TREATMENT OF CONICAL CORNEA*

BY THE LATE

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Keratoconus, according to my experience, is not often seen in the earlier stages, and patients are seldom watched from the very commencement. Slight and early cases may, however, be overlooked, especially in the strenuous work of the out-patient department, and it would perhaps be as well, in case of doubt, to make a more frequent use of Placido's disc. My experience of the disease is when it has reached an advanced stage, a definite and obvious cone has appeared in each eye, either or both of which may be marked by an opacity on the apex, and when vision is extremely reduced, usually to less than 6/60; although by looking through a small aperture or stenopoeic slit and adding concave lenses, it may occasionally be improved to 6/24, or even better. Sometimes, but very rarely, unilateral cases are met with; I had one some years ago in a lady who had very bad health and had been bedridden for years. The deformity developed quite suddenly in the left eye, and did not vary over a period of several years, the right eye remaining perfectly normal. The patient ultimately died of tuberculosis.

Patients with keratoconus are anxious to have something done to improve vision, and it is practically always a question of surgical interference; the only thing to decide is what is the best procedure to adopt.

The disease is somewhat rare and occurs far more frequently in women than men. Taking the last six cases which have come under my care, five belonged to the female sex; of these, however, there were two pairs of sisters. All four sisters may be said to have been of average physical development, except one, a trained nurse, tall, thin and not very robust in appearance, who subsequently developed phthisis. The fifth female was an idiot, stunted in growth as well as in intellect. The solitary male was a well-developed adult; he had never had good sight and was an employee at the Royal Institute for the Blind. He came to the Hospital on account of an opacity which was forming on the apex of the cone in the better eye, the left. This youth, and one of the females as well, had slight peripheral cataractous changes in both lenses. In the case of the two youngest patients it may be assumed

* Read before the Section of Ophthalmology of the Royal Society of Medicine, on January 12, 1923.
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that the affection was due to an error in development, but in the others it is difficult to account for. It seems, as my little series indicates, to have a tendency to occur in more than one member of the same family, and Mr. D. Wilson, of Huddersfield, informs me that he also has two sisters under his care suffering from the disease.

The pathology has not been fully worked out as the opportunity of microscopically examining eyes with keratoconus has seldom occurred. Parsons states that Bowman's membrane is intact, but is thinned and wrinkled and that Descemet's membrane is unchanged. He mentions the following as the main theories of causation:

1. Relative increase of intraocular tension.
2. Malnutrition of the cornea.
3. Inherent weakness of the cornea.
4. Defective embryological development.
5. Disease of Descemet's membrane and epithelium.

Collins and Mayou state that the opacity at the cone apex is due to rupture of Descemet's membrane. Fuchs says that the disease usually commences between the ages of 12 and 20, it attains a maximum but does not recede. Ulceration or rupture of the cone has not been observed. The corneal opacity may be attributed partly to laceration of Descemet's membrane and partly to mechanical injury.

Our knowledge of the pathology is therefore only scanty; what is certain is that the cornea is much thinned and may be reduced to one-third of the normal. The great difficulty is how best to deal with this serious and chronic malady. In the early cases much may be done, according to the late Mr. C. Wray, by general treatment. I should imagine, however, that his method is difficult to carry out, especially in the case of hospital patients, as it is rigorous and involves complete rest for the eyes for long periods. In the more advanced cases where nothing remains but to have recourse to surgery, general treatment must, of course, play a part, but it has no curative action. Two of my cases, the girl and the youth, were brought because of the opacity which had formed on the cone, and which had still further impaired their already very imperfect sight. In the first case surgical methods were out of the question on account of the mental condition. I strongly urged the youth, however, who had cataractous changes in addition to keratoconus, to submit to operation, as there appeared to be a fair chance of improving his vision, but he declined. For both these patients I was compelled therefore to adopt general treatment, which practically resolved itself into a firm pressure bandage and a tonic.
The result was that almost complete corneal transparency was restored in a short time; the improvement was definite and marked, and I have observed the same effect in other cases. While a pressure bandage will, however, from the support it gives to the cornea, restore transparency, it has no curative action in bringing about corneal flattening. The main operative methods are cauteryization of the cornea, with or without perforation, combined if necessary with optical iridectomy and tattooing, cauteryization combined with sclerectomy, as recommended by Fuchs, excision of the apex of the cone, or finally extraction of the lens. Of Critchett's "target" operation my experience has been limited. He has himself reported good results, but stops short of perforation. There can be no doubt that perforation is not free from risk, either of wounding the lens, or of prolapse and incarceration of iris. T. Harrison Butler also reports a case where the anterior chamber remained empty for a long time. Landolt states that perforation is not followed by inconvenience and Terrien's opinion is that cauteryization without perforation is no good.

The case of Nurse S. may be given at this point: She first came under my care in November, 1920. Her age was 24, general health good; her eyes had been all right up to four years previously. Vision was: right eye fingers only, left eye 6/60. She had well-marked conical cornea of both eyes. I first performed sclerectomy on the right eye with a view to establishing hypotony. This was followed by iritis, hypotony and the appearance of a small macula on the apex of the cone, which disappeared as the eye recovered. The patient made a good recovery, but the eye healed without leaving any obvious sign of fistulization and the corneal condition was in no way altered, or vision improved. A small buttonhole iridectomy was the only visible sign of anything having been done. She now returned to work, but eight months later came back, asking for further treatment as she was no better and could not do her work efficiently. Vision was: right eye less than 6/60, left eye with −7.00 6/36. My next expedient was to try the effect of performing the first stage of cataract extraction, making a large flap in the hope that a pressure bandage and post-operative astigmatism would produce a beneficial result. I was again disappointed. The eye healed normally and remained precisely as before. Next, after an interval of four weeks, I tried cauteryization, endeavouring to confine myself to an area not exceeding two millimetres in size; on four separate occasions I cauteryized this long-suffering nurse, perforating the cornea of set purpose three separate times; on each occasion healing occurred under a pressure bandage without any complication, the anterior chamber reforming in a few days' time. For a little while after the first cauteryization with perforation she stated that sight was
definitely improved, but the good effect soon passed away, and at no time was her vision superior to 6/60. Finally, all that remained of her six operations was the small and inconspicuous iridectomy above noted and an insignificant corneal macula below and somewhat to the outer side of the pupil. You might have thought that she would have had enough by this time, but no, she again asked me to do something more. After careful consideration I decided to extract the right lens and had arranged to admit her, but at the last moment she fell ill with pleurisy, and I heard no more until recently, when I received a letter dated December 8, 1922, with the information that she is now a patient in a sanatorium, that her sight is no better, but that she hopes it will improve as she gets stronger. In her case I think it is fair to assume that ill-health has militated, at any rate partly, against a successful operative result.

Terrien states that cauterization with perforation is the best procedure. The perforation should be made through the apex of the cone and the corneal bulge is replaced by a flat opaque scar. Later on optical iridectomy and tattooing may be needed. Vision in one of his cases was improved from 1/10 to 1/2. In my own case, however, perforation on three occasions was a complete failure, and every time the risk was incurred of serious complications. The resulting leucoma must often, moreover, be a serious drawback, it had a very unsightly appearance in one of my cases operated on some years ago, and after such an experience as that just related I shall be very reluctant again to try cauterization if there is anything better to be done.

But is there? I think so, and the case about to be related is a partial answer to the question. It is as follows:—

Mrs. L., aged 49, whose sister, as in Nurse S.'s case, also suffers from keratoconus but not so severely, first consulted me on June 14, 1921. Her sight had been very good up to 12 years previously, and had been gradually failing since. She had never worn glasses. Recently she had been so much handicapped that she was unable to go about by herself. She had suffered from exophthalmic goitre some years previously, but had quite recovered and was in good health. On examination she had conical cornea in both eyes, with a small opacity on the apex of the right. Well-marked peripheral opacities were present in both lenses. Vision was less than 6/60, and could not be improved by glasses. I tried the effect of general treatment at first, but as there was no improvement a year later, and on account of the lenticular opacities, I determined to perform simple extraction. This was done accordingly on August 2 last by my usual method of extraction under a subconjunctival bridge. This has proved, in my experience, a safe
and reliable procedure in cases of simple extraction. The patient made a good recovery and was discharged from hospital nine days after operation with a fair amount of after cataract. This was not unexpected as the lens was almost transparent throughout. She was readmitted for discission on September 25, and I endeavoured with partial success to make an incision in the capsule about a millimetre broad and half a centimetre long in the vertical diameter. This incision was based upon the well-established fact that vision in keratoconus is improved either by narrowing the pupil, or by making it resemble a stenopoeic slit. The honour of applying this principle to keratoconus is due to Mr. G. Critchett, and was embodied by him in the operation of iridodesis or, as the French call it, iridencleisis. The operation was performed by making a small opening at the limbus with a broad needle and snaring the iris as it is withdrawn with a loop of fine silk. One end of the suture was left long in case the iris should slip back into the anterior chamber again! The loop was removed on the second or third day. Eight days later a second iridodesis might be done opposite the first if considered necessary, a linear pupil being thus formed. Vision was then found to be improved. Soelberg Wells, in describing the operation, states that iritis or irido-cyclitis may occasionally be met with, but that for his part he had never met with it. The operation was performed for choice at the extremities of the vertical diameter of the cornea, as the slit was then less disfiguring and gave better vision as it was partly hidden by the lids. Of course, in my case there is no question of any interference with the iris, the idea is merely imitated by making a narrow vertical opening in the capsule. Since the operation the slit has gaped a little owing to elastic retraction, and is now about two millimetres in width. The patient was discharged a few days after discission with a perfectly transparent cornea and is now wearing -5.00 D. sphere with -1.00 cyl. axis horizontal, with which she can read, though with a little difficulty, 6/24. I think this may be regarded as a very satisfactory result; there is no disfiguring leucoma or iridectomy coloboma, such as there might have been if the cautery had been used. She herself is satisfied with the result and can see better than she has done for a long time past, she is now able to get about by herself, and altogether the improvement is material. She is to undergo a similar operation on the other eye shortly, and I shall be satisfied if a similar result is obtained. From the correcting glass one can deduce the fact that the antecedent myopia must have been about 32 diptres.

One case is not much to go upon and in Mrs. L.'s the visual acuity is not high, but the method seems to me to be rational, simple, and devoid of danger; it causes no disfigurement and from start to finish can be done in a reasonable time. I venture to
submit, therefore, that it deserves a trial, either at the outset, or when other means have proved a failure.

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BLUE CATARACT

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Since Koyanagi reported his three cases of blue cataract in the March number of the *Nippon Ganki Zasshi* of 1917, I have come across two such cases at the Royal Eye Hospital, Manchester, which I consider worth while reporting, as this form of cataract is likely to be overlooked in a large ophthalmic clinic unless each and every eye is carefully examined by oblique illumination.

This form of cataract shows no black spots against the red background of the fundus; on the other hand the opacities do not at all interfere with a clear view of the fundus, either by direct or by indirect ophthalmoscopic examination. But by oblique illumination fine greenish blue dots are seen scattered all over in the lens according to the stage of the disease, except at the extreme periphery.

Hess(1) explains the appearance of blue colour in this form of cataract by the observation of Lord Rayleigh who found that in an opalescent medium containing innumerable fine particles of a different refractive index, the dispersion of light is inversely proportional to the fourth power of its wave length. Hence the parts of the crystalline lens presenting such irregularities of refractive index caused the dispersion chiefly of short wave light, green, blue, and purple. By such diffuse light, they are rendered visible, blending into blue the colour of such spots as seen by daylight, or green as seen by yellow artificial light.

It is of practical importance to know that this form of partial cataract generally appears early in life, or, according to some, is congenital, interferes comparatively little with vision and progresses very slowly. It is capable of being removed in toto, and is particularly fitted for extraction with round pupil.(2)

The following are the notes of the cases:—

Case I. Alfred M., printer, aet. 40, came to Dr. J. G. Clegg