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THE EYE IN LEPROSY

BY

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It is estimated that there are between 50 and 100 cases of leprosy in the British Isles to-day.

To meet the needs of these unfortunate victims there has been in existence near Chelmsford since 1914 a Home maintained by voluntary subscription, where some 14 of these patients remain in voluntary isolation—for the disease is not notifiable in this country—and are cared for by Dr. Pirrie and the Mother and Sisters of the Community of St. Giles.

It is my privilege to visit the Homes from time to time to advise on the ocular involvement which is such a striking clinical feature of the disease, and it occurred to me that an outline of this rare and characteristic ophthalmic picture, together with notes of some of the cases I have seen, might be of interest.

I would first remind you that leprosy is a chronic disease, produced by the bacillus leprae, and is characterised by granulomatous infiltrations which may affect predominantly either the skin and subcutaneous tissues or the peripheral nerves; as a result two types, nodular and nerve leprosy occur, though a combination of the two is very usual.

*Proportion of Ocular Involvement in Leprosy.*—With regard to the proportion of lepers who show ocular involvement there is wide variation in the reports of observers. On the one hand de Silva puts the figure as low as 20 per cent., while Rogers maintains the eye is affected in every case if the patient lives long enough, with which Pinkerton agrees. Of the 14 cases I have observed 10 have characteristic eye changes, some of an inconspicuous nature only to be noted satisfactorily with a slit-lamp, a fact which may have some bearing on the low figures reported by some surgeons. There is general agreement that the eye is involved late, and Chance says it is unusual until the disease has been established at least two years. The eye is said to be more vulnerable in nodular than in nerve leprosy, an observation which I have not been able to confirm as these cases were chiefly a mixed type. Men appear rather more liable than women to eye affections.

*The Eyebrows and Lids.*—Thickening of the supraciliary ridges with loss of the eyebrows is a very characteristic early sign of leprosy of any type and is seen in approximately two-thirds of all cases. Diffuse infiltration of the lids beginning in the intermarginal area, and nodules, which rarely attain large

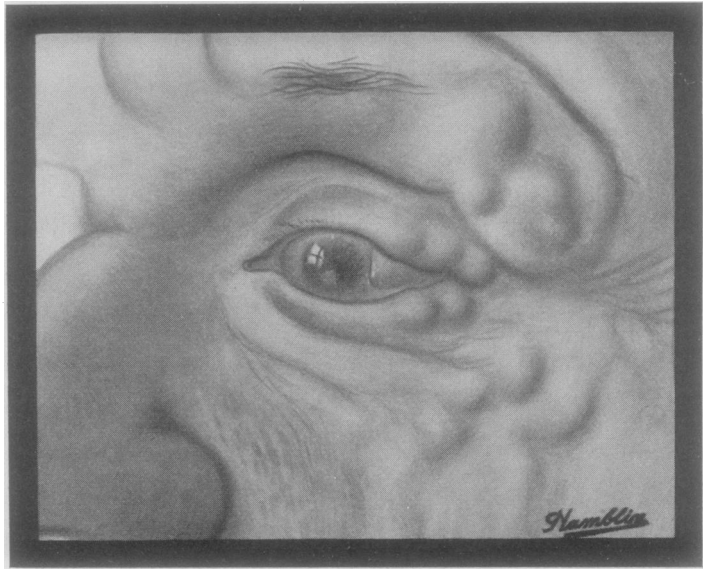


FIG. 1.

An advanced case of leprosy showing diffuse infiltration and nodular formation in the eyebrow, lids and surrounding skin.

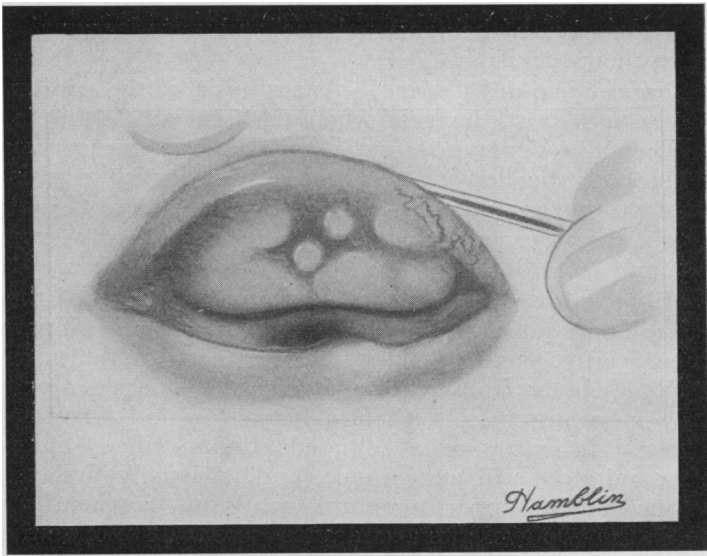


FIG. 2.

Same case as Fig. 1. Upper lid everted to show lepromatous infiltration of the tarsal plate.

size or ulcerate, are also common. This process may extend through the tarsal plate and be visible on the conjunctival surface (Figs. 1 and 2).

The eyelashes tend to become white and downy and to be shed early. In the nerve type anaesthetic patches, hairless and of different colour from the surrounding skin, are seen.

The infiltrated nodular lids tend to droop, and together with the thickened brows, ears, nose and lips are responsible for the characteristic leonine countenance.

*The Extra-Ocular Nerves.*—The terminal filaments of the VII nerve are commonly affected leading at first to diminished blinking, and later to irregular fibrillation of the orbicularis muscle, followed by paralysis with resulting ectropion of the lower lid. The III nerve is much less frequently involved, the ptosis commonly seen being, as I said before, usually a mechanical phenomenon. The IV and VI nerves seem always to escape. Wood considers the branches of the III nerve to the iris and ciliary body may also be affected, leading to a sluggish pupil and premature presbyopia.

*The Lacrymal Apparatus.*—The lacrymal gland may occasionally be the site of leprosy, and its palpebral portion require

excision as a result. Both the nose and eye being such common sites of involvement it is not surprising that the infection should spread frequently to the lacrymal sac and naso-lacrymal duct, leading to their occlusion.

*The Conjunctiva and Cornea.*—Anaesthesia of the conjunctiva and cornea is almost the rule when the eye is affected in any way. When the V nerve is first involved excessive lacrymation occurs but later the flow of tears ceases, leading to xerosis and shrinkage of the conjunctiva, not unlike trachoma. Elliot has pointed out that these changes are parallel to the early excessive sweating and later dry nature of the cutaneous patches.

One of the most characteristic changes in leprosy of the eye is the episcleral nodulē at the limbus. This may apparently arise superficially, as a secondary infection from the lid, or deeply in the ciliary body from whence it spreads forward along the anterior perforating vessels, though Pinkerton and Kirwan deny the former. It occurs as a rounded circumscribed infiltration somewhat gelatinous in appearance, and is usually first seen at the limbus in the upper temporal quadrant, frequently symmetrical in the two eyes; its appearance is very distinctive though it has to be differentiated from spring catarrh or an episcleral

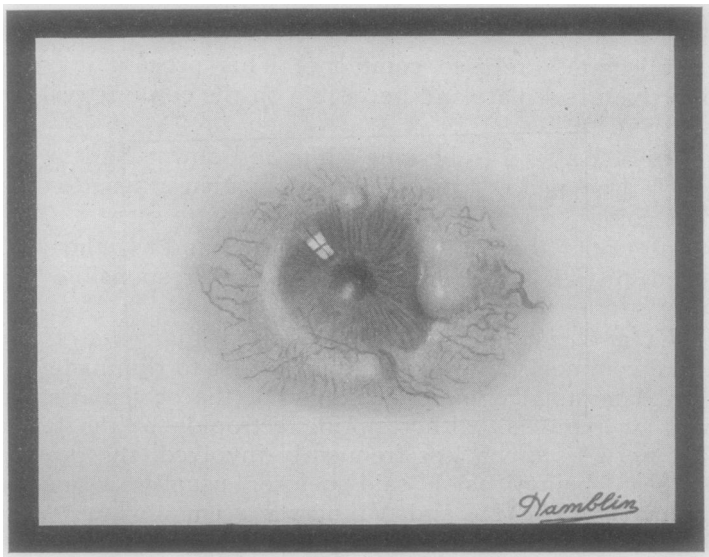


FIG. 3.

Generalised circumcorneal leprotic infiltration with two episcleral nodules. Leprotic pannus below.

growth. It tends to spread round the cornea, producing a characteristic loss of definition of the limbus, posteriorly into the conjunctiva and episclera, and anteriorly into the cornea. In the latter Bowman's membrane rapidly disappears, the substantia propria becomes infiltrated, and an anterior staphyloma may occur, though the central area of the cornea tends to remain clear till relatively late. Descemet's membrane is more resistant, though should it be destroyed perforation occurs and the eye is lost. The resulting corneal opacity becomes superficially vascularised leading to leprotic pannus, which is very like phlyctenular pannus and in no way characteristic (Fig. 3).

The distinctive primary change in the cornea is leprotic superficial keratitis, a bad term for the condition is not superficial and is quite unlike other types of superficial keratitis. It is the commonest, and often the only, eye change in leprosy, and its presence is pathognomonic. An idea of its frequency can be gathered from the fact that of the ten cases I have under observation at the Homes seven show this peculiar corneal involvement

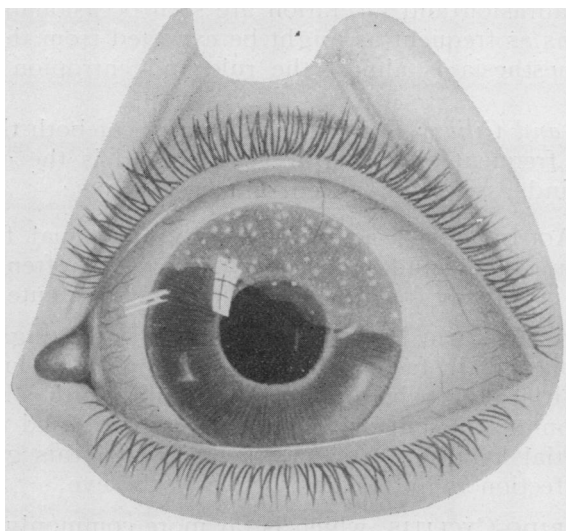


FIG. 4.

Superficial punctate keratitis of leprosy.

in unmistakable form either alone or together with other eye changes. The condition can only be observed satisfactorily with a slit-lamp when there is seen, spreading from the margin of the cornea in its upper half, a grayish milky appearance of the substantia propria, well-defined in extent, in which are scattered

numbers of white dots not unlike grains of chalk. The outline of these is irregular and they vary in size, the largest appearing under the slit-lamp about that of a small pin's head. At the limbus above the whole thickness of the substantia propria is involved, but traced centrally it tends to become more superficial—the whole being cuneiform in shape with the base of the wedge upwards. The overlying epithelium shows little change, and no vascularisation occurs. Pillat has pointed out that in apparently unaffected corneae of a patient with leprosy small nodules at irregular distances may be seen on the nerves; whereas, curiously enough, in an area of superficial keratitis the nerves are invariably normal. Unlike most manifestations of leprosy in the eye this condition of the cornea may disappear to a large extent under treatment. (Fig. 4.)

The other change seen in the cornea is a deep infiltration extending centrally from the periphery known as interstitial keratitis of leprosy. It occurs as a sequel to involvement of the ciliary body, though unlike interstitial keratitis of syphilis, the opacity does not tend to become less dense in the course of time.

Corneal abrasion and ulceration are seen occasionally, but are by no means as frequent as might be expected from the fact that corneal anaesthesia is almost the rule and entropion extremely common.

*The Iris and Ciliary Body.*—Inflammation of both these structures is of frequent occurrence in leprosy and is the commonest cause of blindness. It may occur in several ways:

(a) ACUTE IRITIS, accompanied by the usual features of pain, injection, muddy iris, etc. This is not often seen, and differs in no way from the usual non-specific acute iritis.

(b) A LEPROTIC NODULE IN THE IRIS which, again, is uncommon and, in fact, denied by some. The nodule appears at the iris angle where it is in continuity with a similar infiltration of the ciliary body, and is associated with deep interstitial keratitis. As may be imagined this gross localised infection is highly destructive to the eye.

(c) IRIDO-CYCLITIS, which is far more commonly seen than either of the preceding, and is markedly insidious and chronic in its course. Not infrequently there are co-existing changes of superficial punctate keratitis. The eye is quiet, and the keratic precipitates tend to be non-pigmented and distributed over the whole of the posterior corneal surface rather than in the lower segment as is usually seen in cyclitis. In the iris are seen a number of grayish discrete spots, on some of which may be fine pigment particles, distributed irregularly

between the collarette and the ciliary zone; these are distinctive in appearance and pathognomonic of leprosy. Some observers regard these as points of exudate, others consider them as foci of leprotic infection; I have not been able to find any account of a pathological examination of the iris which might settle this point. Apart from these gray spots the iris may appear quite normal, or may be the site of chronic iritis, the feature of which is its tendency to produce atrophy and depigmentation of the stroma with few, if any, posterior synechiae. This atrophy is at times extreme, the whole structure appearing gray and flat, the normal vessels visible, the crypts lost, and holes appearing in the stroma. As might be expected a soft degenerate eye results, and secondary glaucoma is almost unknown.

Complicated cataract is frequent.

In considering this irido-cyclitis one is struck by the similarity to heterochromic cyclitis—there is the same quiet eye, scattered non-pigmented keratic precipitates, depigmentation of the iris and complicated cataract, though of course the gray spots in the iris are peculiar to leprosy. (Fig. 5.)



FIG. 5.

Specific gray spots in iris in a case of leprosy. No other evidence of iritis.

*The Fundus.*—There is some difference of opinion whether the retina and choroid, apart from local spread from the ciliary body, are ever affected by leprosy; though, of course, examination in

many advanced cases is impossible owing to corneal scarring and the results of iritis. The majority of observers state definitely that no change is ever seen in the fundus. Trantas, however, claims to have observed white dots and pigment proliferation in 68 per cent. of his cases, and Mancione to have discovered leprosy bacilli in the retina and choroid of 5 excised eyes.

*Discovery of Bacilli in Leprotic Eyes.*—It is notorious that lepra bacilli are present in vast numbers in leprotic lesions. They may be found readily in episcleral nodules, conjunctival secretions, scrapings of affected and, in some cases, of apparently normal corneae. They have also been demonstrated in the iris and ciliary body. No bacilli are present in the deep infiltrations of leprotic interstitial keratitis which are probably the result of toxins.

Apart from the 5 cases reported by Mancione, to which I referred above, there appears general agreement that the bacilli are not found in the choroid, retina, optic nerve, lens or vitreous.

*Treatment.*—Treatment must be considered under two headings—GENERAL and LOCAL.

Of the general treatment I will only say that it consists for the most part in the administration of chaulmoogra oil, usually in the form of the ethyl esters. Following these injections there are often severe focal reactions in the eye; initial doses must, therefore, be small when it is involved. Many substances have been injected subconjunctivally, among others bichloride of mercury, ox gall, guaiacol and trypan blue; their very number makes one sceptical, and I suspect Kirwan's statement that all subconjunctival medication is useless, is the truth. The most hopeful surgical procedure is excision of the episcleral nodule in the early stage, which often arrests the condition locally and prevents corneal opacification. Tarsorrhaphy must be done for exposure keratitis, peritomy may be tried for pannus, and an iridectomy for a secluded pupil, though any optical result from the latter is liable to be disappointing as the gap becomes filled by exudate. Extraction of complicated cataract in the late stages may help, and it is noteworthy that the eye in leprosy tolerates surgical interference extremely well; Pinkerton has reported 301 intra-ocular operations without a single case of post-operative infection.

In conclusion I would like to express to Dr. Pirrie, the Medical Officer in charge of the Homes, and to Dr. Cochrane, of the British Empire Leprosy Relief Association, my thanks for their advice and help.

[This paper was read in the Section of Ophthalmology of the British Medical Association at its Annual Meeting at Oxford in 1936.]



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## THE INHERITANCE OF OPAQUE NERVE FIBRES IN THE RETINA (PAPILLA LEPORINA)

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OPAQUE nerve fibres in the retina, though they are abnormal in man, occur as a normal condition in some mammals. Johnson says that they are present in some of the rodents, more particularly in the Leporidae and in some marsupials, such as *Perameles lagotis*. In the rabbit they form a transverse band stretching right across the retina, but do not become visible until ten days after birth, when medullation has taken place. Johnson gives beautiful coloured plates of the fundi of the rabbit and *P. lagotis*, the rabbit-eared perameles. Waardenburg states that they are present in the ox and dog, but this is not shown in