used to treat secondary infections and inflammation. A surgery known as parotid duct transposition is used in some extreme cases where medical treatment has not helped. This redirects the duct from the parotid salivary gland to the eye. Saliva replaces the tears. Dogs suffering from cherry eye should have the condition corrected to help prevent this disease.

Commonly affected breeds include:

- Cavalier King Charles Spaniel
- Bulldog
- Chinese Shar-Pei

- Lhasa Apso
- Shih Tzu
- West Highland White Terrier
- Pug
- Bloodhound
- Cocker Spaniel
- Pekingese
- Boston Terrier
- Miniature Schnauzer
- Samoyed

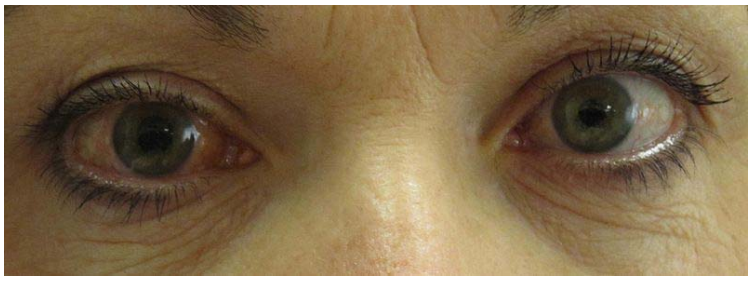
Cats

Keratoconjunctivitis sicca is uncommon in cats. Most cases seem to be caused by chronic conjunctivitis, especially secondary to feline herpesvirus. Diagnosis, symptoms, and treatment are similar to those for dogs.

Chapter 19

Glaucoma

Glaucoma



Acute angle closure glaucoma of the right eye. Note the mid sized pupil, which was non-reactive to light, and injection of the conjunctiva.

ICD-10	H40.-H42.
ICD-9	365
DiseasesDB	5226
eMedicine	oph/578
MeSH	D005901

Glaucoma is an eye disorder in which the optic nerve suffers damage, permanently impacting vision in the affected eye(s) and progressing to complete blindness if untreated. It is often, but not always, associated with increased pressure of the fluid in the eye (aqueous humour).

The nerve damage involves loss of retinal ganglion cells in a characteristic pattern. There are many different sub-types of glaucoma but they can all be considered a type of optic neuropathy. Raised intraocular pressure is a significant risk factor for developing glaucoma (above 21 mmHg or 2.8 kPa). One person may develop nerve damage at a relatively low pressure, while another person may have high eye pressure for years and yet never develop damage. Untreated glaucoma leads to permanent damage of the optic nerve and resultant visual field loss, which can progress to blindness.

Glaucoma can be divided roughly into two main categories, "open angle" and "closed angle" glaucoma. Closed angle glaucoma can appear suddenly and is often painful; visual loss can progress quickly but the discomfort often leads patients to seek medical attention before permanent damage occurs. Open angle, chronic glaucoma tends to progress at a slower rate and the patient may not notice that they have lost vision until the disease has progressed significantly.

Glaucoma has been nicknamed the "silent thief of sight" because the loss of vision normally occurs gradually over a long period of time and is often only recognized when the disease is quite advanced. Once lost, this damaged visual field cannot be recovered. Worldwide, it is the second leading cause of blindness. It is also the leading cause of blindness among African Americans. Glaucoma affects 1 in 200 people aged fifty and younger, and 1 in 10 over the age of eighty. If the condition is detected early enough it is possible to arrest the development or slow the progression with medical and surgical means.

The word *glaucoma* comes from the Greek γλαύκωμα, "opacity of the crystalline lens".

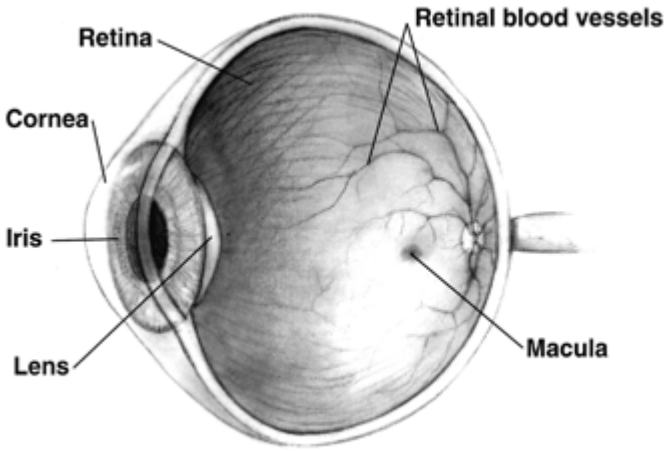
Signs and symptoms

There are two main types of glaucoma: **open-angle glaucoma** and **closed-angle glaucoma.**

Open-angle glaucoma accounts for 90% of glaucoma cases in the United States. It is painless and does not have acute attacks. The only signs are gradually progressive visual field loss, and optic nerve changes (increased cup-to-disc ratio on fundoscopic examination).

Closed-angle glaucoma accounts for less than 10% of glaucoma cases in the United States, but as much as half of glaucoma cases in other nations (particularly Asian countries). About 10% of patients with closed angles present with acute angle closure crises characterized by sudden ocular pain, seeing halos around lights, red eye, very high intraocular pressure (>30 mmHg), nausea and vomiting, sudden decreased vision, and a fixed, mid-dilated pupil. Acute angle closure is an ocular emergency.

Pathophysiology



Human eye cross-sectional view

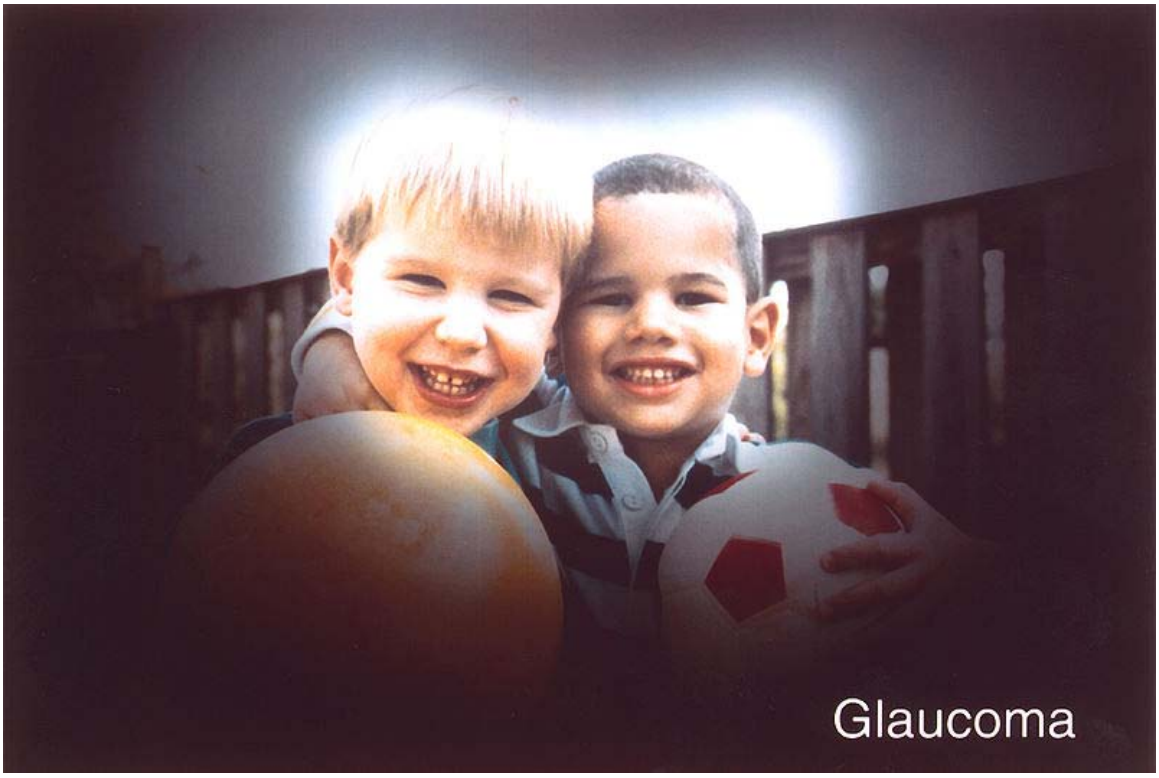
The major risk factor for most glaucomas and focus of treatment is increased intraocular pressure. Intraocular pressure is a function of production of liquid aqueous humor by the ciliary processes of the eye and its drainage through the trabecular meshwork. Aqueous humor flows from the ciliary processes into the posterior chamber, bounded posteriorly by the lens and the zonules of Zinn and anteriorly by the iris. It then flows through the pupil of the iris into the anterior chamber, bounded posteriorly by the iris and anteriorly by the cornea. From here the trabecular meshwork drains aqueous humor via Schlemm's canal into scleral plexuses and general blood circulation. In open angle glaucoma there is reduced flow through the trabecular meshwork; in angle closure glaucoma, the iris is apposed to the lens resulting in the inability of the aqueous fluid to flow from the posterior to the anterior chamber and then out of the trebecular network.

The inconsistent relationship of glaucomatous optic neuropathy with ocular hypertension has provoked hypotheses and studies on anatomic structure, eye development, nerve compression trauma, optic nerve blood flow, excitatory neurotransmitter, trophic factor, retinal ganglion cell/axon degeneration, glial support cell, immune, and aging mechanisms of neuron loss.

The major types of glaucoma are discussed below.

Causes and risk factors

A normal range of vision



The same view with advanced vision loss from glaucoma

There are several causes for glaucoma. *Those at risk are advised to have a dilated eye examination at least once a year.*

Ocular hypertension (increased pressure within the eye) is the largest risk factor in most glaucomas, but in some populations only 50% of patients with primary open angle glaucoma actually have elevated ocular pressure.

Those of African descent are three times more likely to develop primary open angle glaucoma.

Elderly people have thinner corneal thickness and often suffer from hypermetropia. They are also at higher risk for primary open angle glaucoma.

People with a family history of glaucoma have about six percent chance of developing glaucoma.

Many East Asian groups are prone to developing angle closure glaucoma due to their shallower anterior chamber depth, with the majority of cases of glaucoma in this population consisting of some form of angle closure. Inuit also have a twenty to forty times higher risk than Caucasians of developing primary angle closure glaucoma. Women are three times more likely than men to develop acute angle-closure glaucoma due to their shallower anterior chambers.

Other factors can cause glaucoma, known as "secondary glaucomas," including prolonged use of steroids (steroid-induced glaucoma); conditions that severely restrict blood flow to the eye, such as severe diabetic retinopathy and central retinal vein occlusion (neovascular glaucoma); ocular trauma (angle recession glaucoma); and uveitis (uveitic glaucoma).

Primary open angle glaucoma (POAG) has been found to be associated with mutations in genes at several loci. Normal tension glaucoma, which comprises one third of POAG, is associated with genetic mutations.

There is increasing evidence that ocular blood flow is involved in the pathogenesis of glaucoma. Current data indicate that fluctuations in blood flow are more harmful in glaucomatous optic neuropathy than steady reductions. Unstable blood pressure and dips are linked to optic nerve head damage and correlate with visual field deterioration.

A number of studies also suggest a possible correlation between hypertension and the development of glaucoma. In normal tension glaucoma, nocturnal hypotension may play a significant role.

There is no clear evidence that vitamin deficiencies cause glaucoma in humans. It follows then that oral vitamin supplementation is not a recommended treatment for glaucoma.

Various rare congenital/genetic eye malformations are associated with glaucoma. Occasionally, failure of the normal third trimester gestational atrophy of the hyaloid canal and the tunica vasculosa lentis is associated with other anomalies. Angle closure induced ocular hypertension and glaucomatous optic neuropathy may also occur with these anomalies and modelled in mice.

Diagnosis

Screening for glaucoma is usually performed as part of a standard eye examination performed by ophthalmologists, orthoptists and optometrists. Testing for glaucoma should include measurements of the intraocular pressure via tonometry, changes in size or shape of the eye, anterior chamber angle examination or gonioscopy, and examination of the optic nerve to look for any visible damage to it, or change in the cup-to-disc ratio and also rim appearance and vascular change. A formal visual field test should be performed. The retinal nerve fiber layer can be assessed with imaging techniques such as optical coherence tomography (OCT), scanning laser polarimetry (GDx), and/or scanning laser ophthalmoscopy also known as Heidelberg Retina Tomography (HRT3). Owing to the sensitivity of all methods of tonometry to corneal thickness, methods such as Goldmann tonometry should be augmented with pachymetry to measure central corneal thickness (CCT). A thicker-than-average cornea can result in a pressure reading higher than the 'true' pressure, whereas a thinner-than-average cornea can produce a pressure reading lower than the 'true' pressure. Because pressure measurement error can be caused by more than just CCT (i.e., corneal hydration, elastic properties, etc.), it is impossible to 'adjust' pressure measurements based only on CCT measurements. The Frequency

Doubling Illusion can also be used to detect glaucoma with the use of a Frequency Doubling Technology (FDT) perimeter. Examination for glaucoma also could be assessed with more attention given to sex, race, history of drug use, refraction, inheritance and family history.

Management

The modern goals of glaucoma management are to avoid glaucomatous damage, nerve damage, preserve visual field and total quality of life for patients with minimal side effects. This requires appropriate diagnostic techniques and follow up examinations and judicious selection of treatments for the individual patient. Although intraocular pressure is only one of the major risk factors for glaucoma, lowering it via various pharmaceuticals and/or surgical techniques is currently the mainstay of glaucoma treatment. Vascular flow and neurodegenerative theories of glaucomatous optic neuropathy have prompted studies on various neuroprotective therapeutic strategies including nutritional compounds some of which may be regarded by clinicians as safe for use now, while others are on trial.

Medication

Intraocular pressure can be lowered with medication, usually eye drops. There are several different classes of medications to treat glaucoma with several different medications in each class.

Each of these medicines may have local and systemic side effects. Adherence to medication protocol can be confusing and expensive; if side effects occur, the patient must be willing either to tolerate these, or to communicate with the treating physician to improve the drug regimen. Initially, glaucoma drops may reasonably be started in either one or in both eyes.

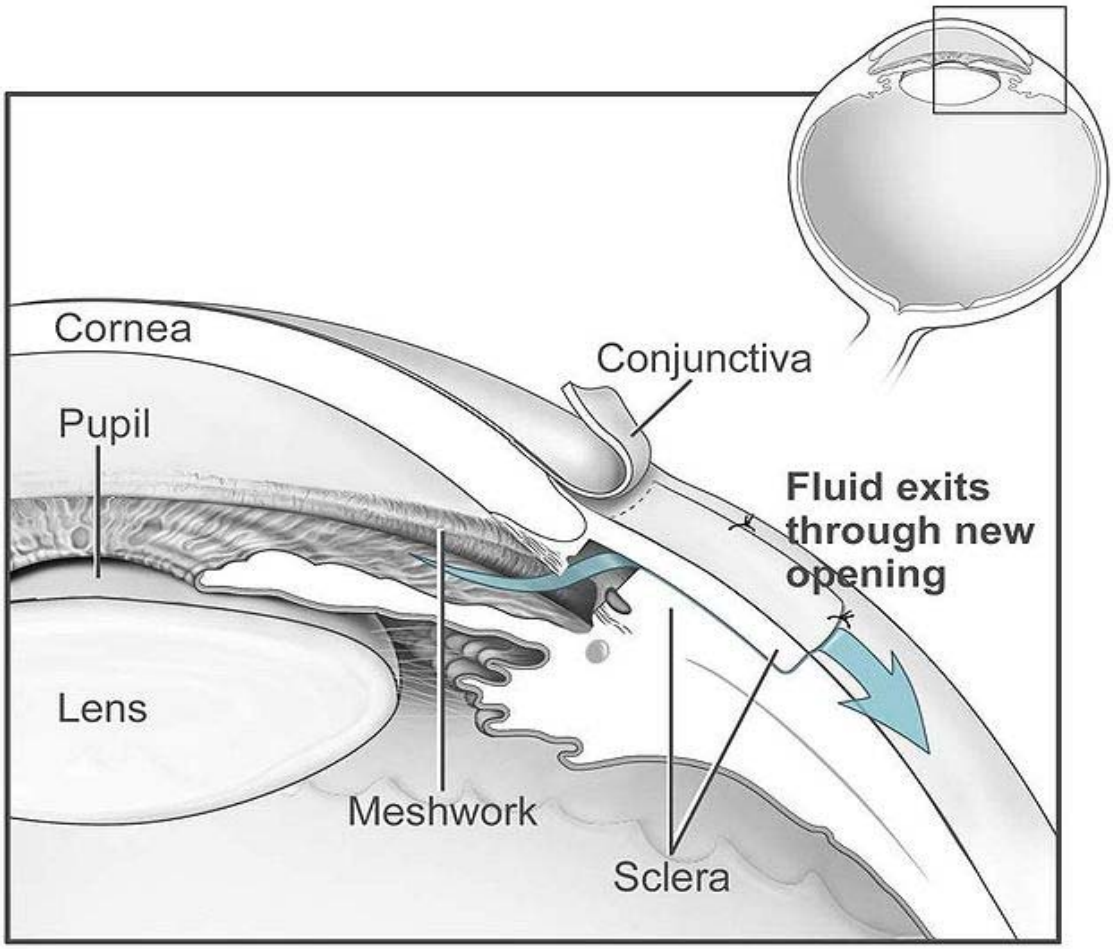
Poor compliance with medications and follow-up visits is a major reason for vision loss in glaucoma patients. A 2003 study of patients in an HMO found that half failed to fill their prescription the first time and one in four failed to refill their prescriptions a second time. Patient education and communication must be ongoing to sustain successful treatment plans for this lifelong disease with no early symptoms.

The possible neuroprotective effects of various topical and systemic medications are also being investigated.

- Prostaglandin analogs like latanoprost (Xalatan), bimatoprost (Lumigan) and travoprost (Travatan) increase uveoscleral outflow of aqueous humor. Bimatoprost also increases trabecular outflow
- Topical beta-adrenergic receptor antagonists such as timolol, levobunolol (Betagan), and betaxolol decrease aqueous humor production by the ciliary body.

- Alpha2-adrenergic agonists such as brimonidine (Alphagan) work by a dual mechanism, decreasing aqueous humor production and increasing trabecular outflow.
- Less-selective sympathomimetics such as epinephrine decrease aqueous humor production through vasoconstriction of ciliary body blood vessels.
- Miotic agents (parasympathomimetics) like pilocarpine work by contraction of the ciliary muscle, tightening the trabecular meshwork and allowing increased outflow of the aqueous humour. Ecothiopate is used in chronic glaucoma.
- Carbonic anhydrase inhibitors like dorzolamide (Trusopt), brinzolamide (Azopt), acetazolamide (Diamox) lower secretion of aqueous humor by inhibiting carbonic anhydrase in the ciliary body.
- Physostigmine is also used to treat glaucoma and delayed gastric emptying.

Surgery



Conventional surgery to treat glaucoma makes a new opening in the meshwork. This new opening helps fluid to leave the eye and lowers intraocular pressure.

Both laser surgeries and conventional surgeries are performed to treat glaucoma.

Surgery is the primary therapy for those with congenital glaucoma.

Generally, these operations are a temporary solution, as there is not yet a cure for glaucoma.

Canaloplasty

Canaloplasty is a nonpenetrating procedure utilizing microcatheter technology. To perform a canaloplasty, an incision is made into the eye to gain access to Schlemm's canal in a similar fashion to a viscocanalostomy. A microcatheter will circumnavigate the canal around the iris, enlarging the main drainage channel and its smaller collector channels through the injection of a sterile, gel-like material called viscoelastic. The catheter is then removed and a suture is placed within the canal and tightened. By opening the canal, the pressure inside the eye may be relieved, although the reason is unclear since the canal (of Schlemm) does not have any significant fluid resistance in glaucoma or healthy eyes. Long-term results are not available.

Laser surgery

Laser trabeculoplasty may be used to treat open angle glaucoma. It is a temporary solution, not a cure. A 50 μm argon laser spot is aimed at the trabecular meshwork to stimulate opening of the mesh to allow more outflow of aqueous fluid. Usually, half of the angle is treated at a time. Traditional laser trabeculoplasty utilizes a thermal argon laser. The procedure is called Argon Laser Trabeculoplasty or ALT. A newer type of laser trabeculoplasty exists that uses a "cold" (non-thermal) laser to stimulate drainage in the trabecular meshwork. This newer procedure which uses a 532 nm frequency-doubled, Q-switched Nd:YAG laser which selectively targets melanin pigment in the trabecular meshwork cells, called Selective Laser Trabeculoplasty or SLT. Studies show that SLT is as effective as ALT at lowering eye pressure. In addition, SLT may be repeated three to four times, whereas ALT can usually be repeated only once.

Nd:YAG laser peripheral iridotomy (LPI) may be used in patients susceptible to or affected by angle closure glaucoma or pigment dispersion syndrome. During laser iridotomy, laser energy is used to make a small full-thickness opening in the iris. This opening equalizes the pressure between the front and back of the iris correcting any abnormal bulging of the iris. In people with narrow angles, this can uncover the trabecular meshwork. In some cases of intermittent or short-term angle closure this may lower the eye pressure. Laser iridotomy reduces the risk of developing an attack of acute angle closure. In most cases it also reduces the risk of developing chronic angle closure or of adhesions of the iris to the trabecular meshwork.

Diode laser cycloablation lowers IOP by reducing aqueous secretion by destroying secretory ciliary epithelium.

Trabeculectomy

The most common conventional surgery performed for glaucoma is the trabeculectomy. Here, a partial thickness flap is made in the scleral wall of the eye, and a window opening made under the flap to remove a portion of the trabecular meshwork. The scleral flap is then sutured loosely back in place. This allows fluid to flow out of the eye through this opening, resulting in lowered intraocular pressure and the formation of a bleb or fluid bubble on the surface of the eye. Scarring can occur around or over the flap opening, causing it to become less effective or lose effectiveness altogether.

Glaucoma drainage implants

There are also several different glaucoma drainage implants. These include the original Molteno implant (1966), the Baerveldt tube shunt, or the valved implants, such as the Ahmed glaucoma valve implant or the ExPress Mini Shunt and the later generation pressure ridge Molteno implants. These are indicated for glaucoma patients not responding to maximal medical therapy, with previous failed guarded filtering surgery (trabeculectomy). The flow tube is inserted into the anterior chamber of the eye and the plate is implanted underneath the conjunctiva to allow flow of aqueous fluid out of the eye into a chamber called a bleb.

- The first-generation Molteno and other non-valved implants sometimes require the ligation of the tube until the bleb formed is mildly fibrosed and water-tight This is done to reduce postoperative hypotony—sudden drops in postoperative intraocular pressure (IOP).
- Valved implants such as the Ahmed glaucoma valve attempt to control postoperative hypotony by using a mechanical valve.

The ongoing scarring over the conjunctival dissipation segment of the shunt may become too thick for the aqueous humor to filter through. This may require preventive measures using anti-fibrotic medication like 5-fluorouracil (5-FU) or mitomycin-C (during the procedure), or additional surgery. And for Glaucomatous painful Blind Eye and some cases of Glaucoma, Cyclocryotherapy for ciliary body ablation could be considered to be performed.

Veterinary implant

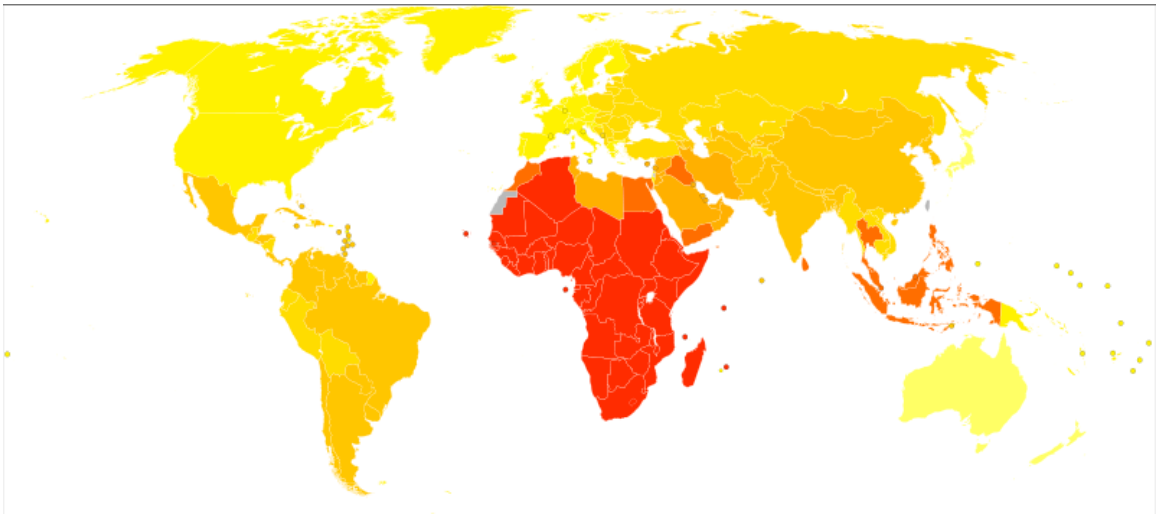
TR BioSurgical has commercialized a new implant specifically for veterinary medicine, called TR-ClarifEYE. The implant consists of a new biomaterial, the STAR BioMaterial, which consists of silicone with a very precise homogenous pore size, a property which reduces fibrosis and improves tissue integration. The implant contains no valves and is placed completely within the eye without sutures. To date, it has demonstrated long term success (> 1yr) in a pilot study in medically refractory dogs with advanced glaucoma

Laser assisted non-penetrating deep sclerectomy

The most common surgical approach currently used for the treatment of glaucoma, is trabeculectomy, in which the sclera is punctured to alleviate intraocular pressure (IOP), the pressure inside the eye. Non-penetrating deep sclerectomy (NPDS) surgery is a similar but modified procedure, in which instead of puncturing the scleral wall, a patch of the sclera is skimmed to a level, upon which, percolation of liquid from the inner eye is achieved and thus alleviating IOP, without penetrating the eye. NPDS is demonstrated to cause a significantly less side effects than trabeculectomy. However, NPDS is performed manually and requires great skill to achieve a lengthy learning curve.

Laser assisted NPDS is the performance of NPDS with the use of a CO_2 laser system. The laser-based system is self-terminating once the required scleral thickness and adequate drainage of the intra ocular fluid have been achieved. This self-regulation effect is achieved as the CO_2 laser essentially stops ablating as soon as it comes in contact with the intra-ocular percolated liquid, which occurs as soon as the laser reaches the optimal residual intact layer thickness.

Epidemiology



Disability-adjusted life year for glaucoma per 100,000 inhabitants in 2004.

- no data
- fewer than 20
- 20-43
- 43-66
- 66-89
- 89-112
- 112-135
- 135-158
- 158-181
- 181-204
- 204-227

227-250
more than 250

Research

- Advanced Glaucoma Intervention Study (AGIS) - large American National Eye Institute (NEI) sponsored study designed "to assess the long-range outcomes of sequences of interventions involving trabeculectomy and argon laser trabeculoplasty in eyes that have failed initial medical treatment for glaucoma." It recommends different treatments based on race.
- Early Manifest Glaucoma Trial (EMGT) -Another NEI study found that immediately treating people who have early stage glaucoma can delay progression of the disease.
- Ocular Hypertension Treatment Study (OHTS) -NEI study findings: "...Topical ocular hypotensive medication was effective in delaying or preventing onset of Primary Open Angle Glaucoma (POAG) in individuals with elevated Intraocular Pressure (IOP). Although this does not imply that all patients with borderline or elevated IOP should receive medication, clinicians should consider initiating treatment for individuals with ocular hypertension who are at moderate or high risk for developing POAG."
- Blue Mountains Eye Study "The Blue Mountains Eye Study was the first large population-based assessment of visual impairment and common eye diseases of a representative older Australian community sample." Risk factors for glaucoma and other eye disease were determined.

Compounds in research

Natural compounds of research interest in glaucoma prevention or treatment include: fish oil and omega 3 fatty acids, bilberries, vitamin E, cannabinoids, carnitine, coenzyme Q10, curcurmin, Salvia miltiorrhiza, dark chocolate, erythropoietin, folic acid, Ginkgo biloba, Ginseng, L-glutathione, grape seed extract, green tea, magnesium, melatonin, methylcobalamin, N-acetyl-L cysteine, pycnogenols, resveratrol, quercetin and salt. Magnesium, ginkgo, salt and fludrocortisone, are already used by some physicians.

Cannabis

Studies in the 1970s showed that marijuana, when smoked, effectively lowers intraocular pressure. In an effort to determine whether marijuana, or drugs derived from marijuana, might be effective as a glaucoma treatment, the US National Eye Institute supported research studies from 1978 to 1984. These studies demonstrated that some derivatives of marijuana lowered intraocular pressure when administered orally, intravenously, or by smoking, but not when topically applied to the eye.

In 2003 the American Academy of Ophthalmology released a position statement which said that "studies demonstrated that some derivatives of marijuana did result in lowering of IOP when administered orally, intravenously, or by smoking, but not when topically

applied to the eye. The duration of the pressure-lowering effect is reported to be in the range of 3 to 4 hours".

However, the position paper qualified that by stating that marijuana was not more effective than prescription medications, stating that "no scientific evidence has been found that demonstrates increased benefits and/or diminished risks of marijuana use to treat glaucoma compared with the wide variety of pharmaceutical agents now available."

The first patient in the United States federal government's Compassionate Investigational New Drug program, Robert Randall, was afflicted with glaucoma and had successfully fought charges of marijuana cultivation because it was deemed a medical necessity (*U.S. v. Randall*) in 1976.

5-HT_{2A} agonists

Peripherally selective 5-HT_{2A} agonists such as the indazole derivative AL-34662 are currently under development and show significant promise in the treatment of glaucoma.

Classification

Glaucoma has been classified into specific types:

Primary glaucoma and its variants (H40.1-H40.2)

- Primary glaucoma
    - Primary angle-closure glaucoma, also known as primary closed-angle glaucoma, narrow-angle glaucoma, pupil-block glaucoma, acute congestive glaucoma
    - Acute angle-closure glaucoma
    - Chronic angle-closure glaucoma
    - Intermittent angle-closure glaucoma
    - Superimposed on chronic open-angle closure glaucoma ("combined mechanism" - uncommon)
    - Primary open-angle glaucoma, also known as chronic open-angle glaucoma, chronic simple glaucoma, glaucoma simplex
    - High-tension glaucoma
    - Low-tension glaucoma
- Variants of primary glaucoma
    - Pigmentary glaucoma

- Exfoliation glaucoma, also known as pseudoexfoliative glaucoma or glaucoma capsulare

Primary angle-closure glaucoma – This is caused by contact between the iris and trabecular meshwork, which in turn obstructs outflow of the aqueous humor from the eye. This contact between iris and trabecular meshwork (TM) may gradually damage the function of the meshwork until it fails to keep pace with aqueous production, and the pressure rises. In over half of all cases, prolonged contact between iris and TM causes the formation of synechiae (effectively "scars"). These cause permanent obstruction of aqueous outflow. In some cases, pressure may rapidly build up in the eye causing pain and redness (symptomatic, or so called "acute" angle-closure). In this situation the vision may become blurred, and halos may be seen around bright lights. Accompanying symptoms may include headache and vomiting. Diagnosis is made from physical signs and symptoms: pupils mid-dilated and unresponsive to light, cornea edematous (cloudy), reduced vision, redness, pain. However, the majority of cases are asymptomatic. Prior to very severe loss of vision, these cases can only be identified by examination, generally by an eye care professional. Once any symptoms have been controlled, the first line (and often definitive) treatment is laser iridotomy. This may be performed using either Nd:YAG or argon lasers, or in some cases by conventional incisional surgery. The goal of treatment is to reverse, and prevent, contact between iris and trabecular meshwork. In early to moderately advanced cases, iridotomy is successful in opening the angle in around 75% of cases. In the other 25% laser iridoplasty, medication (pilocarpine) or incisional surgery may be required.

Primary open-angle glaucoma – Optic nerve damage resulting in progressive visual field loss. This is associated with increased pressure in the eye. Not all people with primary open-angle glaucoma have eye pressure that is elevated beyond normal, but decreasing the eye pressure further has been shown to stop progression even in these cases. The increased pressure is caused by trabecular blockage which is where the aqueous humor in the eye drains out. Because the microscopic passage ways are blocked, the pressure builds up in the eye and causes imperceptible very gradual vision loss. Peripheral vision is affected first but eventually the entire vision will be lost if not treated. Diagnosis is made by looking for cupping of the optic nerve. Prostaglandin agonists work by opening uveoscleral passageways. Beta blockers such as timolol, work by decreasing aqueous formation. Carbonic anhydrase inhibitors decrease bicarbonate formation from ciliary processes in the eye, thus decreasing formation of Aqueous humor. Parasympathetic analogs are drugs that work on the trabecular outflow by opening up the passageway and constricting the pupil. Alpha 2 agonists (brimonidine, apraclonidine) both decrease fluid production (via. inhibition of AC) and increase drainage.

Developmental glaucoma (Q15.0)

- Developmental glaucoma
    - Primary congenital glaucoma
    - Infantile glaucoma

- Glaucoma associated with hereditary of familial diseases

Secondary glaucoma (H40.3-H40.6)

- Secondary glaucoma
    - Inflammatory glaucoma
    - Uveitis of all types
    - Fuchs heterochromic iridocyclitis
    - Phacogenic glaucoma
    - Angle-closure glaucoma with mature cataract
    - Phacoanaphylactic glaucoma secondary to rupture of lens capsule
    - Phacolytic glaucoma due to phacotoxic meshwork blockage
    - Subluxation of lens
    - Glaucoma secondary to intraocular hemorrhage
    - Hyphema
    - Hemolytic glaucoma, also known as erythroclastic glaucoma
    - Traumatic glaucoma
    - Angle recession glaucoma: Traumatic recession on anterior chamber angle
    - Postsurgical glaucoma
    - Aphakic pupillary block
    - Ciliary block glaucoma
    - Neovascular glaucoma
    - Drug-induced glaucoma
    - Corticosteroid induced glaucoma
    - Alpha-chymotrypsin glaucoma. Postoperative ocular hypertension from use of alpha chymotrypsin.
    - Glaucoma of miscellaneous origin
    - Associated with intraocular tumors
    - Associated with retinal detachments
    - Secondary to severe chemical burns of the eye
    - Associated with essential iris atrophy
    - Toxic Glaucoma

Neovascular glaucoma is an uncommon type of glaucoma that is difficult or nearly impossible to treat. This condition is often caused by proliferative diabetic retinopathy (PDR) or central retinal vein occlusion (CRVO). It may also be triggered by other conditions that result in ischemia of the retina or ciliary body. Individuals with poor blood flow to the eye are highly at risk for this condition.

Neovascular glaucoma results when new, abnormal vessels begin developing in the angle of the eye that begin blocking the drainage. Patients with such condition begin to rapidly lose their eyesight. Sometimes, the disease appears very rapidly, specially after cataract surgery procedure. A new treatment for this disease, as first reported by Kahook and colleagues, involves use of a novel group of medications known as Anti-VEGF agents. These injectable medications can lead to a dramatic decrease in new vessel formation and, if injected early enough in the disease process, may lead to normalization of intraocular pressure.

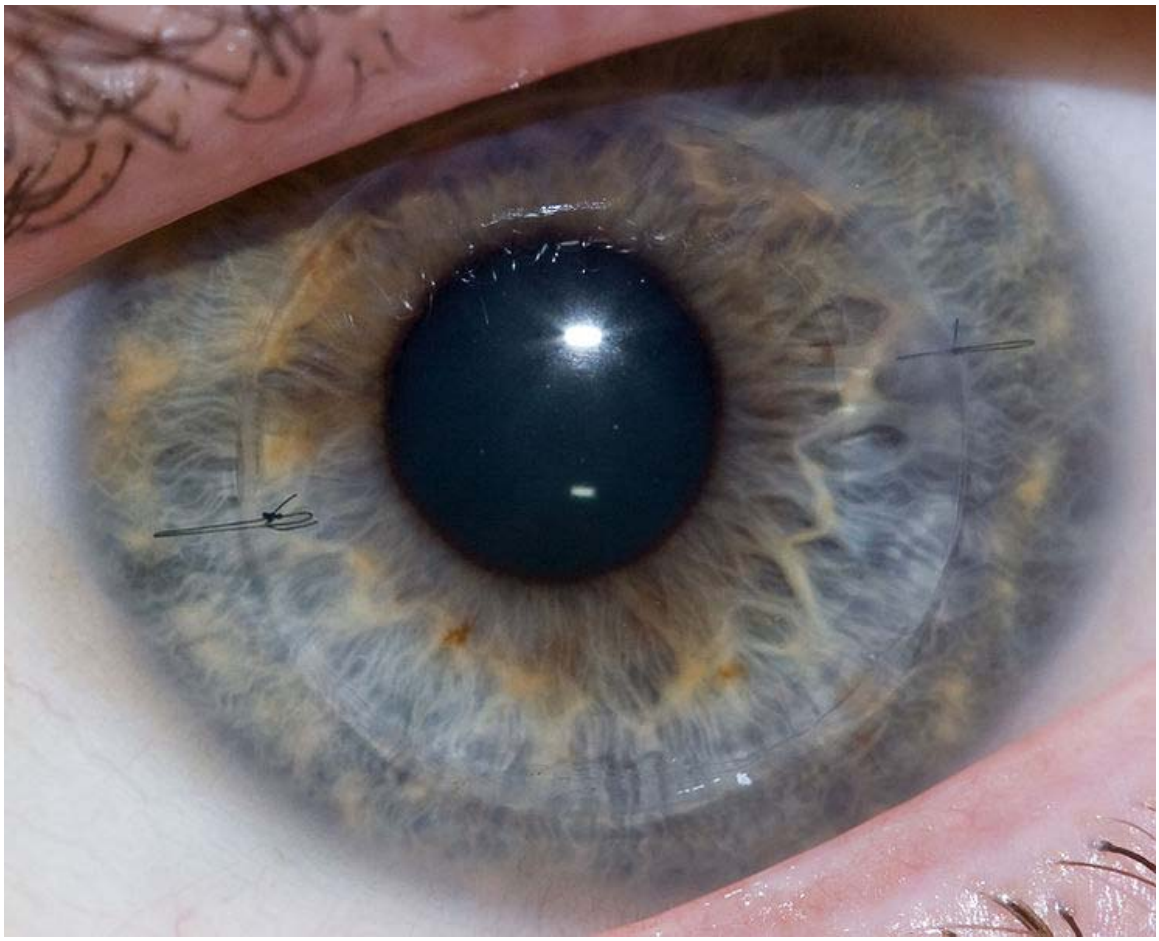
Toxic glaucoma is open angle glaucoma with an unexplained significant rise of intraocular pressure following unknown pathogenesis. Intraocular pressure can sometimes reach 80 mmHg (11 kPa). It characteristically manifests as ciliary body inflammation and massive trabecular oedema that sometimes extends to Schlemm's Canal. This condition is differentiated from malignant glaucoma by the presence of a deep and clear anterior chamber and a lack of aqueous misdirection. Also, the corneal appearance is not as hazy. A reduction in visual acuity can occur followed neuroretinal breakdown. Associated factors include inflammation, drugs, trauma and intraocular surgery, including cataract surgery and vitrectomy procedures. Gede Pardianto (2005) reports on four patients who had toxic glaucoma. One of them underwent phaecoemulsification with small particle nucleus drops. Some cases can be resolved with some medication, vitrectomy procedures or trabeculectomy. Valving procedures can give some relief but further research is required.

Absolute glaucoma (H44.5)

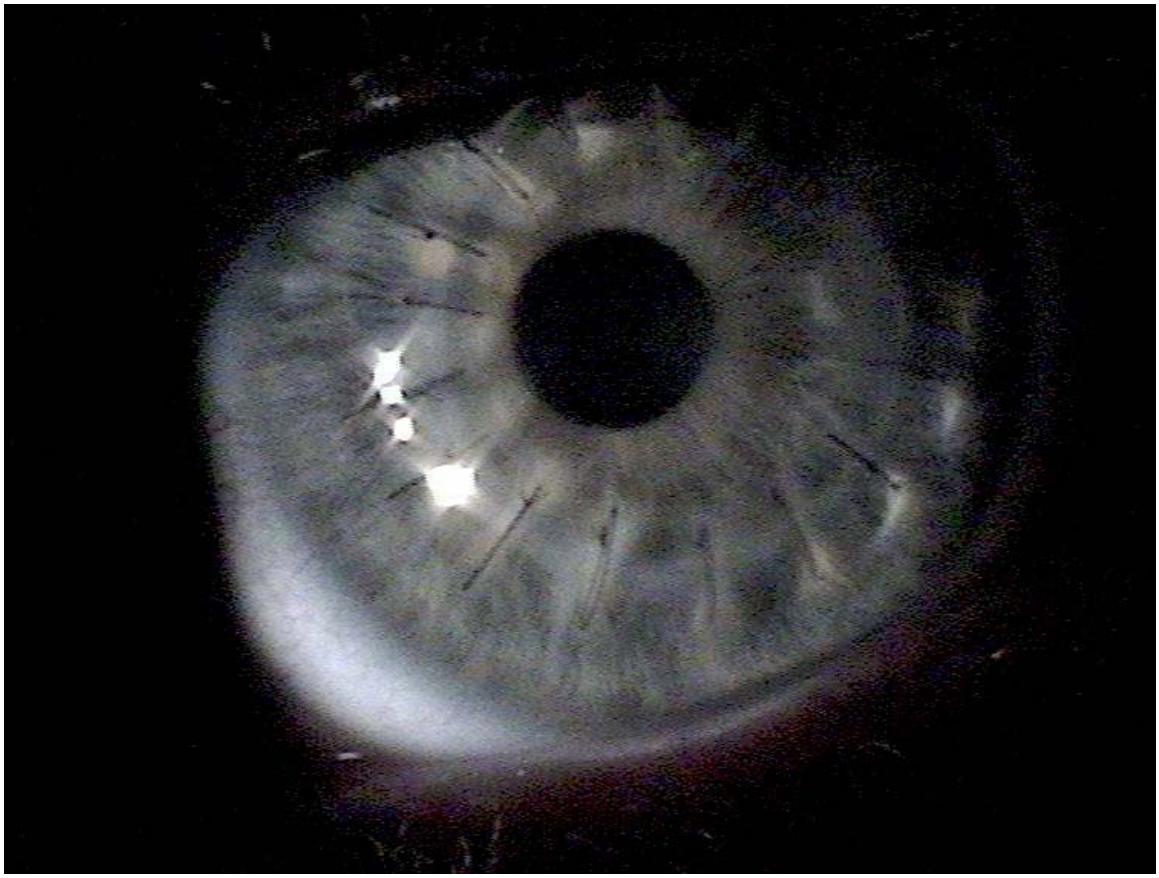
- Absolute glaucoma is the end stage of all types of glaucoma. The eye has no vision, absence of PL and PR, and has a stony appearance. Severe pain is present in the eye. The treatment of absolute glaucoma is a destructive procedure like cyclocryo application, cyclophotocoagulation, or injection of 100% alcohol.

Chapter 20

Corneal Transplantation



Cornea transplant after one year of healing, two stitches are visible



Cornea transplant approximately one week after surgery. Multiple light reflections indicate folds in the cornea, which later resolved.

Corneal transplantation, also known as **corneal grafting**, is a surgical procedure where a damaged or diseased cornea is replaced by donated corneal tissue (the graft) in its entirety (penetrating keratoplasty) or in part (lamellar keratoplasty). The graft has been removed from a recently deceased individual with no known diseases or other factors that may affect the viability of the donated tissue or the health of the recipient. The cornea is the transparent front part of the eye that covers the iris, pupil and anterior chamber. The surgical procedure is performed by ophthalmologists, medical doctors who specialize in eyes, and is often done on an outpatient basis.

Indications

Indications for corneal transplantation include the following:

- Optical: To improve visual acuity by replacing the opaque or distorted host tissue by clear healthy donor tissue. The most common indication in this category is pseudophakic bullous keratopathy, followed by keratoconus, corneal degeneration, keratoglobus and dystrophy, as well as scarring due to keratitis and trauma.

- Tectonic/reconstructive: To preserve corneal anatomy and integrity in patients with stromal thinning and descemetoceles, or to reconstruct the anatomy of the eye, e.g. after corneal perforation.
- Therapeutic: To remove inflamed corneal tissue unresponsive to treatment by antibiotics or anti-virals.
- Cosmetic: To improve the appearance of patients with corneal scars that have given a whitish or opaque hue to the cornea.

Pre-operative examination

In most instances, the patient will meet with their ophthalmologist for an examination in the weeks or months preceding the surgery. During the exam, the ophthalmologist will examine the eye and diagnose the condition. The doctor will then discuss the condition with the patient, including the different treatment options available. The doctor will also discuss the risks and benefits of the various options. If the patient elects to proceed with the surgery, the doctor will have the patient sign an informed consent form. The doctor might also perform a physical examination and order lab tests, such as blood work, X-rays, or an EKG.

The surgery date and time will also be set, and the patient will be told where the surgery will take place. The surgery only takes place when the best corresponding donor tissue is found. This can take weeks and months.

Procedure

On the day of the surgery, the patient arrives to either a hospital or an outpatient surgery center, where the procedure will be performed. The patient is given a brief physical examination by the surgical team and is taken to the operating room. In the OR, the patient lies down on an operating table and is either given general anesthesia, or local anesthesia and a sedative.

With anesthesia induced, the surgical team prepares the eye to be operated on and drapes the face around the eye. An eyelid speculum is placed to keep the lids open, and some lubrication is placed on the eye to prevent drying. In children, a metal ring is stitched to the sclera which will provide support of the sclera during the procedure.

Penetrating keratoplasty

A trephine (a circular cutting device) is then placed over the cornea and is used by the surgeon to cut the host cornea, which removes a circular disc of the patient cornea. The trephine is then removed and the surgeon cuts a circular graft (a "button") from the donor cornea. Once this is done, the surgeon returns to the patient's eye and removes the host cornea.

The donor cornea is then brought into the surgical field and maneuvered into place with forceps. Once in place, the surgeon will fasten the cornea to the eye with a running stitch

(as used in the upper image above) or a multiple interrupted stitches (as in the lower image). The surgeon then reforming the anterior chamber with a sterile solution injected by a cannula, then testing that it's watertight by placing a dye on the wound exterior.

Antibiotic eyedrops placed, the eye is patched, and the patient is taken to a recovery area while the effects of the anesthesia wear off. The patient typically goes home following this and sees the doctor the following day for the first post operative appointment.

Lamellar keratoplasty

This procedure consists in leaving just the patient's own Descemet membrane and endothelium, while transplanting approximately 95% of the cornea. The great advantage of this technique is the virtually "no rejection" post-op. The main disadvantage is that the visual acuity is not as sharp as it is with the full cornea transplantation penetrating keratoplasty). The final visual acuity is usually around 20/40.

Risks

While the cornea is avascular, there is still a potential for some blood loss, usually from suturing the metal ring to the sclera. Any blood loss is typically less than 2 ml (0.07 imp fl oz; 0.07 US fl oz).

There is also a risk of infection. Since the cornea has no blood vessels (it takes its nutrients from the aqueous humor) it heals much more slowly than a cut on the skin. While the wound is healing, it is possible that it might become infected by various microorganisms. This risk is minimized by antibiotic prophylaxis (using antibiotic eyedrops, even when no infection exists).

Graft failure can occur at any time after the cornea has been transplanted, even years or decades later. The causes can vary, though it is usually due to new injury or illness. Treatment can be either medical or surgical, depending on the individual case. An early, technical cause of failure, may be an excessively tight stitch cheesewiring through the sclera.

Prognosis

When the primary purpose of a cornea transplant is to improve visual acuity, the prognosis is dependent upon whether the rest of the eye is healthy. If it is, then it should be possible to recover normal vision.

History

The first cornea transplant was performed in 1905 by Eduard Zirm, making it one of the first types of transplant surgery successfully performed. Another pioneer of the operation was Ramon Castroviejo. Russian eye surgeon Vladimir Filatov's attempts at tranplanting cornea started with the first try in 1912 and were continued, gradually improving until at

6 May 1931 he successfully grafted a patient using corneal tissue from a deceased person. He widely reported of another transplant in 1936, disclosing his technique in full detail. In 1936, Castroviejo did a first transplantation in an advanced case of keratoconus, achieving significant improvement in patient's vision.

Advances in operating microscopes enabled surgeons to have a more magnified view of the surgical field, while advances in materials science enabled them to use sutures finer than a human hair.

Instrumental in the success of cornea transplants were the establishment of eye banks. These are organizations located throughout the world to coordinate the distribution of donated corneas to surgeons, as well as providing eyes for research. Some eye banks also distribute other anatomical gifts.

Synthetic corneas

Boston keratoprosthesis

The Boston keratoprosthesis is the most widely used synthetic cornea to date with over 900 procedures performed worldwide in 2008. The Boston KPro was developed at the Massachusetts Eye and Ear Infirmary under the leadership of Claes Dohlman, MD, PhD.

AlphaCor

In cases where there have been several graft failures or the risk for keratoplasty is high, synthetic corneas can substitute successfully for donor corneas. Such a device contains a peripheral skirt and a transparent central region. These two parts are connected on a molecular level by an interpenetrating polymer network, made from poly-2-hydroxyethyl methacrylate (pHEMA). AlphaCor is an FDA-approved type of synthetic cornea measuring 7.0 mm in diameter and 0.5 mm in thickness. The main advantages of synthetic corneas are that they are biocompatible, and the network between the parts and the device prevents complications that could arise at their interface. The probability of retention in one large study was estimated at 62% at 2 years follow-up. AlphaCor carry lesser risk of diseases that could be transmitted through donor tissue. However, they also cost $10,000.

AlphaCor surgery is reserved for patients who have had traditional cornea transplants either: 1) fail repeatedly, 2) reject due to autoimmune process, or 3) have a highly vascularized cornea that makes traditional cornea transplantation unsuitable.

Use of AlphaCor involved a two part procedure. First the AlphaCor disc is implanted under the tissue. Second, after healing many months later the membrane covering the lens of the disc is removed allowing that eye to see. Glasses will still be needed to optimize vision afterwards.

Osteo-Odonto-Keratoprosthesis

In a very rare and complex multi-step surgical procedure, employed to help the most disabled patients, a lamina of the patient's tooth is grafted into the eye, with an artificial lens installed in the transplanted piece.

Alternatives

Phototherapeutic keratectomy (PTK)

Diseases that only affect the surface of the cornea can be treated with an operation called phototherapeutic keratectomy. With the precision of an excimer laser and a modulating agent coating the eye, irregularities on the surface can be removed. However, in most of the cases where corneal transplantation is recommended, PTK would not be effective.

Intrastromal corneal ring segments

The implants manufactured under the trade name Intacs are the only patented intrastromal corneal implant that has US FDA approval and European CE Mark for both Myopia and Keratoconus. There are over one-hundred clinical articles at for clinical reference (search for Intacs and they all are arranged in chronological order) and are a well documented clinical solution for treating keratoconus. Another version of intrastromal cornea ring segments is manufactured under the trade name KeraRing and is available in South America and Europe.

With this procedure, the implants are placed in the stroma to reshape the cornea into a more natural shape. In mild myopia, this corrects a patients vision. In keratoconus, the goal is to reshape the cornea to where contact lens intolerant patients are able to achieve functional vision with contact lenses or glasses. Although, surgical procedure don't carry a guarantee, one clinically proven benefit of Intacs is that they can be safely removed and the cornea returns to its pre-operative state. Future treatment options are not affected.

Contact lenses

In the early stages and up to the more advanced stages of keratoconus, contact lenses are often used to improve vision. Contact lenses improve visual acuity of the majority of the keratoconus patients. The majority of the patients need to use hard contact lenses. Only 10 to 20% will need cornea tranplantion during their lifetimes due to progression of the disease.

New technology

High speed lasers

Blades are being replaced by high speed lasers in order to make surgical incisions more precise. These improved incisions allow the cornea to heal more quickly and the sutures

to be removed sooner. The cornea heals more strongly than with standard blade operations. Not only does this dramatically improve visual recovery and healing, it also allows the possibility for improvement in visual outcomes.

Since 2004, Amnitrans Eyebank in Rotterdam, The Netherlands, provides donor corneas pre-cut for advanced keratoplasty procedures, such as DSEK, DSAEK, FS-DSEK and DMEK. In 2007, Seattle-based SightLife, one of the leading corneal tissue banks in the world, introduced a process for the preparation of donated corneal tissue using a Femtosecond Laser. This process is known as Custom Corneal Tissue.

DSEK/DSAEK/DMEK

Endothelial keratoplasty (EK) has been introduced by Melles et al. in 1998. Today there are three forms of EK. Deep Lamellar Endothelial Keratoplasty (DLEK) in which the posterior part of the recipient cornea is replaced by donor tissue. Descemet's Stripping (Automated) Endothelial Keratoplasty (DSEK/DSAEK) in which the diseased Descemet's membrane is removed and replaced by a healthy donor posterior transplant. The transplant tissue can be prepared by a surgeon's hand or ordered already prepared for surgery. Ocular Systems Inc. was the first organization to deliver prepared grafts for surgery in 2005. DSEK/DSAEK uses only a small incision that is either self-sealing or may be closed with a few sutures. The small incision offers several benefits over traditional methods of corneal transplant such as Penetrating Keratoplasty. Because the procedure is less invasive, DSAEK leaves the eye much stronger and less prone to injury than full-thickness transplants. New medical devices such as the EndoSaver (patent pending) are designed to ease process of inserting endothelial tissue into the cornea. Additionally, DSAEK has a more rapid rate of visual recovery. Vision is typically restored in one to six months rather than one to two years. Descemet Membrane Endothelial Keratoplasty (DMEK)] is the most recent EK technique in which an isolated Descemet membrane is transplanted. The DMEK procedure combines the anatomical benefits of DSEK/DSAEK with visual rehabilitation to 20/40 or better in 90% of cases and 20/25 or better in 60% of cases within the first three months. In the UK (2010) the omly surgeon offering DMEK procedure under the auspices of the National Health Service is Mr. Ewan Craig of the Royal Shrewsbury Hospital.

Not all patients with diseased corneas are candidates for endothelial keratoplasty. These procedures correct corneal endothelial failure, but are not able to correct corneal scarring, thinning, or surface irregularity. There is currently very little data on long-term survival of DMEK grafts.

Stem cells

There is a bioengineering technique that uses stem cells to create corneas or part of corneas that can be transplanted into the eyes. Corneal stem cells are removed from a healthy cornea. They are collected and, through laboratory procedures, made into five to ten layers of cells that can be stitched into a patient’s eye. The stem cells are placed into the area where the damaged cornea tissue has been removed. This is a good alternative

for those that cannot gain vision through regular cornea transplants. A new development, announced by the University of Cincinnati Medical School in May 2007, would use bone marrow stem cells to regrow the cornea and its cells. This technique, which proved successful in mouse trials, would be of use to those suffering from inherited genetic degenerative conditions of the cornea, especially if other means like a transplant aren't feasible. It works better than a transplant because these stem cells keep their ability to differentiate and replicate, and so keep the disease from recurring, longer and better.

Biosynthetic corneas

On 25 August 2010 investigators from Canada and Sweden reported results from the first 10 people in the world treated with the biosynthetic corneas. Two years after having the corneas implanted, six of the 10 patients had improved vision. Nine of the 10 experienced cell and nerve regeneration, meaning that corneal cells and nerves grew into the implant. To make the material, the researchers placed a human gene that regulates the natural production of collagen into specially programmed yeast cells. They then molded the resulting material into the shape of a cornea. This research shows the potential for these bioengineered corneas but the outcomes in this study were not nearly as good as those achieved with human donor corneas. This may become an excellent technique, but right now it is still in the prototype stage and not ready for clinical use. The results were published in the journal Science Translational Medicine.

Epidemiology and economics

Corneal transplant is one of the most common transplant procedures. Although approximately 100,000 procedures are performed worldwide each year, some estimates report that 10,000,000 people are affected by various disorders that would benefit from corneal transplantation.

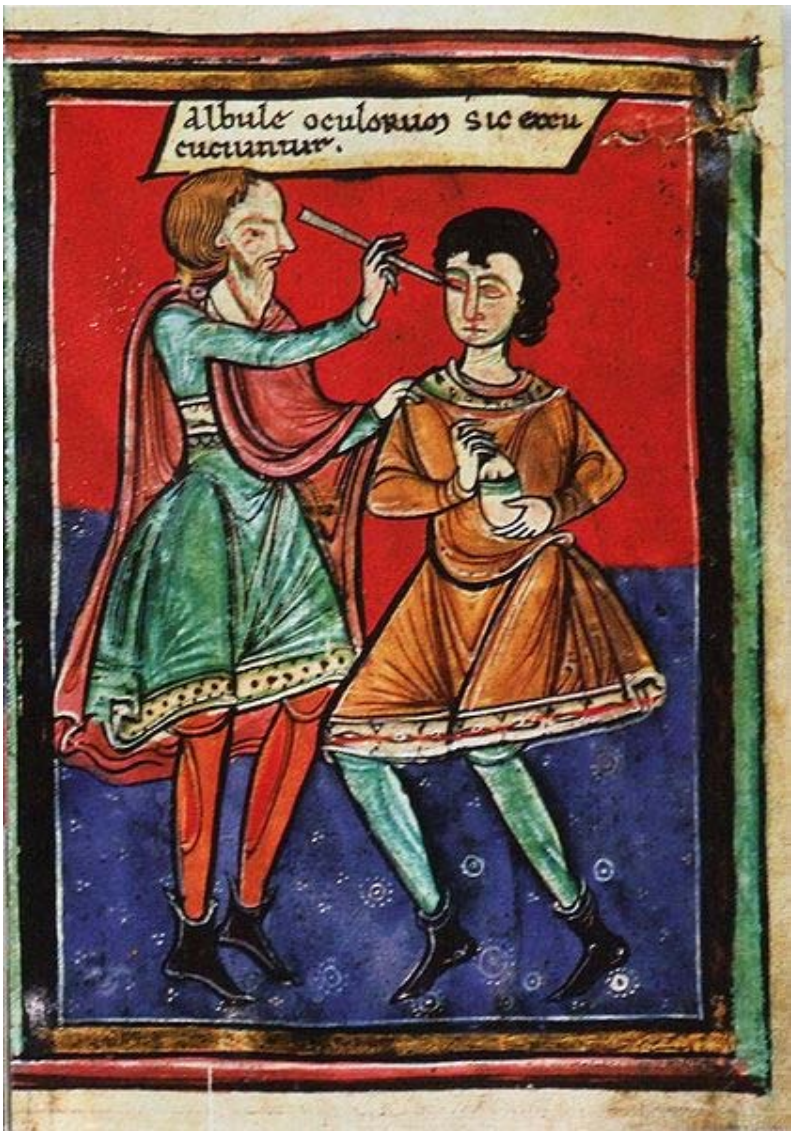
In Australia, approximately 1,500 grafts are performed each year. According to the NHS Blood and Transplant, over 2,300 corneal transplant procedures are performed each year in the United Kingdom. Between April 1, 2005 and March 31, 2006, 2,503 people received corneal transplants in the UK.

In the United States, the cost is usually covered in part by Medicare and health insurers. Reimbursement depends on your personal healthcare provider. Usually 80% of the cost will be covered by your agency. Those on Medicare will be reimbursed up to $1,200 while the remainder is left up to the patient. The average cost of the procedure ranges from $7,500 to $11,000.

In 2005, there were about 32,840 corneal transplant recipients. The estimated first year billed charges per patient, including medications, was $19,100. There were a larger number of transplants for patients over 65 than under, 18,000 compared to 14,840. There were 41,652 corneal transplants performed in the United States in 2008.

Chapter 21

Eye Surgery



Eye surgery in the Middle Ages

Eye surgery, also known as **orogolomistician surgery** or **ocular surgery**, is surgery performed on the eye or its adnexa, typically by an ophthalmologist.

Preparation and precautions

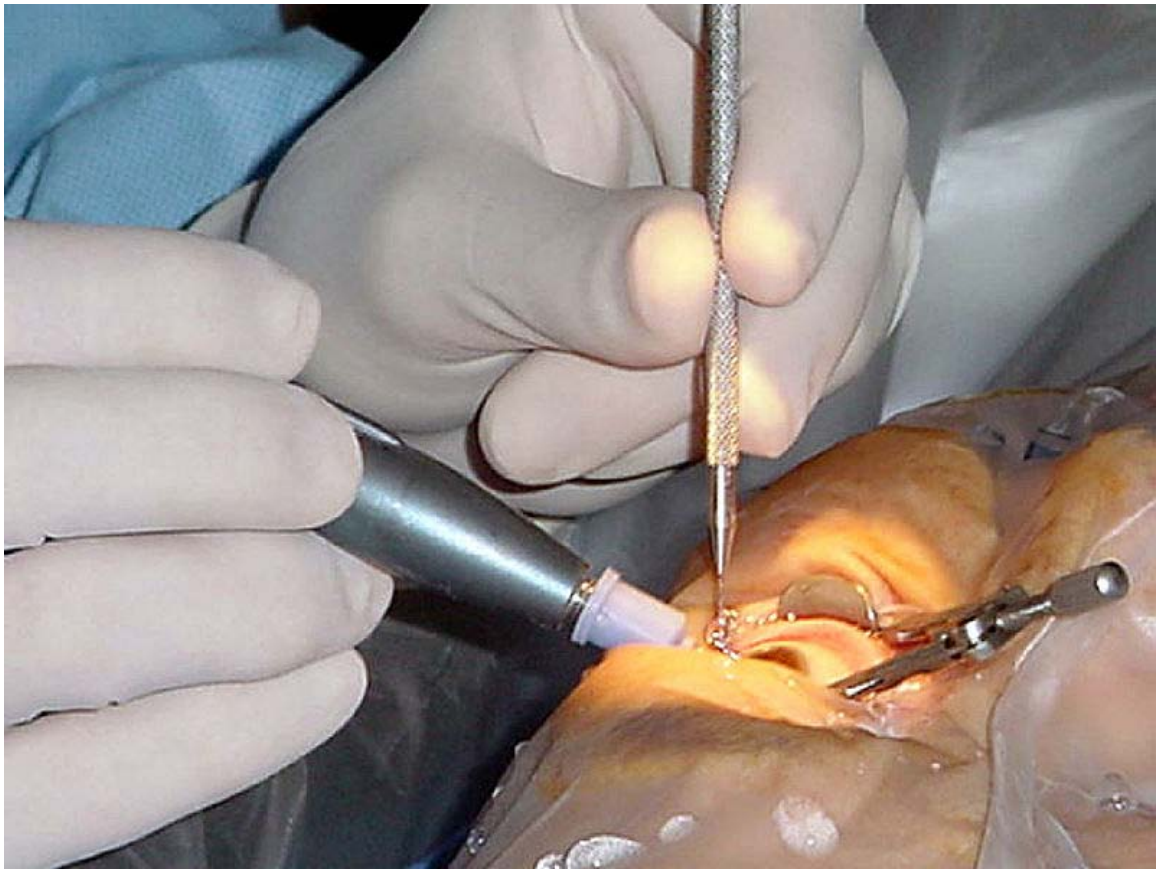
The eye is a fragile organ, requiring extreme care before, during and after a surgical procedure. An expert eye surgeon must identify the need for specific procedure and be responsible for conducting the procedure safely. Many university programmes allow patients to specify if they want to be operated upon by the consultant or the resident / fellow.

Anesthesia is essential for any eye surgery. Local anesthesia is most commonly used. Retrobulbar and peribulbar techniques for infiltrating the local area surrounding the eye muscle cone are used to immobilize the extraocular muscles and eliminate pain sensation. Topical anesthesia using lidocaine topical gel is preferred for quick procedures. In topical anesthesia, patient cooperation is a must for a smooth procedure. General anesthesia is recommended for children, traumatic eye injuries, major orbitotomies and for apprehensive patients. Cardiovascular monitoring is preferable in local anesthesia and is mandatory in general anesthesia. Proper sterile precautions are taken to prepare the area for surgery, including use of antiseptics like povidone-iodine. Sterile drapes, gowns and gloves are a must. A plastic sheet with a receptacle helps collect the fluids during phacoemulsification. An eye speculum is inserted to keep the eyes wide open.

Laser eye surgery

Although the terms laser eye surgery and refractive surgery are commonly used as if they were interchangeable, this is not the case. Lasers may be used to treat nonrefractive conditions (e.g. to seal a retinal tear), while radial keratotomy is an example of refractive surgery without the use of a laser.

Cataract surgery



Cataract surgery, using a temporal approach phacoemulsification probe (in right hand) and "chopper"(in left hand) being done under operating microscope at a Navy medical center

A cataract is an opacification or cloudiness of the eye's crystalline lens due to aging, disease, or trauma that typically prevents light from forming a clear image on the retina. If visual loss is significant, surgical removal of the lens may be warranted, with lost optical power usually replaced with a plastic intraocular lens (IOL). Owing to the high prevalence of cataracts, cataract extraction is the most common eye surgery. Rest after surgery is recommended.

Glaucoma surgery

Glaucoma is a group of diseases affecting the optic nerve that results in vision loss and is frequently characterized by raised intraocular pressure (IOP). There are many types of glaucoma surgery, and variations or combinations of those types, that facilitate the escape of excess aqueous humor from the eye to lower intraocular pressure, and a few that lower IOP by decreasing the production of aqueous humor.

Canaloplasty

Canaloplasty is an advanced, nonpenetrating procedure designed to enhance drainage through the eye's natural drainage system to provide sustained reduction of IOP. Canaloplasty utilizes microcatheter technology in a simple and minimally invasive procedure. To perform a canaloplasty, an Ophthalmologist creates a tiny incision to gain access to a canal in the eye. A microcatheter circumnavigates the canal around the iris, enlarging the main drainage channel and its smaller collector channels through the injection of a sterile, gel-like material called viscoelastic. The catheter is then removed and a suture is placed within the canal and tightened. By opening up the canal, the pressure inside the eye can be reduced. Long-term results are available, published in the *Journal of Cataract and Refractive Surgery*.

Refractive surgery

Refractive surgery aims to correct errors of refraction in the eye, reducing or eliminating the need for corrective lenses

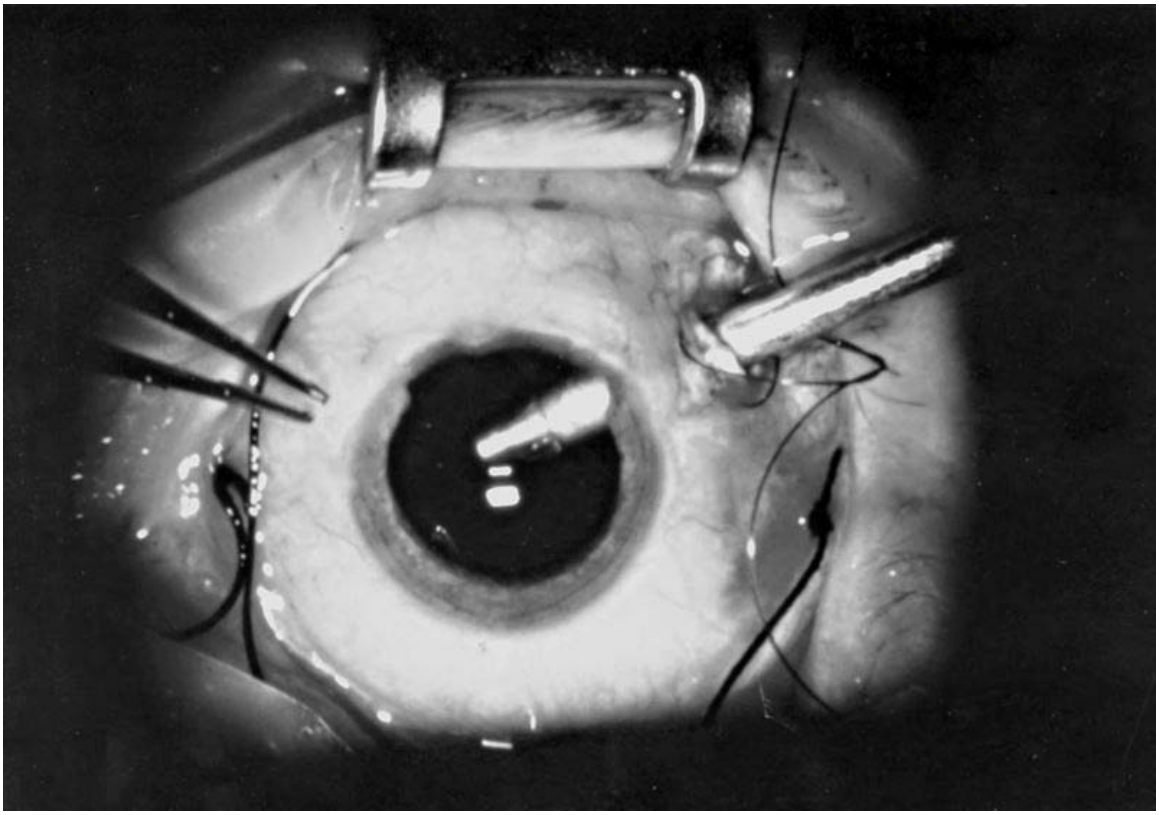
- **Keratomilleusis** is method of reshaping the cornea surface to change its optical power. A disc of cornea is shaved off, quickly frozen, lathe-ground, then returned to its original power.
- **Automated lamellar keratoplasty** (ALK)
- **Laser assisted in-situ keratomileusis** (LASIK)
    - **IntraLASIK**
- **Laser assisted sub-epithelial keratomileusis** (LASEK), aka Epi-LASIK
- **Photorefractive keratectomy** (PRK)
- **Laser thermal keratoplasty** (LTK)
- **Conductive keratoplasty** (CK) uses radio frequency waves to shrink corneal collagen. It is used to treat mild to moderate hyperopia.
- Limbal relaxing incisions (LRI) to correct minor astigmatism
- **Astigmatic keratotomy** (AK), aka Arcuate keratotomy or Transverse keratotomy
- **Radial keratotomy** (RK)
- **Hexagonal keratotomy** (HK)
- **Epikeratophakia** is the removal of the corneal epithelium and replacement with a lathe cut corneal button.
- **Intracorneal rings** (ICRs), or corneal ring segments (*Intacs*)
- Implantable contact lenses
- Presbyopia reversal
- Anterior ciliary sclerotomy (ACS)
- Laser reversal of presbyopia (LRP)
- Scleral expansion bands

Corneal surgery

Corneal surgery includes most refractive surgery as well as the following:

- **Corneal transplant surgery**, is used to remove a cloudy/diseased cornea and replace it with a clear donor cornea.
- **Penetrating keratoplasty** (PK)
- **Keratoprosthesis**(KPro)
- **Phototherapeutic keratectomy** (PTK)
- Pterygium excision
- Corneal tattooing
- Osteo-Odonto-Keratoprosthesis (OOKP), in which support for an artificial cornea is created from a tooth and its surrounding jawbone. This is a still-experimental procedure used for patients with severely damaged eyes, generally from burns.

Vitreo-retinal surgery



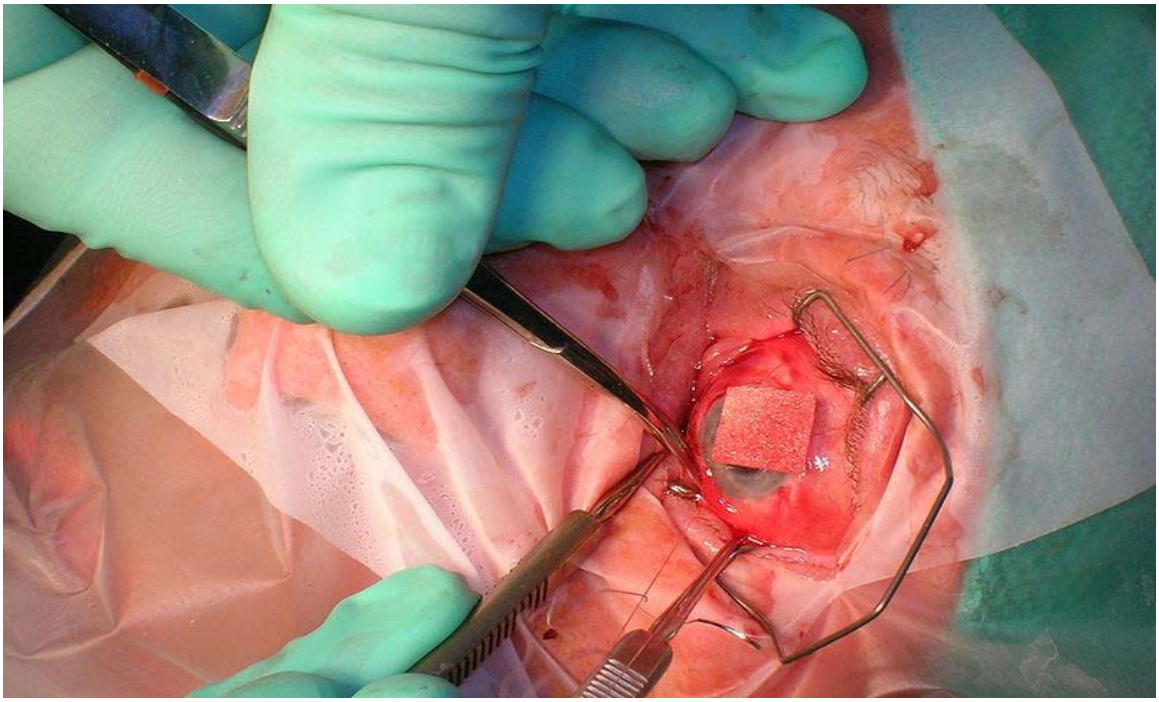
Vitrectomy

Vitreo-retinal surgery includes the following

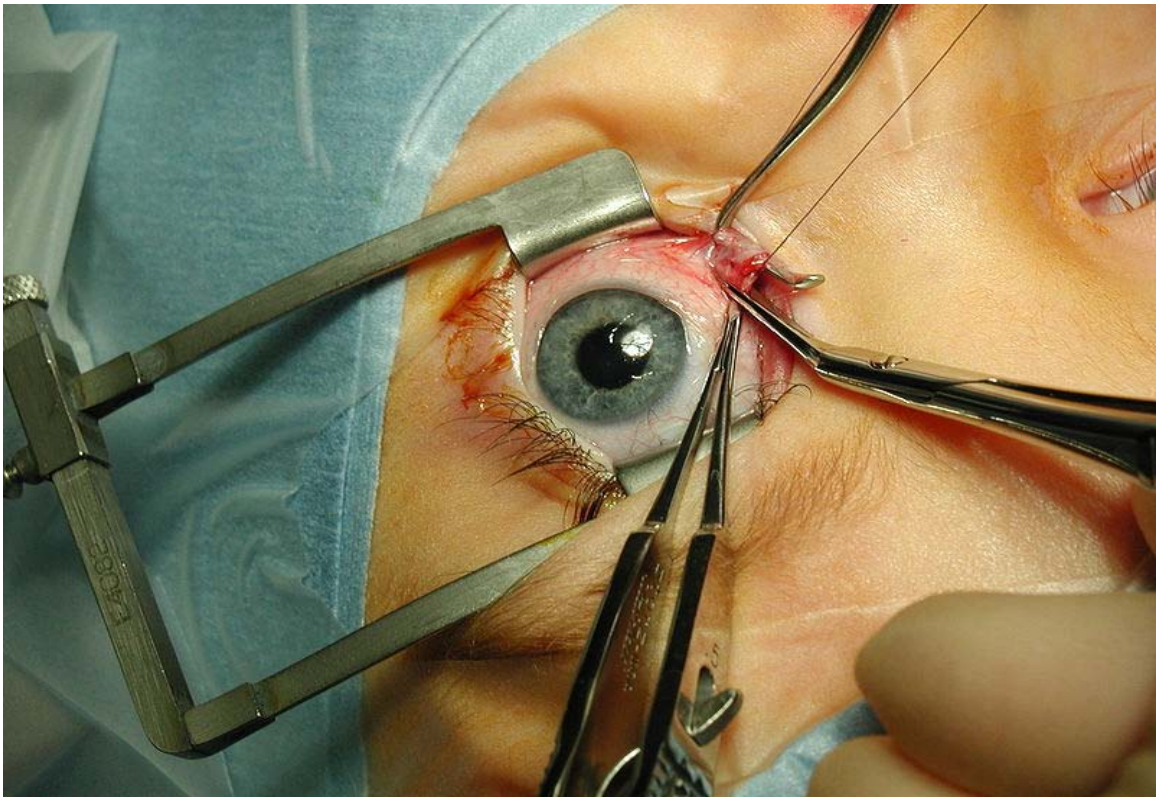
- **Vitrectomy**
  - **Anterior vitrectomy** is the removal of the front portion of vitreous tissue. It is used for preventing or treating vitreous loss during cataract or corneal

surgery, or to remove misplaced vitreous in conditions such as aphakia pupillary block glaucoma.
    - **Pars plana vitrectomy** (PPV), or trans pars plana vitrectomy (TPPV), is a procedure to remove vitreous opacities and membranes through a pars plana incision. It is frequently combined with other intraocular procedures for the treatment of giant retinal tears, tractional retinal detachments, and posterior vitreous detachments.
- **Pan retinal photocoagulation** (PRP) is a type of photocoagulation therapy used in the treatment of diabetic retinopathy.
- Retinal detachment repair
    - **Ignipuncture** is an obsolete procedure that involves cauterization of the retina with a very hot pointed instrument.
    - A scleral buckle is used in the repair of a retinal detachment to indent or "buckle" the sclera inward, usually by sewing a piece of preserved sclera or silicone rubber to its surface.
    - Laser photocoagulation, or photocoagulation therapy, is the use of a laser to seal a retinal tear.
    - **Pneumatic retinopexy**
    - **Retinal cryopexy**, or **retinal cryotherapy**, is a procedure that uses intense cold to induce a chorioretinal scar and to destroy retinal or choroidal tissue.
- Macular hole repair
- **Partial lamellar sclerouvectomy**
- **Partial lamellar sclerocyclochoroidectomy**
- **Partial lamellar sclerochoroidectomy**
- **Posterior sclerotomy** is an opening made into the vitreous through the sclera, as for detached retina or the removal of a foreign body.
- **Radial optic neurotomy**
- macular translocation surgery
    - through 360 degree retinotomy
    - through scleral imbrication technique

Eye muscle surgery



Isolating the inferior rectus muscle



Disinserting the medial rectus muscle, after pre-placing vicryl suture

With approximately 1.2 million procedures each year, extraocular muscle surgery is the third most common eye surgery in the United States.

- Eye muscle surgery typically corrects strabismus and includes the following :
    - Loosening / weakening procedures
        - Recession involves moving the insertion of a muscle posteriorly towards its origin.
        - Myectomy
        - Myotomy
        - Tenectomy
        - Tenotomy
    - Tightening / strengthening procedures
        - Resection
        - Tucking
        - Advancement is the movement of an eye muscle from its original place of attachment on the eyeball to a more forward position.
    - Transposition / repositioning procedures
    - Adjustable suture surgery is a method of reattaching an extraocular muscle by means of a stitch that can be shortened or lengthened within the first post-operative day, to obtain better ocular alignment.

Oculoplastic surgery

Oculoplastic surgery, or oculoplastics, is the subspecialty of ophthalmology that deals with the reconstruction of the eye and associated structures. Oculoplastic surgeons perform procedures such as the repair of droopy eyelids (blepharoplasty), repair of tear duct obstructions, orbital fracture repairs, removal of tumors in and around the eyes, and facial rejuvenation procedures including laser skin resurfacing, eye lifts, brow lifts, and even facelifts. Common procedures are:

Eyelid surgery

- Blepharoplasty (Eyelift)
    - **Blepharoplasty** is plastic surgery of the eyelids to remove excessive skin or subcutaneous fat.
        - **Asian blepharoplasty**
- Ptosis repair for droopy eyelid
    - Ectropion repair
- Entropion repair
- Canthal resection
    - A **canthectomy** is the surgical removal of tissue at the junction of the upper and lower eyelids.
    - **Cantholysis** is the surgical division of the canthus.
    - **Canthopexy**
    - A **canthoplasty** is plastic surgery at the canthus.

    - A **canthorrhaphy** is suturing of the outer canthus to shorten the palpebral fissure.
    - A **canthotomy** is the surgical division of the canthus, usually the outer canthus.
        - A **lateral canthotomy** is the surgical division of the outer canthus.
- **Epicanthoplasty**
- **Tarsorrhaphy** is a procedure in which the eyelids are partially sewn together to narrow the opening (i.e. palpebral fissure).

Orbital surgery

- Orbital reconstruction / Ocular prosthetics (False Eyes)
- Orbital decompression for Grave's Disease. Grave's Disease is a condition (often associated with over-active thyroid problems) in which the eye muscles swell. Because the eye socket is bone, there is nowhere for the swelling to be accommodated and as a result the eye is pushed forward into a protruded position. In some patients this is very pronounced. Orbitial decompression involves removing some bone from the eye socket to open up one or more sinuses and so make space for the swollen tissue and allowing the eye to move back into normal position.

Other oculoplastic surgery

- Botox injections
- Ultrapeel Microdermabrasion
- Endoscopic forehead and browlift
- Face lift (Rhytidectomy)
- Liposuction of the face and neck
- **Browplasty**

Surgery involving the lacrimal apparatus

- A **dacryocystorhinostomy** (DCR) or **dacryocystorhinotomy** is a procedure to restore the flow of tears into the nose from the lacrimal sac when the nasolacrimal duct does not function.
- **Canaliculodacryocystostomy** is a surgical correction for a congenitally blocked tear duct in which the closed segment is excised and the open end is joined to the lacrimal sac.
- **Canaliculotomy** involves slitting of the lacrimal punctum and canaliculus for the relief of epiphora
- A **dacryoadenectomy** is the surgical removal of a lacrimal gland.
- A **dacryocystectomy** is the surgical removal of a part of the lacrimal sac.
- A **dacryocystostomy** is an incision into the lacrimal sac, usually to promote drainage.
- A **dacryocystotomy** is an incision into the lacrimal sac.

Eye removal

- An **enucleation** is the removal of the eye leaving the eye muscles and remaining orbital contents intact.
- An **evisceration** is the removal of the eye's contents, leaving the scleral shell intact. Usually performed to reduce pain in a blind eye.
- An **exenteration** is the removal of the entire orbital contents, including the eye, extraocular muscles, fat, and connective tissues; usually for malignant orbital tumors.

Other surgery

Many of these described procedures are historical and are not recommended due to a risk of complications. Particularly, these include operations done on ciliary body in an attempt to control glaucoma, since highly safer surgeries for glaucoma, including lasers, non-penetrating surgery, guarded filtration surgery and seton valve implants have been invented.

- A **ciliarotomy** is a surgical division of the ciliary zone in the treatment of glaucoma.
- A **ciliectomy** is 1) the surgical removal of part of the ciliary body, or 2) the surgical removal of part of a margin of an eyelid containing the roots of the eyelashes.
- A **ciliotomy** is a surgical section of the ciliary nerves.
- A **conjunctivoanstrostomy** is an opening made form the inferior conjuctival cul-de-sac into the maxillary sinus for the treatment of epiphora.
- **Conjuctivoplasty** is plastic surgery of the conjunctiva.
- A **conjunctivorhinostomy** is a surgical correction of the total obstruction of a lacrimal canaliculus by which the conjuctiva is anastomosed with the nasal cavity to improve tear flow.
- A **corectomedialysis**, or **coretomedialysis**, is an excision of a small portion of the iris at its junction with the ciliary body to form an artificial pupil.
- A **corectomy**, or **coretomy**, is any surgical cutting operation on the iris at the pupil.
- A **corelysis** is a surgical detachment of adhesions of the iris to the capsule of the crystalline lens or cornea.
- A **coremorphosis** is the surgical formation of an artificial pupil.
- A **coreplasty**, or **coreoplasty**, is plastic surgery of the iris, usually for the formation of an artificial pupil.
- A **coreoplasy**, or **laser pupillomydriasis**, is any procedure that changes the size or shape of the pupil.
- A **cyclectomy** is an excision of portion of the ciliary body.
- A **cyclotomy**, or **cyclicotomy**, is a surgical incision of the ciliary body, usually for the relief of glaucoma.
- A **cycloanemization** is a surgical obliteration of the long ciliary arteries in the treatment of glaucoma.

- An **iridectomesodialsys** is the formation of an artificial pupil by detaching and excising a portion of the iris at its periphery.
- An **iridodialysis**, sometimes known as a **coredialysis**, is a localized separation or tearing away of the iris from its attachment to the ciliary body.
- An **iridencleisis**, or **corenclisis**, is a surgical procedure for glaucoma in which a portion of the iris is incised and incarcerated in a limbal incision. (Subdivided into **basal iridencleisis** and **total iridencleisis**.)
- An **iridesis** is a surgical procedure in which a portion of the iris is brought through and incarcerated in a corneal incision in order to reposition the pupil.
- An **iridocorneosclerectomy** is the surgical removal of a portion of the iris, the cornea, and the sclera.
- An **iridocyclectomy** is the surgical removal of the iris and the ciliary body.
- An **iridocystectomy** is the surgical removal of a portion of the iris to form an artificial pupil.
- An **iridosclerectomy** is the surgical removal of a portion of the sclera and a portion of the iris in the region of the limbus for the treatment of glaucoma.
- An **iridosclerotomy** is the surgical puncture of the sclera and the margin of the iris for the treatment of glaucoma.
- A **rhinommectomy** is the surgical removal of a portion of the internal canthus.
- A **trepanotrabeculectomy** is used in the treatment of chronic open and chronic closed angle glaucoma.