Ophthalmic notes / by H. Rosborough Swanzy.

OPHTHALMIC NOTES:

I.—A PECULIAR FORM OF RETINITIS PIGMENTOSA IN CONNEXION WITH INHERITED SYPHILIS.

II.—A CASE OF DERMOID TUMOUR OF THE CORNEA.

III.—AN AFTER-DANGER OF PERIPHERAL PROLAPSE OF THE IRIS.

BY

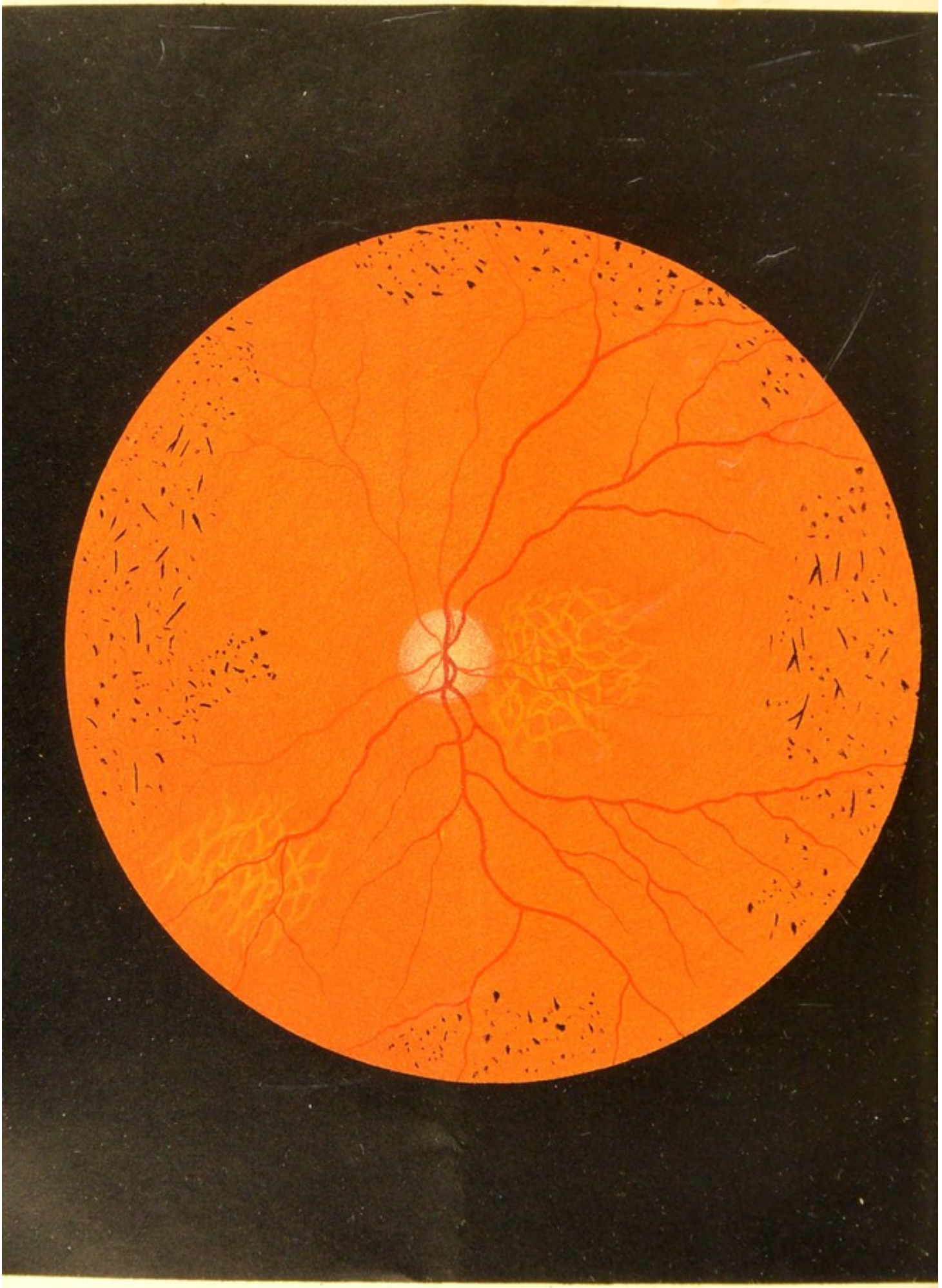
H. ROSBOROUGH SWANZY, M.B.;

SURGEON TO THE NATIONAL EYE AND EAR HOSPITAL;
OPHTHALMIC SURGEON TO THE ADELAIDE HOSPITAL;
LATE ASSISTANT TO THE LATE PROF. VON GRAEFE, BERLIN;
ETC., ETC.

Reprinted from the Dublin Quarterly Journal of Medical Science—May, 1871.

PRINTED FOR THE AUTHOR,
BY JOHN FALCONER, 53, UPPER SACKVILLE-STREET, DUBLIN.

1871.



Dr. C. E. FitzGerald, ad nat. pinx. et lith.

Forster & Co., Imp.

OPHTHALMIC NOTES:

I.—A PECULIAR FORM OF RETINITIS PIGMENTOSA IN CONNEXION WITH INHERITED SYPHILIS.

II.—A CASE OF DERMOID TUMOUR OF THE CORNEA.

III.—AN AFTER-DANGER OF PERIPHERAL PROLAPSE OF THE IRIS.

BY

H. ROSBOROUGH SWANZY, M.B.;

SURGEON TO THE NATIONAL EYE AND EAR HOSPITAL;
OPHTHALMIC SURGEON TO THE ADELAIDE HOSPITAL;
LATE ASSISTANT TO THE LATE PROF. VON GRAEFE, BERLIN;
ETC., ETC.

Reprinted from the Dublin Quarterly Journal of Medical Science—May, 1871.

PRINTED FOR THE AUTHOR,
BY JOHN FALCONER, 53, UPPER SACKVILLE-STREET, DUBLIN.

1871.

OPHTHALMIC NOTES.

I.—*A peculiar Form of Retinitis Pigmentosa in connexion with Inherited Syphilis.*

I HAD the following case for some months under observation at the Eye Dispensary of the Adelaide Hospital:—

Samuel C., aged eleven years and six months, came on the 27th June, 1870, complaining of imperfect vision of the right eye. It had always been weak, but had become much more so of late. It was affected with an external strabismus of about two lines, and with nystagmus. It was somewhat smaller than the left eye. The iris presented a dull, diffused appearance, its fibrillæ not being distinctly marked as in the normal eye. The pupil was perfectly dilatable with atropine. The patient said the sight of the affected eye became much worse in the dusk of the evening. By good light he read with this eye small words of No. 8 Snellen at six inches, and CC at fifteen feet, glasses making no improvement. The eccentric field of vision was unimpaired, for even with a very imperfect illumination he counted fingers in all directions. The left eye was slightly myopic, but its vision was nearly normal, and externally it appeared quite healthy.

The ophthalmoscope revealed those changes in the fundus of the right eye, which are so admirably represented in the accompanying drawing by Dr. C. E. Fitzgerald; and I cannot lose this opportunity of returning him my sincere thanks for the great trouble he has been at, not only in making the original drawing from life with the greatest accuracy, but also in executing with such care the various stones for the lithograph.

The optic disc was of a dirty greyish colour, and its margin indistinctly defined. The veins were normal, but the calibre of the arteries was somewhat diminished. In the retina, towards its periphery, depositions of pigment in the form of numberless small dots and streaks were seen. During six months, while the case was under observation, although these specks and streaks became more numerous, and extended somewhat more towards the centre, yet they had no tendency to become confluent, or to form themselves into stellate or bone-corpuscle shapes. They were not deposited along the course of the arteries; on the contrary, they appeared to avoid the vessels, and to occupy by preference the spaces lying between these. At one place only (above and to the left in the drawing) was I able to discover one or two specks which lay over a vessel. To the inside of the optic disc (in the inverted image), and again in the lower and outer quadrant towards the periphery, patches of choroidal changes were visible. These changes consisted chiefly in the disappearance of the pigment of the epithelial layer, but in the peripheral patch a commencing atrophy of the choroid seemed perhaps to exist. In the fundus of the left eye two patches of choroidal changes similar to those in the right were present, but only two or three small dots of pigment were observable in the retina, and the optic disc and vessels were normal.

The patient was of a sallow earthy complexion, with a flabby skin. One of the upper incisors was very characteristically notched, the other not so; the canine teeth were peg-top shaped, and the lower incisors were vertically notched. The head was large. The patient had been born healthy, but when a fortnight old had suffered from ulcerations about the anus, &c. About a year ago he had had a swollen knee, unassociated with any trauma; the swelling had been painful, but not red or inflamed-looking. This is the eldest of four children, of whom two had been still-born, and one, besides our patient, is living. The last conception had been followed by a miscarriage. The other living child is the youngest, not reckoning the miscarriage, and, with the exception of being somewhat deaf, it is and has been a healthy child. The father and mother had not been blood relations. The latter died a short time ago. Five or six years before his marriage the father had suffered from a gonorrhœa, but he could not say whether he had had a chancre at the same time or not. The medical man, however, to whom he had applied gave him

pills, which made his mouth sore. He said he might have had a chancre between that time and his marriage, but he could not distinctly remember. Six or seven years ago he had ulcerations on the calves of the legs, which have frequently reappeared since, and large scars now mark the place where they were situated. His wife had told him that the dead children had been both "diseased."

The patient was so irregular in his attendance at the dispensary that a steady course of treatment was impossible. I prescribed the iodide of potash in the first instance, and this was repeated occasionally, with apparently good effects, for, at his last visit (5th January) he said he could see much better in the dusk of the evening, and he could read short words of Snellen No. $5\frac{1}{2}$ at 6-8 inches with his right eye, and this, although the pathological changes had distinctly advanced. In the left eye the patches of choroidal changes had progressed somewhat towards a more decided atrophy, but the vision remained as good as at the first visit.

With the exception of one case of Mr. Hutchinson's, to which I shall draw attention further on, I have been unable to find any similar to the above on record. It would be inadmissible to draw a decided conclusion from an isolated clinical observation, and I merely desire to indicate those points in the case which appear of greatest interest, for the purpose of making it more available to the general readers of the Journal, and in the hope of inducing those specially interested in diseases of the eye to record their experiences and opinions with regard to those points.

In the first place, with regard to the ophthalmoscopic appearances. These differed from the usual picture of retinitis pigmentosa (*vide* Liebreich's *Atlas of Ophthalmoscopy*, Plate VI., Fig. 1) in *the shape of the pigmentary depositions, which were in the form of little dots and strokes, but not as stars or Haversian systems*, and, what would still more incline me to regard the disease as a distinct one, *these deposits did not occur along the course of the vessels, but in the spaces between them, a narrow clear interval being commonly left at either side of the vessels*, as is well seen in the drawing. In other respects the fundus of the right eye was not materially different from one affected with ordinary retinitis pigmentosa. Again, with regard to the functional disturbances, a concentric contraction of the field of vision, as the reader is aware, is, I may say, a constant symptom of the typical retinitis pigmentosa; so that, although these patients may be able to read moderately small type, they will be unable to go alone in the street (a state of vision which may be imitated by

looking through a small roll of paper or other narrow cylinder). In fact, by the gradual contraction of the field of vision from its periphery towards its centre (the macula lutea), it is, that these persons ultimately become blind. In our case *the field of vision was perfectly normal.*

The evidence of inherited syphilis in this case will appear insufficient to some, and I confess the proofs might be more conclusive. I give, however, below, Mr. Hutchinson's own authority for regarding them as adequate.

Are the changes in the fundus oculi in this case to be regarded as depending upon and caused by the inherited disease—in fact, as a retinitis pigmentosa syphilitica? Or, are they to be regarded as an unusual form of pigmentary degeneration of the retina, occurring by chance in a person affected with inherited syphilis? Further experience can alone decide these questions.

Having failed to find any parallel case in the literature, I communicated the above to Mr. Hutchinson, with a drawing of the teeth, begging him to acquaint me with his opinion, &c. He kindly sent me the following reply, which I publish with his permission:—

"4, Finsbury Circus, E.C.
Jan. 14, 1871.

"My Dear Sir,—I am very sorry to have been so long in replying to your letter. I am very much interested in the subject, and thank you warmly for sending me the case.

"In the first place, from the history and state of the teeth, I should feel no hesitation in believing your patient the subject of inherited syphilis, and that whatever is present in his eyes is due to that cause.

"I have long taught that conditions very similar to those found in what is called retinitis pigmentosa may occur in consequence of inherited taints. At page 324 of the Ophth. Reports you will find one case, but since then I have seen several much more marked, and more conclusive and I have just had a drawing made of one.

"The syphilitic cases resemble the other forms of retinitis pigmentosa in the existence of hæmeralopia. The arrangement of the pigment in a zone at the periphery. The presence of pigment in the retina. The arrangement of the pigment in stars, &c. The waxy atrophy of the disc, and extreme shrinking of retinal vessels.

"They differ, I think, in the following points:—The rapidity with which the changes advance, and the early age at which blindness is brought about. They are seldom symmetrical, or, at any rate, the two eyes rarely suffer in like degree. The pigment is often in blots or

round specks, as well as in lines and stars. There is often evidence of disease of the choroid as well. The pigment is more often irregular in its arrangement than in the more characteristic cases not due to syphilis.

"Still I have no doubt that inherited syphilis is not infrequently the cause of affections which are spoken of as 'retinitis pigmentosa.'

"Believe me, &c.,

"JONATHAN HUTCHINSON."

The following is the case referred to in the foregoing. *Ophth. Hosp. Rep.*, Vol. v., p. 324:—Case of 'Choroido-retinitis' in a girl, aged seventeen.—"The central incisors most typically notched, but no history of syphilis could be obtained.

"*Ophthalmoscope.*—Each eye the same. The discs of a dirty grey white, and the vessels so small they could only be traced with difficulty beyond the margin. There are but few denuded patches of sclerotic. The pigment was accumulated in 'blots' rather than stellate patches. There were a great number of these 'blots,' and more of them near the centre than at the periphery. Many of the pigment patches were in the retina itself."

I confess I am much inclined to agree in Mr. Hutchinson's opinion, that many cases which are regarded as ordinary retinitis pigmentosa depend probably on inherited syphilis. Attention has been little directed to this point.

Mr. Macnamara, of Calcutta, in his excellent *Handbook of Diseases of the Eye*, p. 401, says:—"I am disposed to look upon the disease [retinitis pigmentosa] as a result of inherited syphilis. . . . . The only facts which, in my mind are opposed to this view of its relation to syphilis are, that I have not noticed the notched teeth, nor have I seen the affection in more than one member of a family; and lastly, the disease does not seem to be influenced in the least by any treatment with which I am acquainted."

II.—*A Case of Dermoid Tumour of the Cornea.*

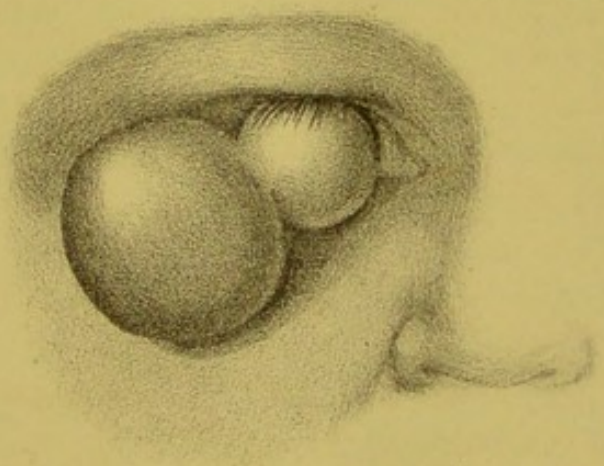
It needs only reference to any text-book of ophthalmology to ascertain that the dermoid is the most common form of tumour of the cornea. Its usual seat is on the outer border of the cornea, extending over to the conjunctiva more or less; indeed, it is as often described under the head of diseases of the latter membrane as among those of the cornea. These tumours are for the most

part congenital, they are rarely larger than a pea, and seldom increase in size after birth. They sometimes extend so far over on the cornea as to interfere with vision, and, whether for this reason or upon cosmetical grounds, their removal is often desirable. This little operation must be practised with great care, as the growth frequently implicates nearly the whole depth of the cornea, and then a radical extirpation would be attended with danger to the eye. Experience, however, shows that the portions of the tumour which are left behind, usually become absorbed, and then a very satisfactory result is obtained. The name of dermoid was first applied to these tumours by Ryba,[a] who found that they were composed of elements similar to those of the skin. Occasionally they partake so slightly of the cutaneous structure as scarcely to deserve their name; or again, when they sometimes increase in size, they do so by the accumulation of fat tissue, and are then termed by some, lipomata, or lipomatous fibromata.

I observed the following remarkable case at the hospital of the late Prof. von Graefe, in December, 1869. It was to have been published by my late master himself, but his lamented death prevented this, and thus it falls to my lot to place it on record. For the notes of the microscopical examination I am indebted to my former colleague Herr Dr. Leber, who had prepared them for von Graefe's use. The accompanying drawing was taken from life, and is a faithful representation of the original.

Martha M., aged eight months, was admitted to hospital on account of a tumour of the cornea of the right eye. The patient was otherwise perfectly healthy. The tumour was congenital, and was remarkable for its size, being, I believe, the largest dermoid tumour of the cornea which has as yet been recorded. It consisted of two portions, an anterior one of about the size of a large cherry, and a posterior one less than half the same size. The two segments were connected by a narrow band. It appears probable that this peculiar shape depended on the constriction the tumour was subjected to by the eyelids *in utero*. The base of the posterior segment occupied the whole of the cornea, with the exception of a small part about a line in width at the inside, but it did not extend over on the conjunctiva in any direction. Both segments were non-translucent and were soft to the touch. They were covered with a delicate cuticle, upon which, with the aid of a loup and in strong light, a

[a] Prager Vierteljahrsschrift, Bd. III., 1853.



MR. SWANZY. DERMOID OF THE CORNEA. (Life size.)

Dwarzaczarki. Del. Forster & Co., Lith.

few fine pigmentless hairs could be discerned. The colour of the entire growth was that of the ordinary integument. The tumour did not impede the motions of the globe, but followed them in every direction. According to the mother's account, the anterior segment had been growing slowly, until at the time of admission it had attained double its original size. The posterior segment had remained stationary.

Prof. von Graefe removed the tumour, but it was not possible to accomplish this in the way usually recommended for these tumours, by merely shaving it off the cornea, for in this instance the tumour occupied the whole depth of the cornea. It was necessary, therefore, to open the globe of the eye, and an aperture was left where the base of the tumour had been situated. Through this a considerable portion of the vitreous humour escaped; the crystalline lens, however, could not be found, and was thus shown to have been congenitally deficient.

The microscopical examination showed that the tumour consisted of an external cutislike layer, and beneath this of a thick layer of fat tissue, of which, in fact, the great mass of both segments consisted; and this tissue differed in no way from the usual subcutaneous fat. The cutislike layer was formed of a superficial epithelial covering, and of a layer of cellular tissue containing hair follicles, and very beautifully developed sudoriferous glands, the latter extending as deep as the fat tissue. There were few papillæ, and these were imperfectly developed; on the other hand, in some places, numerous and tolerably large vessels were observable. It was with difficulty that some minute rudimentary sebaceous glands were discovered in connexion with the hair follicles, but the ordinary acinous sebaceous glands were not present. The cornea, as such, was quite absent, but a thin layer of fibrous tissue which occupied an analogous position, might be supposed to take its place. It could not be ascertained whether the tumour had caused an arrest in development of the cornea, or whether the latter had been gradually destroyed by the growth of the tumour. A rudimentary iris lay close against the pseudo-cornea behind, so that there could have been no anterior chamber. Concerning the size of the globe it was not possible to form an exact estimate, for it was at first so concealed by the tumour, and after the operation it became collapsed. It did not appear, however, to deviate much from the normal size. That the name of dermoid is properly applied to this tumour, is evident from its minute structure.

It is not necessary to try to account for the heterology of the tumour. There is nothing more wonderful in such a growth occurring on the cornea, than in an ovarian tumour which contains hair and teeth. The fact of the deficiency of the crystalline lens has, however, suggested to me a possible explanation for this case, which I may be permitted to mention. As the reader is aware, the crystalline lens, which is an epithelial structure, is developed in the fœtus by the invagination of a portion of the cuticle in a depression of the primary optic vesicle. If we suppose this invagination not to have taken place, or that instead of being folded inwards, the cuticle had been folded outwards, no lens would have been formed, and we may easily imagine how such a growth as this may thus have had its origin.

I must not omit to mention, that Ryba believed many of these dermoids to originate in an arrested development of the eyelids. In our case the eyelids were perfectly normal.

Manfredi, of Pavia, has published[a] an interesting case of dermoid of the cornea accompanied with arrested development of the globe, the latter being in a condition of microphthalmos; but it does not appear that one part of the eye was more imperfectly developed than another, or that any part was completely absent.

III.—*An After Danger of Peripheral Prolapse of the Iris.*

The dangers, immediate and remote, which are most liable to attend prolapse of the iris into a perforating wound or ulcer of the cornea, are so well known, that it is needless to dwell on them here. The three following cases, however, demonstrate a danger incident, I think, to peripheral prolapse, which appears to be less known. I give but a short note of each case.

1. R. B., aged forty, was admitted on the 4th May, 1869, on account of a purulent iritis of the right eye, attended by agonizing pain. The inflammation had commenced two days previously, and until then he had had good use of the eye. About twenty years ago the patient had received a perforating wound of the sclerotic, to the outside from the cornea, and about half a line removed from the latter. Prolapse of a portion of the iris into the wound occurred at the time. The wound healed, and the prolapsed portion of iris remained in it, leaving a coloboma to the

[a] Lipodermoide congenito con difetto di sviluppo dell'occhio. Revista Clinica. 1869.

outside of the true pupil. The cicatrix containing the prolapsed iris was but little raised over the surface of the sclerotic, and there was little or no dragging of the iris towards the cicatrix. The patient had not since then received any injury to the eye that he can remember, certainly none within the last few days. He had of late been a good deal exposed to the weather, and had had much fatigue. At the time of admission the perception of light was good, but the projection imperfect.

The patient remained five days in the house, and was then obliged to leave in consequence of his private affairs. During this time, in spite of treatment, the functions of the eye sank gradually until they had reached a minimum, and all hope of saving the vision was gone.

2. L. M., aged thirty, admitted 6th May, 1869. Eight months before admission he received a perforating wound in the right eye, to the outside of the cornea, in the corneo-sclerotic junction; a portion of iris was prolapsed into the wound, and a coloboma was left in place of the prolapse. The wound had healed without any inflammatory reaction, the iris remaining adherent in it. The cicatrix was not prominent. The iris was dragged a good deal over towards the cicatrix. Since that time the eye had given the patient no trouble, and its vision had been perfect. The day before admission, without any trauma or other immediate cause, the eye became irritated; towards evening this irritation increased and great pain set in, and early next morning the patient came to the hospital. Purulent iritis had already commenced. The process could be distinctly seen to be most intense towards the point of incarceration. The whole iris had, however, assumed a yellowish hue, and the aqueous humour was opaque. The slightest pressure on the eye gave much pain, showing that the ciliary body was inflamed. The perception of light was good. The projection was in every direction uncertain. A very doubtful prognosis was given.

All our efforts to arrest the suppurative process proved unavailing. The power of vision sank daily until it quite disappeared. The eye became rapidly phthisical, and it was soon necessary to enucleate it in consequence of its fellow becoming sympathetically irritated.

3. F. R., aged twenty-seven, ploughman, admitted 25th November, 1869, with panophthalmitis of the right eye. Ten days before, in the forenoon, he had received a slight blow from the hairs of his horse's tail in this eye, while stooping to arrange some part of the

harness. He experienced much pain during the remainder of the day, and by next morning the eye was quite blind. On examining the eye the pupil was found very much displaced upwards, in consequence of an old prolapse of the iris into an ulcer at the upper corneo-sclerotic boundary. The great distension of the globe had now caused the cicatrix to gape. In the left eye a symmetrical ulcer had been present, leaving a similar condition of the pupil and iris. It now appeared that two years previously he had suffered from an acute conjunctival blennorrhæa in both eyes, which had been the cause of these ulcers. Since then he had had good use of his eyes.

The treatment adopted in Cases 1 and 2 was actively antiphlogistic. Mercurial inunctions until the gums were affected, emetics, purging, and locally, periodical warm fomentations, the pressure bandage, and atropine.

It is remarkable that the first two cases should have been under treatment simultaneously, and should have been so similar in many points. They differ, indeed, considerably in the length of time which had elapsed since the receipt of the trauma and the occurrence of the inflammation. In one 20 years, in the other 8 months.

In Case 2 it was evident that the inflammatory process originated in the iris at the place of incarceration. In Case 1 the suppuration had advanced too far to decide this with certainty from the appearances themselves, but in the absence of any reason for so violent a process, and with the aid of Case 2, all reasonable doubt as to the prolapse being the starting point of the iritis, must be removed. With regard to Case 3, such an injury would never produce an inflammation of this kind in a healthy eye, whatever other serious consequences it might have.

It would seem then—*a*. That the presence of a peripheral prolapse of the iris renders the eye always liable to be suddenly attacked by a purulent iritis *foudroyante*; *b*. That the immediate exciting cause may be very slight or even not discoverable; and *c*. That, in consequence of the great rapidity of the process, the prognosis in these cases is of the worst.

Why peripheral prolapse should expose an eye to such a danger, it is not easy to understand. Is the proximity of the ciliary body somehow in play? It cannot depend on the tension and dragging of the iris, for in Case 1 this was nil.