Relations

There is one last point which I should like to make. In dealing with the congenital syphilitic you have not discharged your duty when you have treated him. His parents and brothers and sisters ought to be tested and if necessary treated.

Fourteen years ago I treated a child for interstitial keratitis. I advised the doctor that the mother's blood ought to be tested. It was not done: I saw her this year—she now has an aortic aneurysm. There is little doubt in my mind that such a tragedy could have been avoided quite easily. And, as with the congenital, so with the acquired. The wife or any other contact of acquired syphilis ought to be considered, and not left to rot for fear of a social or family trouble. I know the difficulties, but I also know that they ought to be faced.

The Future

War brings many horrors in its train; not the least of these is an increase in syphilis.

I indicated at the outset that syphilis had shown a definite annual decline in the years preceding the present war. I regret that the number of new acute cases is now beginning to show an alarming increase; this will not be seen in ophthalmology until later on, but unless the acute cases are properly treated it will surely occur, it may be not for ten or fifteen years, when the congenital syphilitic children reach puberty or the half treated cases arrive with lesions of the central nervous system.

CHORIO-RETINITIS JUXTA-PAPILLARIS (E. JENSEN)*

BY

DR. R. G. POSTHUMUS

HAARLEM

THE above-mentioned affection has of late years been brought comparatively little to the notice of the readers of great ophthalmological periodicals, though its syndrome is not yet completely known to its full extent and in all its details. This is apparent from the mere fact that besides the one mentioned above no less than three other names have been suggested, on the assumption that these names give a better description of the said syndrome. Thus we can also speak of chorio-retinitis para-papillaris of

* Received for publication, 1939.

Blessig, neuro-fibrillitis retinae of van der Hoeve and finally of neuritis retinae of Zeeman. Rönne rightly wrote in 1915, "Unzweifelhaft ist die Klinik dieser Krankheitsform bei weitem nicht ausdebatiert." In the main it is a matter of three points requiring further elucidation, viz. the aetiology, the portal of entry and the primary site of the inflammation. The difficulty of approaching this syndrome more closely lies in the fact that no pathologico-anatomical material is available, because such eyes need not be enucleated as, with very few exceptions, the prognosis of the disease is favourable. Favourable with respect to sight, but nevertheless this form of inflammation may result in big patches lacking from the field of vision after cicatrization of the focus (foci). These defects of the visual fields are among the most characteristic manifestations of the affection, for the rest it resembles an ordinary chorio-retinitis and according to some investigators it is identical with this. The fact that in many arbitrary choroidal foci the perimeter research is not or cannot be rightly performed owing to the turbid media, is probably the cause that these defects are not much more frequently met with in all kinds of chorio-retinitis.

Originally the syndrome as to its fundus changes has been described under the name given to it by the first investigator, who looked upon it as a special change, viz., chorio-retinitis juxta-papillaris, in which the foci lay beside or leant against the papilla of the optic nerve. After the communications made by Jensen there followed others in which there were also described foci which had led to radial or arched scotomata, which foci were more, sometimes even considerably more, peripherally situated. Thus Blessig suggested the name para-papillaris. In all these syndromes stress was always laid on the second part of the name chorio-retinitis, i.e., the retinitis.

Van der Hoeve proposed, however, to change the name into neuro-fibrillitis retinae, by which he indicated that the cause did not lie in the vessels of the retina, as was originally assumed, but in the nervous fibres. According to him there was no question of a lesion caused by compression of a vessel secondarily causing a nutritional derangement in a special area of the retina, which bad nutrition gave rise to derangements far outside the area of the focus proper (Jensen, Fleischer), but it was an affection of certain fibres situated within a sharply circumscribed area. So it should not be looked upon as a retinal ischaemia, a retinal infarction, which the syndrome also resembled far less in its clinical symptoms, but rather as an interruption of the irritability of nervous fibres because of an inflammation. The same idea was kept in view, albeit to a somewhat wider extent, by Zeeman, in using the name neuritis retinae.

In addition to cases from personal observation, I was also, owing to Prof. Zeeman's kindness, provided with data about some twenty patients from the Amsterdam clinic, see below.

For the sake of convenience I have left out additional and less important facts and brought the chief data together in one table*, so that one would be able to survey at a glance the most important features in their order.

In connection with the above-mentioned cases I wish to go a little farther into these things. First of all the question should be put whether the primary site of the disease may not be found in the choroids or in both posterior sclerotics, as Wagenmann, Gilberts, Loewenstein, Schieck and Rönne have thought, among others. With regard to this Schieck draws attention to the fact that the irido-cyclitis often attending it, stamps the disease as an ordinary uveitis. To this the objection may be raised that in the large number of cases of iritis, respectively irido-cyclitis, met with in the larger clinics, the combination with a single circumscribed fundus-focus and corresponding defect of the visual field (Fig. 1), is a comparative (rather great) rarity, so that by this combination a separate stamp is put on the cases; and in the symptoms just mentioned we may not in the least see an ordinary, frequently occurring, complication of irido-cyclitis. On the other hand one would sooner infer from the fact that the combination of a solitary fundus-focus with one of the above-mentioned defects of the visual field not seldom shows a sign of cyclitis (turbidity of the vitreous humour in 19 out of 22 patients) that the first-mentioned symptoms

* This table is drawn up in Dutch. Under present conditions we regret that it is not possible to reproduce it—EDITORS.

FIG. 1.

Visual fields. First five representations of visual fields of the first patient. (A) In which the course of the defect of the visual field can be traced. These visual fields belong to the fundus shown in Fig. 2. Distinct spreading from the centre to the periphery. The sixth visual field pertains to the second patient, whose fundus has been represented in Fig. 3. In all visual fields the fact becomes manifest that the course of the destroyed part of the visual field is linked up with the course of the fibrils and not with the supply of the region in question by a definite blood-vessel.

not too seldom seem to lead to irido-cyclitis. This supposition is obvious also for the reason that distinct symptoms of cyclitis in the beginning were lacking in some of our patients and only manifested themselves later on (patients N and O). Besides the number of cases of secondary iritis in affections of the posterior half of the globe is rather great. We see it in detachments of the retina, in tumour necroses, and we have been well enough acquainted with this development since Fuchs's investigations.

FIG. 2.

Fundus of the first patient. (A) In the focus blood-vessels of the choroid are to be seen as indicative of the fact that the destruction has only affected the retina and the pigment-layer with superficial choroid-layer. Besides it is clearly to be seen from the colour that more has been saved than the chalk-white sclera. Through the focus runs a retinal vessel showing no changes except a few slight haemorrhages situated beside the vessel, just outside the focus. No turbidity of the vitreous humour of any importance. Fundus further normal, with the exception of a small old focus not to be seen in the picture, which is more peripherally situated, very slight and also rather superficial. This focus is older than the one situated beside the papilla and might be a primary affection; more probably, however, a process originated via the blood-flow at an earlier period, perhaps through the same aetiological factor

Consequently the association of symptoms in the choroid, the iris and the ciliary body can certainly not be alleged as an argument in favour of the origin of the affection as a primary uveitis. This does not at all preclude, however, that the process may primarily be a case of choroiditis. The distinct changes in the choroids, which always remain behind after cure, at first made

FIG. 3.

Fundus of the second patient. (B) A deeper focus, in the centre reaching down to the sclera, with a truncated blood-vessel on its border, becoming somewhat thinner there, but afterwards resuming its normal size. This phenomenon has been described before. In front of the focus 8 D. inwardly, consequently about 2 and two thirds mm. more towards the centre, a sausage-shaped turbidity in the vitreous humour, remainder of the turbidity of the vitreous humour originated on that spot, which at first withdrew the whole fundus-syndrome from observation, so that at the time no diagnosis could be drawn up, respectively no visual field be obtained. The extension both inward and outward and the greater defect in the field of vision correspond with one another, also when compared with the other case without turbidity of the vitreous humour, shallower focus and a smaller defect in the visual field.

this supposition seem quite acceptable. In proportion to the violence of the process only the inner layers or the entire thickness of the choroid coat were supposed to be affected (Figs. 2 and 3).

Now in primary choroiditis an affection of the outer layers of the retina, of the percipient elements, is to be expected in the first place, however; and, indeed, in choroiditis one sees cases of scotoma pointing to this. In the affection described here the shape of the scotomata indicates, however, an early impairment of the nervous fibres, so that in the first place one must think of a disease of the retina. The syndrome described here, on the other hand, does not in the least resemble a typical retinitis of the kind well-known to us in diabetes, uraemia, and hypertension (Fig. 4). Neither does it look like the cases of retinitis attending infections,

FIG. 4

Fundus-photograph. I. Fundus-syndrome of patient D. Fresh focus with veiled vessels, papilla also seen to be locally blurred. II. Fundus-photograph of patient G. Juxta-papillary focus. Old para-papillary focus with close to it a new recurrence. III and IV. Two photographs taken a month after each other. Old para-papillary foci, of which one large one, quite atrophic, by the side of it a recurrence, on the first photograph fresh, with apparently thinner vessels; on the other photograph atrophy already beginning on the border. V. Fundus of patient M. Large, fresh, parapapillary focus; blood-vessels for a part completely veiled, papilla also slightly blurred. VI. Same fundus, after three years a small local recidivism in the old focus. Vessels taken up again into the inflammatory mass.

sepsis and diseases of the blood. Particularly there are as a rule lacking the multiple diapedetic haemorrhages and exudations which in the said affections are always met with between the nuclear layers and elsewhere in the retina. There are also lacking here the varicose nerve fibres and their ganglionic degeneration, which by their situation along and over and under the retinal vessels sometimes makes one suppose a degenerative process in the layer of the nerve fibres. The differences mentioned give no ground, however, for the rejection of a localization in the retina; at most it may be inferred from this that the diffuse and the diapedetic, haemorrhagic, angiospastic or toxic character of the said retinitides is lacking, and point to the solitary and more exudative or inflammatory character of the focus (*Cf.* Fig. 4). There is a striking resemblance between this ophthalmoscopical syndrome and the fundus-syndrome one may meet with after a perforating lesion (Fig. 5), in which an iron-splinter has hit the retina close to the papillary border and has infected the eye; here, too, there is exudation on and in the retina, attended with the loss of a sector of the visual field as a consequence of the cutting through

of a bundle of nerve fibres. Formerly the defect in the visual field in this case was sometimes ascribed to interrupted circulation in the quadrant in question; the visible symptoms of such a disturbance, *i.e.*, stricture of the arteries and dilatation of the veins, haemorrhages and exudations, afterwards collaterals, are always lacking here. The shape of the defects in the visual field in case of interrupted circulation is also different as a rule. In contradistinction to this the defects in the disease discussed here perfectly resemble the missing parts in the field of vision after lesions in several quadrants. As to the course of the nerve fibres in the retina and optic nerve I would refer to the experimental investigations of Sjaaff and Zeeman and the literature mentioned by

FIG. 5.

Fundus-photograph of a patient with lesion caused by iron. The little lump is situated para-to juxta-papillary and has formed an exudation. Thus the syndrome of the same inflammation arose in quite another way. This process also perimetrically gave the syndrome of the disease of Jensen.

them, as well as to the discussions held between van der Hoeve and Igersheimer. On accurately inspecting the series of photographs of the visual field of our first patient, one sees that destruction arises from the centre to the periphery in centrifugal direction; this observation seems to indicate that also in man there is some system in the arrangement of the nerve fibres from different areas of the retina.

About the aetiology, opinions also greatly differ. Gilbert believes the cause is not known with certainty. Köhne ascribes it to influenza, Schertlin and A. Loewenstein to tuberculosis, Pavia to lues (combined with abuse of alcohol and tobacco), Higuchi to tuberculosis, Tristaino is of opinion that the cause often lies in the latter disease. Groes-Petersen also looks upon lues as one of the causes. Igersheimer, without making use of the name of Jensen, describes a typical case having lues as its cause, Wolff met with a case with empyema of the jaw-cavity, Leber already at an earlier time in the pre-Jensen days described a case in agreement with the description given here of a woman with a pulmonary affection.

Wilmer looks upon its origin in tonsils or secondary cavities as possible, Fleischer thinks of multiple sclerosis. Zeeman has described a case of distinctly luetic origin, but points out that in looking for aetiological factors it will be useful to think of the analogies and seeming relationship between this change, which he would like to call neuritis retinae, and neuritis optica.* In our cases a cause can mostly not be given with certainty; sometimes one would be inclined to think of a focal infection on the ground of affections of the teeth; in a few cases a slight trace of tuberculosis can be discerned, even though it be at a far distance. Whether pregnancy may lessen the resistance owing to which the disease may come into being, is perhaps demonstrable in two cases. In none of the cases was there any indication of multiple sclerosis, to which special attention has been given. The assertion of Fleischer, that the sclerosis and plaques are of importance in this syndrome, finds no support in our cases. For the rest it is not known either whether people with whom multiple sclerosis was diagnosed with certainty, suffered from this ocular affection in a much greater measure or even in a somewhat more considerable way. For that matter the whole neuro-trophic character of the virus or of the causation cannot without more proof be accepted as certain.

One is rather inclined here to look for an analogy with a perivascular affection of the lymphatic vessels. That the retinal vessels are of importance in spreading the causation is quite acceptable. The occurrence of one or more vessels in or near every focus and the origin in a small number of cases of a few localized haemorrhages, give rise to more than a supposition in this direction. That the vessel itself should be the cause and that consequently the focus should come into being *via* the diseased vascular wall is not altogether incontestable; considerable haemorrhages, even more than a few punctiform small bleedings and that only in a limited number of cases are lacking. Also the distribution by means of the blood flow would rather be expected in places with slow flow, in the final arteries and in places where the vessels bend acutely or are compressed, and surely in the last place in the large and extensive vessels close to the papilla. Also the inclination to recidivism, which we see more in perivascular affections (I am thinking of recurring juvenile haemorrhages of the vitreous humour), and just as in these the apparently benign character of the affection itself as to cure and destruction (for with the latter the haemorrhage is the danger, as we know, not the affection itself) remind us more of the surroundings of a vessel than of the innermost vascular layers. Finally all typical dissemination is also lacking; seldom is there more than one focus in a fresh condition.

* *Arch. of Ophthalm.*, Vol. CVI, p. 4

Notwithstanding all this, in default of any anatomical substratum, in but three of our cases of periphlebitis, this conception is not more than a hypothesis on acceptable grounds. Besides such a periphlebitis may be secondary just as well as primary.

Further it should be kept in mind that the whole lymph-flow through the retina is far from completely known as yet. Aetiologically it may, as it is, be of importance that the affection arises exclusively at a rather young age, at least for the first time. At the first attack the age lies between 13 and 33, perhaps a period of increased susceptibility. In one of our patients the disease probably took place before birth as an inflammation having the same cause, which in this child (patient J), seeing the occurrence of lupus and tuberculosis in the father's family, might have been tuberculous.

There were two pregnant women among our patients, one with dysmenorrhea and an infantile uterus and one with hereditary brachyphalangia on the father's side. Slight indications for tuberculosis were also met with in a few other patients.

P. Sweating, lymphomata, catarrh of the lung apices; uncle died of tuberculosis.

Q. Father tuberculous.

R. Very strong reaction of von Pirquet, mother tuberculous uveitis.

A. Family-anamnesis suspect, von Pirquet plus B.S. 9 mm.

S. Under control of Welfare Centre (thick knees).

D. Swollen hilus-glands, mother pulmonary tuberculosis, father protracted bronchitis.

H. Swelling of the hilus gland and tuberculosis in the mother's family.

V. Much coughing, strong pulmonary markings, calcareous foci.

O. The specialist for internal diseases considered, in view of the clinical picture of the blood, locally weak breathing, rhonchi, intensified pulmonary markings, old primary affections and severe hilus tuberculosis certainly possible.

Remarkable in the case of K is the origin of the change after tonsillectomy and in the case of W after mastoiditis, from which she had a running ear, whereas with three other patients the connection with bad teeth or a diseased mouth-cavity had to be considered (A, E and F).

Summarizing one can say, therefore, that there is a question here of a not very frequently occurring syndrome, at least one not often diagnosed, with a rather favourable prognosis*, characterized by and consequently feared because of a great inclination

* Ormond found as final vision an average of 6/36, Appleman 5/6, van der Hoeve 6/12 f. and the average of the cases cited here is 1/2-5/4; in drawing up a favourable prognosis some reserve should be made in cases of paramacular scotomata and of more serious affection of the anterior segment.

to recidivism, situated round and near the retinal vessels, mostly against or close to the papilla, sometimes attended with slight haemorrhages along these vessels, in which, also in the cicatrix, the vessels show a contracted and dented appearance, a change perhaps occurring slightly more in women, but certainly not seldom in men, preferably manifesting itself in the second and third decennium and the aetiology of which is not known with certainty, but may be different. Primarily the affection is situated in the innermost layer of the retina; in fresh condition it consists of a focus projecting inward into the vitreous humour up to $2\frac{1}{2}$ mm. and perhaps more in other cases of exudative character including strings and turbidities; in part surely also caused by a cyclitis or irido-cyclitis sometimes arising later on, which may, however, also be lacking or at least be very slight. From the primary site the focus also extends outward into the deeper layers of the retina and the choroid. Characteristic is the sector, arch or paramacular limitation of the visual field attributable to destruction of neuro-fibrils. The affection occurs only at the earlier time of life. To finish up with a word about the name. Seeing this disease has as many as four names it seems bold to want to suggest another, which, in that case, might be "retinitis interna circumscripta."

REFERENCES

- GROES-PETERSEN.—*Klin. Monatsbl. f. Augenheilk.*, Vol. L, II, p. 159, 1912.
 VAN DER HOEVE.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LIII, II, p. 487, 1914.
 KÖHNE.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LXV, p. 882, 1920.
 LODBERG.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LVI, p. 301, 1916.
 LOEWENSTEIN.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LXXVI, p. 812, 1926.
 RÖNNE.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LIV, I, p. 455, 1915.
 SCHERTLIN.—*Klin. Monatsbl. f. Augenheilk.*, Vol. LVII, p. 60, 1916.
 EISLER.—*Kurzes Handbuch*, Vol. I, p. 156.
 SCHIECK.—*Kurzes Handbuch*, Vol. V, p. 382 and 546.
 JENSEN.—*Arch. f. Ophthal.*, Vol. LXIX, I, 1908.
 IGRSHEIMER.—*Auge u. Syphilis*, p. 441.
 LEBER.—*Graefe-Saemisch.*, Vol. VII, A, p. 786.
 POSTHUMUS.—*Beschouwingen over retinitis septica*, 1933.
 GILBERT.—*Kurzes Handbuch*, Vol. V, p. 105 and 118.
 VAN DER HOEVE.—45th meeting N.O.G. Ned. Tydschr. v. Geneesk, p. 424, 1914.
 DUKE-ELDER.—*Textbook of Ophthalmology*, Vol. I, p. 108.
 BUSACCA.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXIX, p. 124.
 PAVIA.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXI, p. 474 and 699.
 GRANDI.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXIV, p. 708.
 BENCINI.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXV, p. 523.
 HIGUCHI.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXVI, p. 411.
 TRISTAINO.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXVII, p. 505.
 TRISTAINO.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXVIII, p. 303.
 WOLFF.—*Zentralbl. f. d. ges. Ophthal.*, Vol. XXXIX, p. 333.
 BLESSIG.—*Graefe's Arch.*, Vol. LXXIV, p. 284, 1910.
 FUCHS.—*Graefe's Arch.*, Vol. CVII, p. 15, 1921.
 FLEISCHER.—*Deutsch. Ophthal. ges.*, p. 140, 1922.
 SJAFF and ZEEMAN.—*Graefe's Arch.*, p. 192, 1924.
 SJAFF.—*Vezelverloop in het netvlies en oogzenuw*, Diss., Amsterdam, 1923.
 ZEEMAN.—*Graefe's Arch.*, p. 1, 1921.
 WILMER.—*Atlas fundus oculi*, 1934.
 IGRSHEIMER.—*Arch. f. Ophthal.*, Vol. CI, p. 105.
 VAN DER HOEVE.—*Arch. f. Ophthal.*, Vol. CII, p. 184.