Current Practice

THE SPECIAL SENSES

Corneal Disease

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The transparency of the cornea is dependent on a number of factors, such as its state of turgescence, the regular arrangement of its collagen bundles, and the maintenance of its curvatures. The slightest interference with any of these will lead to loss of transparency and therefore diminution of vision.

The list of diseases which may affect the cornea directly or indirectly to cause a lowering of visual acuity is very long. Many of these diseases are, fortunately, self-limiting, and many of the changes in the cornea which they produce are reversible. There are, however, certain corneal disorders which characteristically lead to permanent and more or less gross loss of corneal function. When such a disease affects only one eye the result may be unfortunate for the patient, but when both eyes are affected it is a disaster. The natural history of some of these major corneal disorders will now be described.

Inflammations

Chemical Irritants.—Of the various chemical agents which cause severe corneal and conjunctival damage the alkalis are the most dangerous. Lime and ammonia burns have in general a much worse prognosis than most others. The first-aid treatment for all such accidents is thorough irrigation with water. This can usually be done most easily by holding the eye open under a running tap. Lime particles should be removed piecemeal as soon as possible with the aid of topical anaesthetics. The inactivation of the lime by the chelating agent trisodium ethylenediamine tetra-acetate applied as eye-drops can do much to mitigate the severity of the corneal changes.

Bacterial Infections.—These have caused much corneal blindness in the past and are still among the major blinding diseases in underdeveloped countries. It is doubtful, for instance, whether measles keratitis ever leads to serious corneal changes in healthy and well-nourished children, but because of the susceptibility to secondary infections which is such a prominent feature of measles countless eyes are lost annually in Africa and India from "measles" keratitis.

Ophthalmia neonatorum is probably the most important bacterial keratoconjunctivitis at the present time in the western world. It is a notifiable disease. The organisms which have been found to be responsible are many, but the staphylococcus is by far the most common. The TRIC virus and the gonococcus are the second most common, and all others are rare. The effects of the ophthalmia (an old word for conjunctivitis) are enhanced by the facts that babies produce no tears, that the infantile conjunctiva is without the lymphoid barrier layer, and that the eyes are kept almost constantly closed. The cornea frequently suffers badly and, indeed, may perforate, allowing the organism access to the inner eye. Treatment consists in preventing access of infection to the conjunctival sac at birth by cleaning the lids with sterile swabs, and in treating at once any discharge with local antibiotics, of which penicillin is probably still the best first choice. Bacteriological investigation should be carried out and may show that some other antibiotic is necessary. The routine prophylactic instillation of Argyrol or some similar drop (Credé's method) has much to commend it, but a number of accidents have occurred owing to mistaken ideas about the concentration of some of the agents used, and the method is no longer in general use.

Viral Infections.—Trachoma is the world's most important eye disease, and it has been estimated that 400 million people suffer from it. It is due to the *Chlamydia trachomatis* virus and thrives in conditions of poverty and squalor. Trachoma is rare and sporadic in north-west Europe, but even there is not confined to cases from abroad.

Herpes simplex infection of the cornea is the single most common cause of corneal blindness in the western world. Fortunately, bilateral corneal herpes is by no means common, and the disease, though usually recurrent, tends to recur in the same eye on each occasion. The disease provides a pitfall for the unwary practitioner, and many are misled into treating the red, painful eye it produces with steroid drops. Fluorescein staining will show the typical dendritic pattern on the cornea, and it must be remembered that apart from 5-iodo-2′-deoxyuridine no antibiotic has any effect on this virus, whereas steroids will cause the disease to spread in the most rapid and destructive manner throughout the cornea.

Allergic Keratoconjunctivitis.—Two diseases of an allergic type can give rise to serious corneal disease, though both tend to run a more mild course without visual effects. These are phlyctenular disease, which is an allergic reaction to endogenous protein (usually tuberculo-protein), and spring catarrh, which may be regarded as an allergic reaction to exogenous allergens, though such a simple explanation accounts for only some of the facts. Both tend to affect children and young adults. The topical application of steroid drops has revolutionized the treatment in these disorders.

Degenerations

Pterygium.—An encroachment of conjunctiva on to the cornea is found most commonly in sunny, hot climates, but heredity appears to be another important factor. It occurs in the British Isles among out-door workers in particular. Medical treatments are not effective, and surgical removal has a recurrence rate of some 50%. Early detection and adequate surgical excision are probably favourable to the long-term cure rate.

Band-shaped Keratopathy.—This is characterized by the deposition of a grey opacification in the superficial cornea in the area of the palpebral aperture. The material deposited consists of a mixture of calcium carbonate and phosphate, with silicon and sulphur also present in small quantities. The change may occur in any severely damaged eye. It is characteristically present in the corneae of children who have uveitis associated with any rheumatic disease. It may be associated with hyperparathyroidism or vitamin D poisoning, Bourneville's disease (tuberous sclerosis), and Fanconi's syndrome.

A number of cases, nevertheless, appear to be primary. If treatment of the causative condition is impractical or fails to

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improve the vision, the deposits can be removed most successfully by dissolving them out with trisodium ethylenediamine tetra-acetate drops after first removing the overlying epithelium.

Corneal Dystrophies.—There are hereditary degenerations. They are bilateral and come on at an age characteristic for the particular dystrophy, usually before 20 years. Some are slowly progressive and interfere seriously with vision. Each dystrophy has its own characteristic appearance and behaviour. Treatment, when it is needed, usually consists in corneal transplantation. The combined dystrophy of Fuchs, however, occurs much later in life, usually in females over the age of 50. It leads, if untreated, to blindness with pain due to rupture of the corneal bullae which are a characteristic feature of the disease. Though corneal grafting at an early stage in the disease is the most effective treatment the defect tends to recur in the graft.

Corneal Manifestations of Systemic Diseases

Collagen Diseases

Lupus erythematosus may be associated with corneal changes, but these are not usually severe.

Polyarteritis nodosa, particularly Wegener's granulomatosis, may present as a sclero-keratitis with a typical furrow-like ulcer at the limbus. Treatment is rarely effective, and the eye is often blinded.

Scleroderma and *dermatomyositis* have no corneal complications of significance.

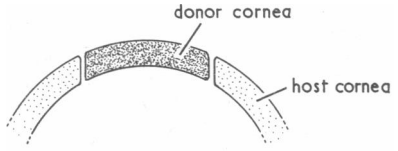


Fig.1.—Full-thickness (perforating) keratoplasty

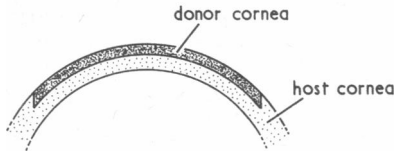


Fig.2.—Lamellar keratoplasty

Rheumatoid Diseases.—The most common ocular complication is keratoconjunctivitis sicca. The lacrimal secretion is suppressed by atrophy of the glandular elements. The dryness of the eye can be demonstrated by the staining of conjunctival and corneal epithelial cells with Rose Bengal, a dye, which normal non-keratinised cells do not take up. Treatment consists in supplying the eye with a preparation of artificial tears or in occluding the lacrimal punctae surgically to conserve such moisture as is available. Severe corneal change is by no means the rule, but vascularized tissue may spread from the limbus to cover the whole cornea, greatly reducing vision.

Crystalline Deposits

These occur in certain systemic diseases. They are occasionally associated with gout. A different crystal is deposited in cystinosis, a diffuse cloudiness of the cornea occurs in Hurler's disease, and the dysproteinaemic diseases, particularly multiple myeloma, are sometimes associated with crystalline deposits in the cornea and conjunctiva.

Treatment of Corneal Opacities

Whatever may have been the cause of a corneal opacification which is making an eye partially blind, once the opacity has become established treatment consists in trying to mitigate the effect by optical means, or if that fails in trying to remove the opacity by surgical means.

Contact Lenses

If the corneal disease has altered the refractive power of the cornea in an irregular manner, a contact lens may improve the vision by correcting the resulting irregular astigmatism.

Corneal Surgery

The removal of the superficial opacification in band-shaped keratopathy by chelating agents has been mentioned. This is a highly effective and safe method of improving vision in suitable cases. Where the opacity involves deeper layers of the cornea, however, much more radical surgery is necessary.

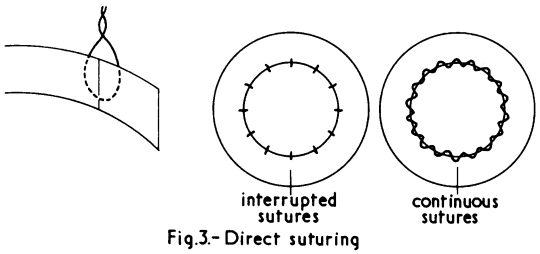


Fig.3.—Direct suturing

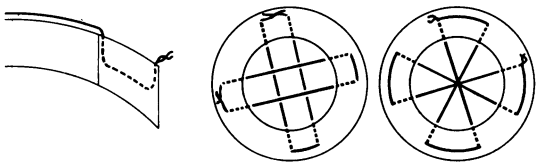


Fig.4.—Indirect suturing

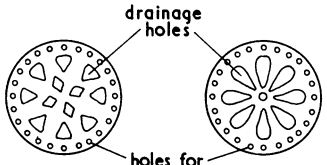
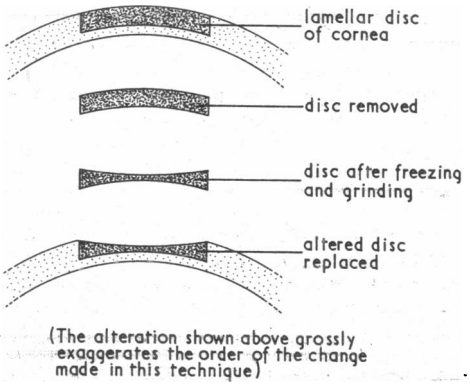


Fig.5—Fixation by splint (two types are shown)

Fig.6. Keratomileusis



This may take the form of corneal transplantation or the insertion of some form of transparent prothesis into the opaque host cornea.

Corneal Transplantation.—The first corneal transplant on a human patient was apparently performed in 1844 and was a failure. Since then techniques, instruments, anaesthetics, suture materials, microscopes, and corneal needles have been developed to make the procedure a safe and useful method of relieving corneal blindness.

The grafted material may be obtained from the individual who is to undergo the operation if he happens to have a blind eye with a clear cornea and a potentially sighted eye with an opaque cornea. Such an arrangement affords a greater prospect of success, in that immune responses to the graft probably do not occur, but unfortunately these circumstances are rare.

Several reports have appeared in which corneae have been taken from a different species and grafted into man. The failure rate, however, appears to be much higher than when the donor cornea is also human. In almost all clinical cases, therefore, homogenous donor material (from eyes donated for the purpose) is used.

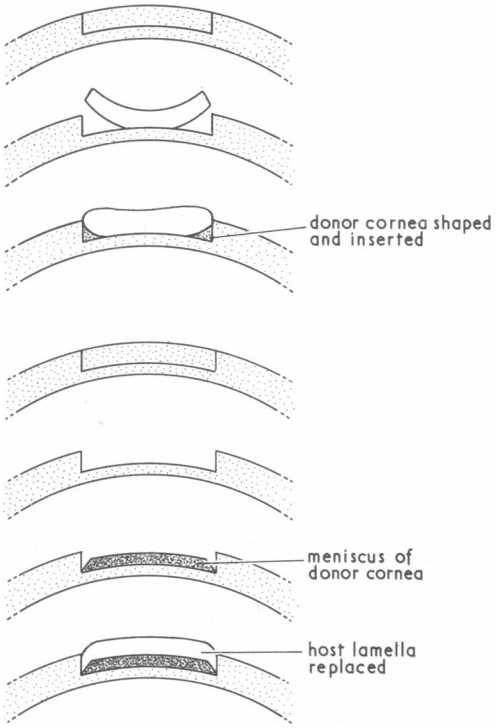


Fig.7 – Other refractive keratoplasties using donor material

Types of Graft.—The bed for the new donor cornea may be cut in different sizes, shapes and thicknesses. If the whole thickness of the host cornea is to be replaced, the graft is said to be perforating (Fig. 1) and it may be round or square. Its diameter will usually be 5, 6, 7, 8, 9 or 10 mm. The graft may replace only the outer layers of the cornea, in which case it is said to be lamellar (Fig. 2).

Of the two types, perforating and lamellar, the former gives usually a superior visual result but carries decidedly more risk since the interior of the globe is exposed. Lamellar grafts have a small range of usefulness, since many opacities involve the full thickness of the cornea, and they give a definitely inferior visual result, but they are very much less prone to complications.

The graft having been cut and put in place in the host cornea it needs to be fixed. Various methods are used. Direct sutures, either interrupted or continuous (Fig. 3), indirect sutures which avoid injury to the graft (Fig. 4) and one or other form of supporting splint (Fig. 5) all have their advocates.

Corneal grafting has been further refined recently by attempting to use it as a means of correcting very high errors of refraction. In the techniques of keratomileusis a corneal homograft is frozen and ground to a desired optical shape before being replaced in the eye (Fig. 6). In other techniques of refractive keratoplasty homogenous-donor corneae are ground to form menisci or prisms which are then inserted inside the host cornea, thus altering its optical behaviour (Fig. 7). None of these techniques of refractive keratoplasty appears as yet to have attained such a degree of accuracy as to warrant general clinical application.

Corneal Prostheses.—In certain corneal conditions the chance of a successful corneal transplant is so low as to be negligible. In general these unfavourable situations tend to occur in advanced trachoma, severe previous ulceration, bullous keratopathy, severe alkali burns, vascularization of the cornea, and disease of the inner or endothelial layer.

In an attempt to provide a substitute for biological corneae, various workers have devised prostheses of many different types, and a good degree of success is claimed for some of them. These implants in general aim to provide a clear window through the whole thickness of an opaque cornea. As might be predicted, the major problem is that of extrusion of the prosthesis. Such extrusion usually means consequent severe damage to an already very poor eye. Bearing in mind that the eye which receives such an implant will in general be one which will see by no other means, the fact that the vision may be more or less short-lived can be accepted if a careful choice of patients is made. One of the most ingenious of these prostheses, the osteo-odonto-keratoprosthesis, is prepared from one of the patient's canine teeth together with bone from its socket. The tooth, plus its socket, is cut into 0·5 mm discs, of which the most suitable is selected. The outer borders are made regular by polishing and the root canal of the tooth is enlarged with a drill to take an acrylic lens. The prosthesis then consists of an outer ring of the patient's own bone—which is capable of healing into the host cornea without reaction—and an inner ring of dentine. The dentine is firmly adherent to the bone by the alveolodental ligament, to which the lens can be fixed with dental cement. Though the technique has many attractive features it has the disadvantage of being extremely difficult to carry out to perfection.

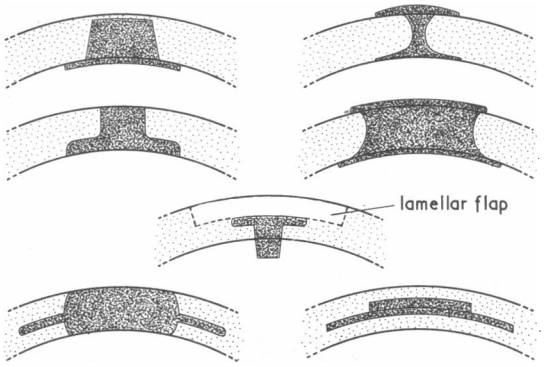


Fig. 8 – A selection of acrylic keratoprostheses

Fig. 8 shows a few of the many prostheses in recent or current use. Their variety is an indication that the technique in general is not yet of established value.

In summary, at the present time, corneal homografts are the most satisfactory answer to corneal blindness. Their limitations are definite, however, and the techniques of corneal prostheses are constantly being improved.