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COMMUNICATIONS

SOLITARY RETINO-CHOROIDITIS WITH REFERENCE TO RETINO-CHOROIDITIS JUXTA- PAPILLARIS—JENSEN

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ALTHOUGH the subject of solitary retino-choroiditis has been considerably discussed, under a variety of different names, on the Continent and in America, it seems to have attracted very little attention in this country and such accounts as are given even in most of the recent text-books are both inadequate and inaccurate. As it is a disease of no great rarity and of considerable interest it was hoped that a survey of the literature and the investigation of a number of cases might serve to clarify our views about the condition and even to throw some light on its aetiology. These ambitions have been imperfectly realised.

History.—The credit for the first recognition of the disease is usually accorded to Jensen, who described four cases in 1908 as constituting a separate type of choroiditis. It seems to have been consistently overlooked that Friedenwald¹ gave a far more comprehensive account of similar cases six years earlier and mentioned that the condition had already been recognised by Griffith, but had up till that time been only imperfectly described. Members of the American Ophthalmological Society retorted that the disease was already well known to them. Pallarés mentions that

the typical field-defects had been recorded by Macnamara in his "Manual" in 1876, but there appears to be little in that work to justify the claim.

Griffith called the disease "choroiditis with Descemetitis." He gave a brief but accurate description of its appearance and apparently confined this group to those lesions in the neighbourhood of the disc. He states that he had drawn attention to this condition eight years before, but it has not proved possible to trace this reference. Friedenwald gave a very full account of his observations on thirty-eight cases seen in the acute stage. He described them as "acute circumscribed exudative chorio-retinitis" and included similar lesions wherever situated in the fundus. He and Griffith both realised the importance of keratic precipitates, a point overlooked by Jensen and many of his successors. Jensen named the disease "retino-choroiditis juxtapapillaris" and restricted its application to those cases in which the lesion is in contact with the disc and produces a sector-shaped defect in the field of vision. Groes-Petersen, in 1912, reported a series of fourteen cases and once more included lesions which were not near the disc and did not produce a sector-defect in the field of vision. He thought that the disease should be called "Jensen's retino-choroiditis," a name which has been generally accepted. In the same year Hepburn¹ attempted to classify all choroiditis into five groups. He admitted that there might be some difficulty in deciding whether any particular case should be considered as "deep localised" or "superficial localised" choroiditis and it appears that most cases of solitary retino-choroiditis would fulfil the criteria for both groups. Two years later he³ described the disease under the title of "cases of deep inflammatory deposits in the choroid." The next year Rönne² went even further and considered that all cases of acute choroiditis are probably of the same type, but divided them into two classes, one in which the lesions are in one or more isolated groups, and the other where the lesion is large and solitary. Van der Hoeve, meanwhile, had re-named the disease "neuro-fibrillitis retinae," a name adhered to by Riehm in 1933. Zeeman in 1921 described one case as "syphilitic neuritis retinae." In 1922 Fuchs reclassified choroiditis into three groups, of which the first included all those cases with single, large, circumscribed lesions.

Friedenwald³ attempted to restore order in 1924, twenty-two years after his first account. In a comprehensive paper he surveyed the literature and gave the results of his observations on a further series of sixty-two cases. He seems to have adopted a very broad basis for the diagnosis of this disease and unfortunately, as none of the cases is described in detail, it is impossible to decide which of the more striking manifestations recorded should

or should not be included here. This paper also has not received the attention it deserves and did not prevent Pallarés, in 1931, from reporting a case as "neurofibrillitis tuberculosa retinae." Finally in 1935 Tristaino brought us back to Jensen by stating that all cases in which the lesion is not touching the disc and does not produce a sector-defect in the temporal field of vision are atypical.

What should the disease be called now? To multiply names increases confusion, but none of those in use is satisfactory. "Juxtapapillary retino-choroiditis" is obviously unsuitable for many cases. "Jensen's disease" is convenient but unfair to Friedenwald, whilst "Friedenwald's disease" would only cause misunderstanding. "Neuro-fibrillitis retinae" is succinct, but may well be based upon unsound pathology. "Acute circumscribed exudative retino-choroiditis" is safe and descriptive but most cumbersome. "Solitary retino-choroiditis" has already been used for this condition, is of convenient length and should not cause confusion; it will be used in this paper.

From the history of the disease will be seen the difficulties which confront one in trying to piece together an orderly account of it. Not only has no agreement been reached on the nature of the condition, but there are even no generally accepted criteria by which it may be diagnosed. For this reason it has not seemed worth while to attempt to make a statistical analysis of the recorded cases; for while some authors have included cases whose claim to belong to this group seems not beyond cavil, others have omitted all cases where the lesion was not at, or near the disc, or where the disease involved the macula, or where there was no sector-defect in the field of vision, thus rendering comparison on these points valueless. Before going on, therefore, to discuss the intricacies of the situation it may be as well to describe briefly what would appear to be understood by all as a typical case of solitary retino-choroiditis.

Typical Case.—An apparently healthy young adult complains that the sight of one eye has been blurred for two or three days. The vision in that eye is found to be reduced to about 6/12. External examination shows nothing amiss. Examination of the fundus shows a vitreous haze which obscures the finer details but close to the disc is seen a patch of shining, cream-coloured exudate shading off gradually into the normal retina on all sides. A week or more later the vision in that eye has deteriorated to about 6/36. Fine keratic precipitates are present and the vitreous haze is thicker than before. The exudate and the rest of the fundus appear unchanged. The condition remains the same for one or two weeks and then steadily improves. In three months the media are clear and vision is restored to normal. The fundus shows a

pigmented scar corresponding to the site of the exudate but is otherwise normal. The field of vision shows an absolute sector-defect extending from the blind-spot to the periphery.

Investigation

A series of twenty-seven cases diagnosed as "Jensen's" or "juxtapapillary" retino-choroiditis has recently been investigated; of these twenty-four seem to have warranted that title and three should probably not be included in this category. No useful purpose would be served by giving a complete account of each case; brief histories of all the cases are appended, only those features which seem of especial interest being described in detail.

The following is a summary of the findings of the twenty-four cases.

Type of Patient.—The patients have shown a remarkable uniformity of type. Not only were they all young and apparently healthy but also, and even more strikingly, they were all intelligent persons. With the exception of the three housewives and the roundsman they were all employed in skilled trades. It is a curious fact that, in a hospital where the patients in general are poor and not very intelligent, the twenty-four investigated with this disease should all have been of the more well-to-do and intellectual class. They were neither undernourished nor unkempt nor likely to harbour gross unattended sepsis. They were, in short, patients in whom one would not expect to find a high incidence of tuberculosis.

General Health.—Without exception the patients stated that their health was in general excellent. The majority were comprehensively examined by a physician, an ear, nose and throat surgeon and a dentist when first seen. All were re-examined this year. A few could associate an attack of the disease with some concurrent indisposition, but the majority could find nothing to account for it. Two patients have been seen five years, two seven years and two ten years since the first known attack, and have developed no other disease; as will be mentioned again it is probable that fourteen of them had already started the disease in childhood.

The family histories disclosed only that the father in two cases and a sister in one case were believed to have died of tuberculosis. In childhood three patients had had discharging ears, one pleurisy and one pneumonia. In their youth two had occasional attacks of tonsillitis, two had had a small crop of boils and one had had gonorrhoea; these diseases were considered to be either cured or in abeyance before the eyes were known to be affected. Tonsillectomy had been performed in four cases, appendicectomy in two

and resection of the nasal septum in two earlier in life. Several had an occasional attack of "flu" which, on closer questioning, usually resolved itself into a severe cold. Seven patients suffered rarely from colds, six frequently and eleven ordinarily. Three were myopes.

When considering the findings in connection with the acute attacks it must be remembered that this is an analysis not of only twenty-four patients but of thirty-five known attacks. In one case (VII) an aural discharge was a most suggestive factor to which two others (I and XVIII) offer a salutary contrast. One patient (VIII) attributed the third attack to "flu" another (XXII) associated the second and third attacks with a cold; another (XXIII) attributed the attack to benzene; one patient (XXIV) felt rather "run down"; one patient was pregnant, one was slightly anaemic and one suffered intermittently from an idiosyncrasy of digestion to certain foods. The aggregate of focal sepsis discovered is no more impressive. The teeth were definitely septic in two cases; in two others the extraction of two and in two the extraction of one tooth was advised. In one case the tonsils were unhealthy; in another they had already been removed in childhood but, as no other focus of infection could be found they were further dissected. In no case was any clinical evidence of tuberculosis discovered.

Tests.—The Wassermann reaction was performed on seventeen patients and was without exception negative. The Mantoux reaction was performed on six cases only and was positive in one. Two patients were given a course of tuberculin injections, which produced a mild local and general reaction in one of them; no change was noticed in the eyes. A blood-count was performed in four cases; two were normal; in one there was slight hypochromic anaemia; in the other out of 8,000 white cells 52 per cent. were lymphocytes. This last patient was fully investigated for tuberculosis without any evidence being found.

The sedimentation-rate of the blood was examined in six acute cases; in five of them it was normal and in one at the upper limit of normality. In Reichel's book on the sedimentation-rate Löffler has summarised the results in diseases of the eye. From this it appears that in conditions localised within the globe the rate is never increased, so that a raised rate indicates a general process secondarily involving the eye. The reliability of the test for indicating the presence of an outside focus of infection does not seem to be high except in marked cases. A normal result in a large number of cases, however, would constitute fair evidence that the disease is confined to the eye. Although this is too small a series to be of any significance it does suggest that the process is neither tuberculous nor septic in origin.

All these investigations have revealed a smaller proportion of disease or sepsis in this series than one would expect to find in average hospital patients. Apart from five patients (VII, VIII, XVIII, XXII and XXIV) even those whose teeth or tonsils were not surgically normal considered themselves in perfect health. In no less than nine patients no history or sign of any malady whatever could be elicited, not even one septic tooth.

Incidence.—Ten of the patients were male, fourteen female. In two cases (X and XIV) the condition was bilateral.

In assessing the age incidence of the known acute attacks one case (VII) introduces a slight difficulty, to overcome which it has been assumed that of six attacks three occurred before the age of twenty and three after. Including these, out of thirty-five attacks 5 occurred under the age of 20 years (VII, VIII and XXIV); 15 from 20 to 24 years; 7 from 25 to 29 years; 8 from 30 to 34 years. Fourteen patients already had an old scar in the fundus when first seen. As these scars and the field-defects differ in no way from those produced by recognised acute attacks, and as three patients remembered having had something wrong with their sight years before, it seems reasonable to suppose that fourteen acute attacks had occurred at an age when they would be likely to pass unnoticed, probably under ten years. The attacks are unlikely to have passed unnoticed in later life in these patients, as they well might have done in others, on account of the general mental alertness already mentioned. To the above age-incidence, therefore, should be added 14 under 10 years. This then constitutes the largest group but one and introduces a curious breach of continuity, but one which it appears that we must accept.

Of the known acute attacks one patient (XI) had had none; 17 had 1 each; 3 had 2 each; 2 had 3 each; 1 had 6. Adding in the fourteen presumed previous attacks, of which ten occurred in patients with only one known attack, gives the very different result that 8 patients had only 1 attack each; 11 had 2 each; 3 had 3 each; 1 had 4; 1 had 6. Even this does not represent the true liability to recurrence, for only two patients were seen after the age of thirty-five, up till which time the chances of another attack are not inconsiderable, and only seven others after the age of thirty.

Only ten patients were seen having what was probably an initial attack; of these one was aged 14 years; one was 16 years; three were 20 years; four were 21, 23, 25 and 26 years respectively; one was 32 years.

In only six cases were the intervals between successive attacks known; one case (II) recurred after 5 years, one case (III) after 3 years, one case (VII) at an average interval of about 1 year, one case (VIII) after 6 years and again after 5 years, one case (X)

FIG. 1. CASE IX.

The appearance in this case is atypical in the irregular outline of the large atrophic scar, in the presence of several small scars, and in the surrounding area of pigmentary disturbance.

For the sake of clarity only the arching part of the "veil" has been represented and several of the smaller vessels have been omitted. The "veil" is shown much more white and opaque than it in fact appeared in order to emphasise the arch of the contained artery.

Three of the larger veins terminating in the scar are clearly seen.

FIG. 2. CASE XVIII.

These scars are typical in appearance and situation, but unusual in the scarcity of pigment. The normal retinal vessels overlying them are well shown. It is probable that each of the two large scars was produced by a separate attack of inflammation whilst the satellite scar occurred simultaneously with one of them. Of especial interest is a comparison of these scars with the associated defect in the field of vision (Fig. 8).

FIG. 3. CASE XXIII.

This scar bears a strong superficial resemblance to a coloboma of the macula, but was undoubtedly inflammatory in origin. It is unusually large, but otherwise typical in appearance. A lightly pigmented rim is present. The preservation of the choroidal vessels is well seen. There is marked temporal pallor of the disc.

after 10 years, one case (XXI) after 1 year and again after 7 years.

The approximate time taken for vision to return to normal is known for fifteen attacks; of these 3 took 1 month; 5 took 2 months; 4 took 3 months; 2 took 6 months; 1 took 7 months. There appears to be no obvious correlation between the features of the attack and the time taken to recover from it.

Situation.—In each recurrent attack the fresh site of inflammation was continuous with or close to the previous scar; in analysing the site of the attack, therefore, it is not necessary to take into account each attack individually. Thus in the twenty-six eyes affected the lesion was touching the disc in 9, less than 1 disc-diameter away but not touching in 5, more than 1 but less than 3 disc-diameters away in 6, and more than 3 disc-diameters away in 6, the two furthest being 6 and 8 disc-diameters away respectively (I and XX). In seven cases there were one or more small outlying or satellite scars or islands of exudate, excluding pigment-cysts seen in two cases (X and XI).

In only two cases (V and XIII) was there a separate site of inflammation comparable in size to the first, and in both of these it was less than one disc-diameter away. In the cases in which successive scars have become confluent it is almost always possible to recognise by their outline the number of scars concerned. By comparing the number of attacks with the number of scars present it seems probable that each attack produces typically only one large scar. Thus one can assess the number of attacks accurately in almost every case provided that one ignores small and outlying scars. It is probable that two large, separate areas of exudate may occur in some cases, but this must be very rare.

Type of Scar.—The amount of pigment present in the scar appears to bear no relation to the degree of destruction, the age of the scar, or the colour of the patient. In one case (XVIII) (Fig. 2) large scars with marked atrophy of the choroid, occurring in a dark man, contained only the faintest trace of pigment; in another case (V) very little pigment was present.

In one case (XV) pigment appeared amongst the exudate fourteen days after the onset and in another (XXIII) in nineteen days. No evidence was found to suggest that the scar, once formed, underwent any further alteration until disturbed by a recurrent attack. Islands of pigment seen in two cases (X and XII) did not resemble the scars of inflammation but looked as though they had been deposited without any disturbance of the surrounding tissues.

One case (IX) (Fig. 1) was atypical in the irregularity of the outline of the scars and in the large surrounding area of change.

Macular Involvement.—The macula was involved in five cases. In only one case (XXIII) (Fig. 3) the macula was the primary

site of inflammation and suffered complete destruction. In every other respect this appeared to be a typical case. In two cases (VII and IX) the macula was involved in the outskirts of the exudate during the acute attacks and in each case vision was reduced to counting fingers. In the latter case vision returned to 6/12; in the former it is probable that it would greatly improve if the condition were to become quiescent. In the other two cases (XIV and XXIV) oedema of the macula at some distance from the lesion was seen and in the latter shining dots, as of an incipient star-figure, were present.

Papillary Changes.—Papillitis is extremely difficult to assess in these cases for the vitreous haze makes the disc appear more ruddy than normal and obscures its outline. Particularly is this the case when the exudate is close to the disc. Of the seven cases seen during an acute attack in which the exudate was touching the disc definite papillitis was present in three (II, V and VIII); the other four were seen some time ago and there is no note on this point. There was papillitis in three cases (IX, XXII and XXIV) in which the exudate was within one disc-diameter of the disc but not touching it, in two cases (XIII and XVII) in which the exudate was between one and three disc-diameters away, and in four cases (I, III, XVI and XX) in which the exudate was more peripheral. Included in these figures are two cases (I and III) in which the changes at the disc appeared to be more in the nature of exudation into or oedema of the adjacent retina than true papillitis.

Thus some change at the disc was present in at least half the cases. The occurrence of papillitis seems to have affected neither the course nor the duration of the attack.

Keratic Precipitates.—Keratic precipitates were observed in every case watched throughout an acute attack but three. These three are probably attributable to the absence not of keratic precipitates but of observation. To be seen in every case they must be looked for frequently and diligently. In two cases they were so fine that it is probable that without the use of a slit-lamp they would have escaped detection. In one case they were described as "mutton-fat." In most cases they were evanescent but in some they lasted for several weeks.

Vitreous Changes.—Vitreous haze, usually intense, has been present in every acute attack. This haze is always thickest in the neighbourhood of the exudate and also often appears particularly dense over the disc. In three cases coarse vitreous opacities or "floaters" were present during the acute attack.

In one case (XXIII) a large vitreous opacity remained permanently in the anterior vitreous.

In eight cases were seen permanent vitreous opacities whose

appearances were quite unlike that just mentioned (XXIII). In each case, although called " opacities " for want of a better term, they were transparent, so that their outline was difficult to determine, and they all appeared to be composed of the same type of material. In seven cases the opacity was tethered to the fundus and in six of these the attachment appeared to be wholly or partly over the disc; in one case (IX) there appeared to be a broad attachment over the scar as well; in one case (XI) the attachment was probably just beyond the scar alone. In one case (XXII) no attachment could be seen. In no case was there an attachment to the lens. Although these opacities showed a considerable uniformity of constitution and disposition they differed markedly in size and shape. In two cases (XIV and XXI) the opacity was small; in the one it looked like a firm, curved handle to the disc, in the other like a short club. At first sight another case (XIX) rather resembled the latter (XXI) in that a knobbed head could be seen in front of the disc; by careful focusing, however, it appeared that this was the apex of a tent-shaped " veil " taking origin from a ring rather less in diameter than the disc and placed eccentrically upon it and the adjacent scar. In two cases (XI and XXII) the opacities were like strands or narrow fronds and in both it was doubtful where or whether they were tethered. In two cases (I and VIII) the opacity resembled a broad frond or veil attached to the disc and waving freely in the vitreous. One case (IX) (Fig. 1) exceeded all the others in size and pattern, the smaller part resembling that in one other case (XIX), although considerably larger, in being apparently tent-shaped, whilst the larger part was as great as the rest put together.

Vascular Changes.—Spasm of one or more vessels during the acute attack was seen in four cases (X, XIII, XIV and XVII). Apart from one case (IX) no changes in the vessels were seen after healing had taken place. " Perivasculitis " associated with spasm occurred in one case (XVII).

Retinal haemorrhages were seen in two cases (VI and XII) and in the latter occurred two months after the onset.

Curious alterations in the circulation were seen in one case (IX) (Fig. 1) which was also unique in that retinal ischaemia was present in the acute attack.

Pain.—Pain occurred with the onset of the acute attack in seven cases (I, V, VII, VIII, XVII, XXII and XXIV). It appeared that if present in one attack it was likely to be present in each successive one. In each case but one there was papillitis; in one case (VII) there was no papillitis but the macula was involved. In three cases (V, VII and XVII) the pain was said to be worse on movement of the eyes.

Headache was present with the acute attack in five cases (X,

XII, IX, XIII and XX); of these papillitis was present in the last three cases.

Thus twelve out of the twenty-four patients had pain or headache with one or more acute attacks, and only three cases with definite papillitis had neither.

Other Appearances.—Tension was increased in the affected eye in three cases (I, XV and XXIII) and in each case returned to normal without special treatment.

Ciliary injection was seen in two cases (V and XXII), in the former appearing first three weeks after the onset. Three other patients stated that their eyes had been "blood-shot" before they came to be examined.

Iritis with synechiae occurred in two cases (IX and XXI) and in each subsided leaving no sequelae.

Definite pallor of the disc was present in one case (XXIII) (Fig. 3).

In two cases (XIII and XVIII) hyaloid remnants were present in the affected eye.

Field of Vision.—It has not seemed desirable to reproduce the fields of vision of all these cases as types similar to all but three (III, XIV right and XVIII) have been illustrated and discussed elsewhere. In this paper a "sector-defect" is taken to mean any defect extending throughout the length of the fibres passing through the lesion, that is to say a nerve-fibre-bundle defect. Thus an arcuate scotoma is included as a sector-defect in that its mode of production is similar, its extent being modified only by the course of the fibres.

Of the twenty-six eyes eighteen showed sector-defects and eight scotomata only. In two cases (V and XVIII) (Figs. 6 and 8) the sector-defect was double; in both cases the sectors had a common origin but in the one they communicated with the blind-spot whereas in the other they did not. This latter type does not appear to have been previously described; it is remarkable both for the shape and for the method of production of the intervening island of retained vision. In one case (XIX) a sector-defect and a separate scotoma were present. In one case (II) the sector was unusually narrow and in one (XIX) broad. Of the twenty sector-defects eleven originated at the blind-spot and nine were separate from it; in three cases (V, IX and XXIV) the sector originated from the blind-spot although there appeared to be an area of normal fundus between the disc and the scar. In seven cases (I, III, IV, XII, XVI, IX and XVIII) the sector was in the nasal field and in all but the last two the apex was medial to the fixation-point; in each of the former cases the nasal extent of the defect was far greater than would be expected from the conventional conception of the distribution of the nerve fibres (Fig. 4).

In one case (III left) (Fig. 5) the sector was relative throughout its entire extent after the first attack and became absolute after the second; this also appears not to have been recorded before.

FIG. 4. CASE I. Test object—5/330 white.

FIG. 5. CASE III. Test object—5/330 white.

In three cases (V, X left and XI) the sectors were relative centrally. Three cases (V, X left and XII) showed considerable diminution in the area of the defect over a long period.

The scotomata were absolute in every case and corresponded in area with the lesion in all but two cases, one (XIV right)

(Fig. 7) where what appeared to be a truncated sector-defect was present, and the other (XV) where the increased area was probably due to oedema during the acute stage. The former (XIV right)

FIG. 6. CASE V. Test object—5/330 white.

FIG. 7. CASE XIV.

Test objects—Periphery 5/330 white—Scotoma 15 1,000 white.

is the only case in this series in which a lesion touching the disc did not produce a sector-defect extending to the periphery. The presence of a peripheral defect corresponding to the direction of the curtailed sector is difficult to understand; the most probable explanation seems to be that there was a small outlying scar in that situation which escaped detection.

FIG. 8. CASE XVIII. Test object - 5/330 white.

The implications of these findings are discussed more fully below.

Excluded Cases.—The three cases A, B and C are described for comparison.

In case A no keratic precipitates were seen in the acute attack. The chief reason for its exclusion, however, is the scar, which differs markedly in two aspects from the typical appearance. First the scar is not discrete and circumscribed, having no definite peripheral limit. Secondly the pattern of the scar does not resemble the characteristic lesion, which will be described in detail below.

In case B neither of the acute attacks was seen but the patient's age was sufficiently unusual to arouse doubt as to their nature. The scar, extending from the disc to the periphery and showing marked sclerosis of the choroidal vessels, is totally unlike anything seen in the other cases in this series, as is also the gross irregularity of the defect in the field of vision. The presence of arterio-sclerosis suggests some different aetiology for the condition.

Case C has little in common with solitary retino-choroiditis except for the age of the patient. The large but indefinite area of change and the gross reduction of vision without any apparent choroidal atrophy are not features of this condition.

It may well be that in the first of these three cases, and possibly even in the second, the condition was produced by a disease process similar to that responsible for solitary retino-choroiditis. Until, however, we have some knowledge of what that process may be it is convenient to distinguish between these cases for reasons which will be given later.

Discussion

Owing to the lack of uniformity in diagnosing this condition it has proved unusually difficult to correlate or reconcile the results obtained in the present series with the findings of previously recorded cases. Many points in the literature have doubtless been overlooked or misunderstood.

Incidence.—Jensen considered the disease rare and Dèrer in 1928 still held that view. Most authorities believe that it is by no means uncommon and Friedenwald¹ stated that it is the commonest form of acute choroiditis. Rönne² thought it common but often misdiagnosed. Where there is an intense vitreous haze and the lesion is peripheral it may easily be overlooked and the condition passed over as "cyclitis," as has certainly happened in some cases and possibly in many.

The sex-incidence appears to be equal. The age-incidence is not easy to determine because so many cases when first seen already have a scar presumably from a previous attack. Friedenwald mentioned this point in his first paper but it has received surprisingly little attention from other authors. One of Jensen's patients remembered having misty vision in childhood and, although this is no proof, it does appear probable that the disease often starts when the patient is too young to be troubled by it. Groes-Petersen saw an acute attack in a patient of thirteen. He also saw acute attacks in patients of forty-four and forty-five, but these were both recurrences, as also was one of Jensen's. Friedenwald¹ mentions an acute attack at fifty but gives no details. Omitting the evidence previously discussed the highest incidence for all recognised acute attacks seems to fall between the ages of twenty and thirty-five and that of recognised first attacks rather earlier.

The type of patient suffering from this disease does not seem to have attracted much attention except that they are usually said to be young and apparently healthy. Friedenwald mentions that most of his cases were seen in private practice. It seems likely that very poor and unhealthy persons are seldom afflicted by it.

Bilateral cases have been recorded by Friedenwald, van der Hoeve, Rönne² and Zentmayer,² but are uncommon. In Rönne's case there was an acute attack in both eyes simultaneously. Fuchs' and Jessop's cases with bilateral involvement of the macula were probably not examples of this disease.

Fundus Appearances.—In the acute stage the characteristic feature of the disease is the patch of shining, cream-coloured exudate standing out above the retina. In several descriptions the exudate has been called "greyish-white"; the explanation of this may be that in some cases after the first few weeks pigment can

be seen hazily through the exudate, giving a patchy grey appearance. Schertlin described green exudate in a most atypical case which should probably not be included in this group. The patch is often round or oval but may have irregular edges in contradistinction to the scar, which has a rounded, regular outline.

The exudate is situated often touching or near to the disc but is quite commonly more than three disc-diameters away and may be much further. In Friedenwald's two series the lesion was close to the disc in twenty-nine cases, near the macula in twenty-nine and peripheral in thirty-eight. There is, however, some doubt as to the type of disease in all these cases, as four are stated to have been very extensive and one from the disc to the periphery. In Groes-Petersen's series nine were close to the disc and six further away, but all within five disc-diameters. Jensen stated that the exudate covers an area about equal to that of the disc and this statement has been copied in several accounts; in fact it often does, but is quite commonly larger and may be four times that size.

The exudate shades off gradually into the surrounding oedematous retina, so that no definite edge can be determined. The retinal vessels may be seen on the surface of the exudate but are frequently entirely obscured. The height of the exudate in these cases is probably never so great as to simulate a tumour. There is typically only one large patch of exudate in each attack, but not infrequently small, outlying islands may be seen at some considerable distance from the main site and according to Riehm these may appear some weeks after the onset. In one of Rönne's cases there were two main patches of almost equal size, but this seems to be a unique example. A recurrent attack usually occurs on the edge of the previous scar but is sometimes separate from it although always in its immediate neighbourhood.

Small retinal haemorrhages are fairly frequent during the acute attack. They may occur in the edges of the exudate or at some little distance away. Köhne states that the haemorrhages may appear some weeks after the onset of the attack, as also seen in this series. Large retinal haemorrhages have only been recorded in cases whose claim to inclusion is most doubtful, such as those of Abraham,² Junius and Loddoni, and probably do not occur in this disease.

Alterations in the calibre of the retinal vessels in the neighbourhood of the inflammation have been recorded by most observers. It is almost impossible to be sure of the details of a vessel actually in the midst of the exudate but it is common to see marked spasm in an artery at one or two places after its emergence. Sometimes vessels not actually involved in the exudate may show spasm for some distance around. The arteries are usually said to be affected,

but Friedenwald stated that the veins often showed spasm too and this has been well illustrated in the present series. The question of whether the changes in the vessels may be permanent has occasioned some controversy. Blessig, Gertz and Zentmayer¹ recorded cases in which spasm was seen after the inflammation had subsided, whilst Fleischer, who believed that the arteries were the seat of the inflammation, said that he had found eight observers who had seen persistent alterations. Abraham² considered that permanent changes in the vessels were present in twenty per cent. of all cases, but this does not seem to have been the general experience. Ham and Schertlin recorded cases in which small retinal vessels entirely vanished during an attack.

The anomalous vessels disappearing into the scar which were seen in one case in this series, appear not to have been previously recorded in this condition. It may well be that the association of this appearance and the preceding retinal ischaemia, both extremely rare, is more than a coincidence. Feingold has recorded three cases of this alteration in the circulation, which he attributes to a retino-choroidal anastomosis. His second case is of especial interest in this connection in that the scar appears to have been a typical product of solitary retino-choroiditis and a tethered vitreous opacity was also present.

Friedenwald¹ saw cases showing fine flecks of exudate bordering the vessels, as occurred in one case in this series, and held them to be a "unique perivasculitis." Schertlin and Löwenstein considered this appearance characteristic of tuberculosis but Fleischer did not.

The papilla is frequently involved in the acute stage. Exudate overlapping on to the disc has been recorded by several authors; Köhne and Statti described cases where the disc was completely covered by exudate whilst in one of Riehm's cases the exudate appeared to be confined to the disc alone. Apart from the presence of exudate or oedema of the adjacent retina the disc itself often shows changes which have been described by most observers and given a variety of names; the term "papillitis" seems most suitably non-committal. Commonly the disc appears slightly swollen and redder than normal and its edges are blurred, whilst there is congestion of the entering veins. The swelling is not usually gross but in Appleman's case amounted to three dioptres and in Statti's case to four or six dioptres and in each closely resembled optic neuritis. As has been mentioned above the appearance of the disc is deceptive in these cases. However, it seems probable that there is papillitis in the majority of cases where the inflammation is juxta-papillary and Tristaino considers it to be typical. What is more interesting is that changes occur at the disc when the exudate is

not touching it, as has been recorded by Riehm, Bencini and others and was clearly seen in the present series even when the exudate was quite peripheral.

The typical scar left after subsidence of the exudate is quite characteristic, but there are occasionally modified forms which are almost unrecognisable. In shape a single scar is round or oval with its long axis usually radial. Small, circular, outlying or satellite scars may be present from the islands of exudate mentioned above. When recurrent attacks produce multiple scars these may remain separate, although close to one another, or coalesce to form a large area in which, nevertheless, the individual scars are still recognisable. From the manner of their formation the outline of these conglomerate scars is always rounded and serpiginous and it is unlikely that such an appearance as that seen, for instance, in Junius' sixth case could be produced by this disease.

Usually the scar is outlined by a rim of heaped retinal pigment. Sometimes this rim is very poorly marked and occasionally is entirely absent. The degree of destruction of the tissues in the floor of the scar varies widely. The whole area may be entirely atrophied so that the floor appears to be composed of sclera alone; even in such cases normal retinal vessels may be seen bridging the gap. There may be no pigment in the floor but apparently unsupported choroidal vessels are seen as a red trellis against the white of the sclera. Attention was drawn to this apparent immunity of the choroidal vessels by Groes-Petersen, and many others have discussed it. There may be choroidal pigment amongst the vessels, looking as though the intervening retina alone has disappeared leaving the normal choroid. Heaping of retinal pigment may be present also within the rim. All these appearances are common and may often occur together in individual or neighbouring scars; perhaps most often there is a gradual increase in destruction from intact choroidal structures at the periphery to complete atrophy in the centre. Rarely there is much less change than this, so that the fundus appears stippled or only slightly greyer than normal, as has been recorded by Rönne,² Lodberg and Statti and in the present series. Disappearance of the exudate leaving no scar at all has been recorded by van der Hoeve, Pallarés and Riehm. Isolated pigment-spots like those seen in two cases in this series have been recorded by Köhne and others. They do not appear to be small scars but heaps of pigment deposited in the normal retina.

Site of Inflammation.—The primary site of the disease has been much debated, one side, represented especially by Friedenwald, Hepburn, Rönne and Löwenstein, maintaining that this is essentially a disease of the choroid, whilst their opponents, notably

van der Hoeve, Fuchs, Fleischer and Riehm, consider that it must start in the retina, van der Hoeve, Riehm and others going so far as to say that the nerve-fibrils themselves are primarily involved.

Unfortunately the only eyes which have been examined pathologically have come from most suspect cases and differ widely in their appearances, so that the final proof is yet to come. Verhoeff¹ described a case which has been much quoted, but at the time he doubted whether it should have been included in this group and subsequently it has been generally agreed that it was an unrelated syphilitic inflammation. Twelve years later he² reported another case which proved to be tuberculous and concluded therefrom that all are of similar origin. Unfortunately this case was not seen in its earliest stages but the facts that it occurred in a woman of sixty-seven, that it caused progressive loss of sight over a period of six months, that the vision was reduced to a doubtful perception of light and that it was finally considered advisable to enucleate the eye seem sufficiently unusual to cast doubt upon the validity of his findings. Four years later Abraham³ published what he claimed to be the first histological report on a case of juxta-papillary retino-choroiditis; on reading the clinical notes one cannot help concluding that this, too, was unjustifiable. There is a history of attacks of blurred vision in either eye, lasting only a half to one day each, for years before the patient was seen; in the final attack there is no mention of keratic precipitates or vitreous haze; there were no scars in the fundus in spite of the presumed previous attacks; the "exudate" appeared as a slate-coloured tumour of irregular outline having an elevation of four dioptries and resembling a sarcoma; vision deteriorated steadily during two months to "hand movements"; the pictures do not resemble those of any other recorded cases. These facts, together with the subsequent course of the other eye seem adequate grounds on which to dispute the diagnosis; it would seem to have more in common with cases described by von Graefe as central relapsing retinitis. The case is, however, of great interest in that the pathological findings have many points of similarity with those of a case described by von Hippel as exudative retinitis, but not considered by Coats to belong to that group. Moreover, both of these cases seem to have resembled clinically the fifth of the six cases recorded by Junius.

Fuchs formulated criteria of retinal inflammation which are frequently fulfilled in this disease. That the retina is much involved in the acute stage is shown by the superficial exudate, which always appears to cover a considerably greater area than the resulting scar, by the surrounding oedema of the retina, by the involvement of the retinal vessels, by the presence of retinal

haemorrhages and by the damage to the nerve-fibres as shown by the sector-defects. On the other hand it has been argued that the presence of keratic precipitates, the extensive destruction of the choroid and the fact that often scotomata only are found in the field of vision point to a choroidal lesion. The case for the inflammation being primarily in the nerve-fibre layer seems to rest on the frequent involvement of the papilla, especially in that exudate may appear upon it, on the scar being sometimes minimal or absent, that is to say, without causing any disturbance of pigment, and yet producing a sector-defect, and on the statement of Pallarés that in one case he could see that the inflammation was confined to the innermost layer of the retina. On the other hand there is no doubt that severe choroidal destruction may be present and the field of vision show only a scotoma.

It seems that the only alternative explanations by which these facts may be reconciled are either that the disease may start sometimes in the choroid and sometimes in the retina, or that the inflammation lies primarily in the deeper layers of the retina, but above the pigment, and produces the greater destruction sometimes above and sometimes below it; of these the latter seems by far the more probable solution.

Field of Vision.—The sector-shaped defects of the visual field commonly produced by an attack of this disease have been the subject of much discussion. Jensen, and Friedenwald¹ before him, Ormond, Ham and Zentmayer considered that the defect was due to ischaemia produced by narrowing of a retinal vessel. Hepburn² believed that thrombosis of the choroidal vessels was the cause. Groes-Petersen first suggested that the sector was produced by destruction of all the nerve-fibres over the site of the inflammation and his paper contained charts of many different types of sector-defect which upheld this view. Rönne agreed with this and he and van der Hoeve worked the matter out very fully, tracing the course of the nerve-fibres from the sectors produced. Subsequently it has been generally recognised that the nerve-fibres and not the vessels are at fault and Friedenwald was convinced of the correctness of this view. Groes-Petersen admitted that it was somewhat surprising that pallor of the disc could not be seen even when a large sector was deficient but supposed that this was due to the comparatively small number of fibres affected. Friedenwald states that he saw pallor of the disc in several cases and there was a most striking example in this series (Fig. 3).

Jensen laid down that the sector-defect was a diagnostic necessity for this disease and a few other authorities up to Tristano have adhered to this dictum. Friedenwald pointed out in his first paper that there appeared to be no essential difference between

those cases which produced a sector-defect and those which left only a scotoma. In his later paper he quoted a case in which there was a sector-defect in one eye and a scotoma in the other, and another in which the defect began as a scotoma and later developed into a sector. These facts, together with the evidence of Blessig and Abraham¹ that a sector-defect and scotoma may co-exist in the same eye, seem to be convincing evidence that no distinction can be made on these grounds.

The relative frequency of sector-defects and scotoma is difficult to assess on account of the discrimination which has been exercised against the latter. Friedenwald found the sector-defect comparatively rare; Groes-Petersen found it more than twice as common as a scotoma, with which observation the present series agrees. Sector-defects have been described in every part of the visual field starting from the blind-spot or peripherally; an extremely narrow one has been recorded by Fischer and a very large one by Groes-Petersen and Ormond, the former including more than half the field of vision. Groes-Petersen described a case in which two separate sector-defects were present in the same eye, each being produced by a separate attack of the disease and in the present series a double sector was seen in two cases (Figs. 6 and 8). In the second of these cases it is difficult to understand how the island between the two sectors has survived. The two scars (Fig. 2) which presumably cause the sectors are confluent and it would seem unlikely that any of the overlying retina had escaped destruction. The shape of the nasal defect and of the island is also unexpected.

Traquair has described in detail the probable course of the fibre-bundles, derived largely from a study of these sector-defects. Whilst, however, the defects originating in the blind-spot accord accurately with this conventional conception, those starting more peripherally may not. In the present series five cases of peripheral sector-defects in the nasal field showed a medial extent not to be explained by this distribution (Figs. 4 and 8) and such fields have been recorded by Groes-Petersen and others so that there can be no doubt of the accuracy of the observations. In one case in this series a sector-defect in the nasal field had its apex medial to the fixation-point and extended equally on both sides of the horizontal meridian (Fig. 5). There appears to be no description elsewhere of such a defect and it is difficult to understand how it may be reconciled with the accepted theories, by which such a lesion should produce only a local scotoma. It is clear that we do not yet fully understand the arrangement of the fibre-bundles.

Jensen stated that the visual defect was absolute and permanent and many authors have quoted this statement with agreement.

Friedenwald observed in his first paper that the defects diminish with healing and, at any rate in the case of scotomata, this is so much to be expected that it is curious that it has not more often been observed. During the acute stage the vision is impaired locally not only by actual destruction but by the exudate, which covers an area greater than that of the subsequent scar, and by surrounding oedema. If, therefore, the final scotoma is to correspond in extent with the size of the scar, as is usually the case, it is clear that it must previously have been larger. Similarly a sector may show a considerably greater extent during the acute stage not only at its apex, which corresponds to the local oedema, but in its breadth, due presumably to oedema of the fibres passing through the outskirts of the inflammatory area. Also there may be general contraction of the peripheral field during the acute attack when papillitis is present. Blessig first drew attention to this regression of the defect only two years after Jensen's paper. Zeeman's case is most interesting in this respect and Pallarés shows the resolution of an arcuate scotoma, which is in effect a sector-defect, to leave only a very small circular scotoma corresponding to the scar. Blessig's case suggests that this improvement may continue steadily for some time and this is also indicated in the present series.

Van der Hoeve and Rönne² described relative sector-defects and Bencini one in which a part was relative and a part absolute. It seems possible that the borders of a sector-defect might often be found to be relative if carefully investigated and the fact that some may be relative towards the apex in particular, suggests that this may be the result of a gradual diminution in size taking place for some considerable time after the acute attack.

As Traquair has pointed out we do not know whether the most superficial fibres in the retina are distributed to the central field or to the periphery. Those cases in which the defect is greater towards the periphery and in which improvement occurs from within outwards would seem to support the former alternative, in that the deeper fibres are more severely affected, if we believe that the inflammatory process attacks from the deeper layers of the retina. This view, if taken to its logical conclusion, leads us to expect that such a scar might produce a peripheral defect separated by an area of normal perception from the scotoma corresponding to the lesion. Such a defect does not appear to have been described. In direct contrast to this view one case in this series showed a "proximal sector-defect," in that the scotoma greatly exceeded the area of the scar but fell far short of the periphery, which, however, showed a corresponding indentation of debatable origin (Fig. 7). If we assume that this peripheral defect was produced by a separate lesion the proximal "truncated

sector " appears to be a bundle-defect involving only part of the thickness of the fibre-layer, but leads to the opposite conclusion to the preceding. The latter alternative, that the superficial fibres supply the periphery, seems in general to be the more probable.

Course.—The disease is usually announced by the patient complaining of misty vision and at this stage a vitreous haze and the patch of exudate are already present. If the onset is associated with pain this is usually the first symptom and may precede the mistiness by a day or two. In one case in the present series the patient was seen in this condition when the fundus showed only papillitis. Unfortunately, it was not re-examined until a week later, by which time the exudate was fully developed. In Appleman's case the exudate was present when first seen, but the vitreous haze did not appear until a week after the onset so that the mistiness of vision was presumably due to the papillitis. Schertlin and Loddoni have recorded cases in which they watched the exudate appear, but both in somewhat unusual circumstances.

During the first week or more the sight usually becomes rather worse, probably due to increasing vitreous haze and oedema. The degree of deterioration of vision seems to depend mainly upon the nearness of the inflammation to the papillo-macular bundle. The keratic precipitates may be present at the onset but are commonly not seen until after the first week and may not appear for two or three weeks; they are usually transient but may last occasionally for several weeks. There are few data about the earliest appearance of pigment amongst the exudate. Usually as the exudate disappears it reveals pigment lying in the base of the scar. Riehm records a case in which pigment was visible twenty-six days after the onset and in two cases in this series it was seen on the fourteenth and nineteenth days.

All subjective symptoms have usually subsided before the exudate has entirely disappeared. Vision returns to normal commonly in two or three months; several cases which have recovered in one month have been recorded and a few taking six months or longer. By the time the media are clear the scar is usually plainly visible with a slight haze of exudate still upon it, which clears completely in a further few weeks. Once the condition has subsided the scar probably does not atrophy further but remains unchanged until disturbed by a further attack.

The incidence of recurrence is not easy to assess as many patients are not watched until the likelihood of further attacks is past. Friedenwald stated that only nineteen out of sixty-two of his patients had recurrences but this is almost certainly an understatement. It seems probable that recurrences occur in over half the cases.

Keratic Precipitates.—Jensen did not observe keratic precipitates

in any of his four cases and it is remarkable in how many recorded cases no mention of them has been made. Griffith stated originally that they were constantly present in the acute stage; Friedenwald agreed with him and added that if keratic precipitates were not seen it was either because the diagnosis was wrong, or because they were looked for too late, or because the attack was too mild. The "mildness" of an attack is difficult to assess; Riehm described three cases which he considered to be very mild and in which no keratic precipitates were observed, which seems to support this contention. Griffith added that they are dust-like and that they fade quickly, and these are probably the real reasons why they are overlooked. As mentioned above they may occur at the beginning of the attack or some weeks later and are often ephemeral so that unless frequent observations are made with a slit-lamp they may well be missed. It has seemed that they are sometimes finer and much more widely distributed than the precipitates usually associated with cyclitis, in fact in one case they were almost evenly spread over the whole cornea, as though there might be some essential difference between the two varieties; owing to insufficient experience this observation cannot, unfortunately, be of much value. Friedenwald found that they are fine early but later may become thick, whilst Hepburn states that they occur simultaneously with an increase in the vitreous haze, but these facts do not appear to have been confirmed.

The origin of these precipitates does not appear to have aroused much speculation. Griffith believed that they are formed in the choroid and Friedenwald agreed that they are not due to a cyclitis. Hepburn¹ stated that the ciliary body is always involved in deep, circumscribed choroiditis, but his evidence appears to have been only the presence of keratic precipitates. Most authorities seem to believe that they are due to a concurrent "quiet" cyclitis on the ground that any disease of the uveal tract is likely to imply a generalised uveitis. This assumption does not seem particularly justifiable in this condition. Iritis is uncommon. One might expect ciliary injection in many cases if there were really a generalised uveitis, but in fact it is not usual. Moreover the choroidal inflammation is fulminant and if the ciliary body were to be involved in a similar process it would scarcely be likely to be "quiet." Above all it seems most improbable that a generalised uveitis should involve the ciliary body at a considerable distance from the primary inflammation whilst a large area of intervening choroid remained apparently undisturbed.

Vitreous Opacities.—Some vitreous haze is always present during the acute attack. Usually it becomes more dense during the first few weeks and may form visible, discrete opacities and then gradually clears, seldom leaving any residue.

As with the keratic precipitates neither the composition nor the origin of this haze has been determined but it is usually assumed that both are of similar production, to which Hepburn's observation has lent colour, and that both are probably derived from the ciliary body. It seems more logical to suppose that both are in reality derived directly from the exudate. The probability that the primary site of inflammation lies in the retina has been discussed above. Those cases in which characteristic exudation occurs and yet on resolution no disturbance of the retinal pigment is seen suggest that the exudate is formed entirely in the retina, and, although we have no knowledge of the histological findings in a typical case of this disease, the reports of cases with a similar exudate, especially that of Hancock, suggest that this is a reasonable view and that leucocytes of one sort or another play a large part in the process. If there are great numbers of leucocytes heaped upon the retina it seems most likely that they should wander into the vitreous and thence into the anterior chamber. The observation that the haze appears thickest over the lesion and the more or less related waxing and waning of the exudate, the haze and the precipitates provide some circumstantial evidence in favour of this view.

The permanent vitreous "opacities," taking the form usually of strands or veils, which were present in no less than eight cases in the present series, were mentioned by Groes-Petersen and since then by Abraham, Statti and others. They are, of course, also seen in other forms of choroiditis, but their formation does not seem to have aroused much discussion. Knapp has recently described three outstanding cases, of which the one illustrated appears to have been an example of solitary retino-choroiditis. The variable appearances which they may present have been described above. There seem to be at least five possible explanations of their occurrence. They might be produced by organisation of the exudate or vitreous opacities, they might be fibrinous strands, a form of retinitis proliferans, vitreous degeneration or detachment of the vitreous. "Organisation" is a vague term which is usually taken to imply the production of fibrous tissue. Such organisation is accompanied by a considerable process of vascularisation and proliferation which appears never to have been observed in this condition. Developed fibrous tissue has a white or grey glistening appearance well seen in congenital remnants on the papilla and quite unlike the pale yellow transparency of these "opacities." Moreover fibrous tissue attached to the retina commonly leads to a detachment, which seldom, if ever, occurs in this disease. These considerations make fibrous organisation or retinitis proliferans improbable. On the other hand free fibrin or a fibrinous organisation might produce such an appearance,

and strands in the anterior chamber not unlike the shorter ones seen on the disc are attributed to it, but we have no evidence that this may occur in the vitreous. That vitreous degeneration may produce such changes is possible, but owing to the difficulty of studying its structure we cannot tell what alterations it may undergo. Detachment of the vitreous has been recently described in great detail by Rieger, who concludes that it is usually associated with choroidal changes and may follow choroiditis. It was unfortunately not possible in this series to use the special methods necessary for its diagnosis but many of the pictures in that paper are extremely like the appearances seen in the present series and one of Rieger's cases shows a scar similar to those seen in this condition. In any case it is a suggestion which deserves further consideration. It is possible that the short, thick strands and the diaphanous veils are not produced by the same process.

A case of unusual interest in the present series is that in which a branch of the central retinal artery was seen arching forward into the vitreous apparently in the edge of a "veil" (Fig. 1). It seems inexplicable that this large vessel should really be free in the vitreous and much more likely that it is included in a fold of retina obscured by the "veil." This would then seem to resemble cases described as congenital anomalies more closely than those of the results of inflammation. The simultaneous presence in the same situation of two such unusual conditions, however, appears a most improbable coincidence.

Complications.—Macular involvement in various forms is not uncommon. For the macula to be the focus of inflammation and suffer permanent destruction is very rare; it has been recorded by Rönne,² Friedenwald² and once in the present series (Fig. 3). Involvement of the macula in the outskirts of the exudate has been described by Friedenwald,¹ Rönne, van der Hoeve, Zeeman, Fleischer, Bencini and others. In all these cases there was gross reduction of vision, usually to counting fingers, in the acute attack and usually complete recovery later; in Zeeman's case vision was only partially restored. Oedema of the macula when the lesion was some way away has been recorded by several authors. A star-figure at the macula in similar circumstances was observed by Ormond, Lodberg, Fuchs, Friedenwald and Statti; it does not appear to affect the prognosis.

Pain in the affected eye at the onset of the acute attack is fairly frequent. It may occur with the mistiness of vision or precede it by some days. It appears to be of two types, a constant dull ache, often referred to the orbital walls, or a sharper pain on movements of the eye. The two types may occur together; both appear usually to be associated with papillitis.

Headache may accompany the onset and be severe. It is often intermittent and may last for several weeks.

Ophthalmic migraine in association with the acute attack has been described by Friedenwald² and Blessig.

Increased tension in the affected eye during the acute attack is recorded by Groes-Petersen and Zentmayer amongst others. This complication has occasionally necessitated interference but usually recovers without especial treatment.

Ciliary injection is unusual but has been observed in some cases. In two, described by Groes-Petersen, it occurred with iritis, but this association is by no means constant.

Iritis occurs occasionally. It appears usually to be mild and to recover completely.

All the above complications have been mentioned by Friedenwald² and in addition scleritis, cataract, optic atrophy, detachment of the retina, diminished tension and phthisis bulbi, which do not appear to have been recorded elsewhere, and it is doubtful whether such severe sequelae occur in cases strictly belonging to this group.

Diagnosis.—In the absence of any positive knowledge of the aetiology of this condition it is questionable whether there is much to be gained by trying artificially to segregate this type of case from all others. For the sake of prognosis alone, however, the attempt seems not only justifiable but desirable.

Jensen's original concept was undoubtedly too narrow and has been almost universally abandoned. Friedenwald's was, in contrast, too broad if we are to accept prognosis as the measure of success. Hepburn³ made by far the best attempt to tabulate the features of the disease, but his classification, as he himself admitted, was not entirely satisfactory and presented few features of diagnostic value. Most authors have evaded this unattractive task.

Before venturing on such uncertain ground it will be well to consider which other conditions most nearly resemble this disease, for, in the present state of our ignorance we can only hope to distinguish them on an entirely empirical basis. In this respect the six cases described by Junius are particularly instructive, especially those three which he termed "parapapillary"; his fourth case gives much cause for thought and the two following have many points of similarity. Then the recorded cases of solitary retino-choroiditis which have been regarded above with suspicion must be discounted on good grounds to justify such high-handed treatment. Lastly the disease must be distinguished from tuberculosis. How firmly intermixed the two have become is well shown by three cases described by Hancock, Clement and Bywater as presumably tuberculosis. Although there is much similarity between these cases the first certainly was not, the third

possibly was and the second probably was a case of solitary retino-choroiditis, and yet this alternative appears to have been overlooked. From a consideration of such cases the following points emerge which may serve, for want of better, as criteria for diagnosis.

The disease occurs in young people and may start in childhood. Any acute attack in a patient over forty-five or any first attack in a patient over thirty-five makes the diagnosis doubtful. The acute stage is characterised by the presence of the exudate. The patient may be seen before the exudate has appeared, in which case papillitis alone may be present and the immediate diagnosis almost impossible. The exudate is creamy white or, later, greyish. Exudates of other colours do not occur. The patch seldom exceeds three times the area of the disc and is not so heaped as to simulate a tumour. If there is a scar already present the exudate will be upon or near it. Although there is presumably a time during which the exudate appears and spreads, after the first fortnight the patch does not increase in size. Exudates which steadily encroach upon the surrounding retina should be excluded. Two separate large patches very rarely occur and more than two never. There may be small, outlying islands of exudate but apart from these, papillitis, oedema and vascular changes the rest of the fundus is normal. Small haemorrhages may be present but large haemorrhages do not occur. Vessels do not become thrombosed. The vision may be reduced to 6/36 but not lower unless the papillo-macular bundle is involved in the exudate or by oedema. Keratic precipitates are almost always present and their absence after prolonged searching makes the diagnosis doubtful. Pigmented precipitates are suspect. A vitreous haze is always present during the acute stage. Any of the complications mentioned above may occur. Severe iritis should arouse suspicion. The condition is not progressive. The vitreous haze may increase for some weeks but after the first month gradual amelioration occurs. Increasing loss of vision or general deterioration after this time is not a feature of the disease. The field of vision may show one or more sector-defects or scotomata and there may be general peripheral contraction.

In the quiescent stage the typical "clean" scar described above is present, or rarely a much less definite one. There may be small outlying scars and occasionally two large separate ones. The scars are sharply defined and round, oval or serpiginous in outline. Choroidal vessels, if visible, are not sclerosed. The scar, once formed, does not alter in appearance without an exacerbation. Isolated spots of pigment may be present but apart from these and perhaps localised vascular changes the rest of the fundus is normal. The field of vision may show one or more absolute or

relative sector-defects or scotomata. The defect may slowly diminish in area but the outline is always regular. Vision returns to normal unless the macula is involved in the scar.

Prognosis.—The acute attack may take from one month to a year to subside, usually two or three months. There appear to be no clear indications in the early stages of how long this period may be. Increased tension may rarely become serious but the other complications lead to no permanent damage, if Friedenwald's cases be excluded. Friedenwald stated that iritis or optic neuritis make the prognosis worse; these observations do not appear to have been confirmed, nor does the contention appear in general to be true. Vision always returns ultimately to normal unless the macula has been involved in the inflammation; there seems to be no record of any case where central vision was destroyed by a lesion of the papillo-macular bundle not extending to the macula. The defect in the field of vision may be of almost any extent but very seldom interferes subjectively with the patient's vision. The patient's consciousness of the "blind spot" appears to depend neither upon the size of the defect nor upon its relation to the fixation-point but upon the mentality of the patient. The likelihood of a recurrence cannot be estimated. The majority of patients have two or more attacks but the first recognised attack may often be the last. After the age of thirty-five attacks are uncommon and after the age of forty very rare.

Treatment.—Without knowledge of the aetiology rational treatment is not possible. Atropine is commonly instilled and may well be beneficial although the indication for its use is not very clear unless iritis be present. It does not appear to relieve pain or headache. Fischer allowed a typical case to take its natural course, using only one drop of atropine for examination of the fundus; vision returned to normal within a month. It seems undesirable to continue atropinisation whilst the condition is subsiding so that vision remains impaired by cycloplegia longer than by the disease. Increased tension should be treated on the usual lines.

There is no good evidence that tuberculin injections ameliorate or shorten the course of the attack. On general grounds it is obviously desirable to treat any constitutional disorder or focus of sepsis, if such can be found, although it cannot be shown that this has any specific effect upon the course of the attack or the liability to recurrence.

Aetiology.—Although usually accorded a higher place in the consideration of any disease it has seemed convenient in this instance to reserve the discussion of the aetiology until the end, that the evidence may be examined in the light of what has gone before. A large number of conditions have been described occurring coincidently with an attack of this disease and therefore

credited with its causation. Amongst such are trauma, chill, pregnancy, parturition, influenza, pleurisy, erysipelas, intestinal infection and colds. Loddoni's case is remarkable in that the exudate appeared in a patient with a fractured skull and consequent gross disturbance of the fundus and was attributed to latent tuberculosis.

It is a rational point of view to maintain, as did Rönne, that all acute choroiditis forms one single group, or that there are but five main groups, of which this condition is a subdivision, as held by Hepburn. If, however, one believes that these cases constitute a clinical entity, as many authors do, it seems illogical to suppose that it may be produced by a variety of entirely dissimilar disease processes. For this reason the position of Köhne Löwenstein, Gilbert, Bencini and others who hold that either tubercle or syphilis may cause such an inflammation seems entirely untenable.

Dérer recorded one case with syphilis and Igersheimer described as "syphilitic papillo-retinitis" what was probably an example of this disease, but the association is so rare and the Wassermann reaction has so often been found negative that syphilis can no longer be considered seriously in the aetiology.

Tuberculosis stands on an entirely different footing. It is so commonly present amongst the adult population of civilised countries that it would be surprising to find that less than three quarters of all adults with choroiditis had been infected at some time with the tubercle bacillus. For this reason it is generally believed on the Continent that almost all ocular inflammations of unknown origin are directly or indirectly attributable to tuberculosis. Thus Schertlin, Pallarés, Riehm, Tristano and Bossalino with the majority of Continental authors and Verhoeff² state that this condition is a manifestation of tuberculosis, and several others have agreed that it is a frequent if not the sole cause. Before this opinion became popular Groes-Petersen and Rönne had recorded that they were unable to find any evidence of tuberculosis in the majority of these cases and Friedenwald agreed with them. Certainly the age-incidence, which is one of the stumbling-blocks in the aetiology of this condition, accords well with that of tuberculous infection. On the other hand the distribution of the two in the population seems entirely incompatible.

The appearances during the acute attack in this disease are not the same as those of solitary or miliary tuberculosis of the choroid, the iritis which occasionally accompanies it is totally unlike a known tuberculous iritis and the prognosis for both is consistently much better. For this condition to occur in association with manifest tuberculosis elsewhere in the body is rare; it has been

recorded by Hepburn³ and a case described by Leber as tuberculosis of the retina should probably belong here. To overcome these difficulties it has been explained that the condition occurs in people who have developed a high immunity to the tuberculous process so that a fresh infection may well produce a picture entirely different from that which we are accustomed to expect. In this case one would be entitled to imagine that every patient would show a marked reaction to tuberculin, but whereas many have done, as would any average sample of the adult population, a surprisingly large number have been entirely negative to such injections, and this might appear to be an almost insuperable objection to the hypothesis, although Woods and others deny this contention. Again, if such were the case, one might confidently expect that a series of tuberculin injections would produce not only a marked general reaction but some definite manifestation in the affected eye. In fact, in spite of the large number of patients who have been subjected to such treatment, no definite reaction in the eye has ever, apparently, been recorded, those of Schertlin and Riehm, and a case of Bywater's which might possibly be included here, being open to considerable criticism.

The other evidence for tuberculosis seems to rest upon the occasional presence of perivascularitis, the tuberculous origin of which is itself debatable, and upon the effects of tuberculin treatment. Striking claims for the latter have been made by Pallarés and Riehm among many. It is well known that injections of tuberculin or other foreign protein may favourably affect the course of non-tuberculous diseases. Even so, equally dramatic recoveries have occurred in this condition without any such treatment and the frequent use of tuberculin in other cases has not produced such remarkable results. On the evidence of a large number of cases it does not appear that the exhibition of tuberculin in general influences the course of the disease for better or for worse. Rönne goes so far as to state that all recorded cases of tuberculous choroiditis cured by tuberculin injections were probably in reality cases of solitary retino-choroiditis.

Focal sepsis rests on yet more treacherous ground. On account of the dominating tuberculous influence on the Continent teeth and tonsils have received little attention there. Groes-Petersen, who did not believe in the tuberculous aetiology, mentioned that a focus of sepsis could often not be found, and this, in a population whose adults commonly harbour some culpable site, is a serious objection. Moreover the general prevalence of focal sepsis occurs at a later age than does the incidence of this disease. Ormond, Hepburn, Zentmayer and Friedenwald have attributed attacks to infections in various parts of the body but the evidence is not impressive. If teeth and tonsils were a common cause of

the disease one might expect that thorough attention to them would safeguard the patient from an attack and prevent recurrences. It appears to do neither. To circumvent the latter difficulty it has been suggested that once the primary inflammation has occurred and settled, the process may not become extinguished but lie dormant, ready to be rekindled by the next trivial, perhaps unnoticed infection which the patient incurs; in which case eradication of the primary focus of sepsis would not prevent recurrences. The objections to this theory are twofold. In the first place if some sub-clinical infection could produce a repullulation surely one would expect a cold or influenza or other recognised febrile disturbance to do so with much more certainty, whereas in fact exacerbations usually occur apart from such maladies. Secondly one would not be surprised if eradication of the primary focus by tonsillectomy or extraction of teeth were to produce some reaction in the eye; this would indeed constitute good evidence for the contention. It appears never to have been recorded. The very healthiness of the vast majority of these patients has been attested to by almost all who have written about this condition.

In the present state of our knowledge, however, any attempt to suggest a cause which accords with the known facts involves a further step in the descent to the abyss of conjecture. The frequent and severe involvement of the vessels has led many to suppose that the condition is primarily vascular. The disease does not appear to resemble or often to be associated with any known vascular affection. One case has been seen which subsequently developed Raynaud's syndrome but confirmatory evidence of such sequelae is entirely lacking. The recovery of the vessels after so severe an inflammation is rather against such a hypothesis.

The prominent and perhaps primary part played by the retina in the inflammation has led to the suggestion that this is a disease of the nervous system. The frequent involvement of the disc, shown either by exudation, papillitis or surrounding oedema, strongly supports this view, whilst changes at the macula, often at some distance from the primary site of inflammation, seem to indicate that this may be no localised embolic phenomenon. Groes-Petersen originally mentioned the possibility of a specific virus infection, which has occasionally been echoed but with little enthusiasm. It seems certainly to be open to no more objections than other theories. Fleischer believed that this condition is a manifestation of disseminated sclerosis or some allied process; he had often seen similar lesions at the periphery of the fundus in patients with that malady and also symptoms suggestive of it in patients with solitary retino-choroiditis. Another case has been

seen in which the condition occurred in a patient with definite disseminated sclerosis, but this association must be very rare and proves nothing. Certainly the age incidence shows an unusual similarity, but disseminated sclerosis is more than twice as common in women as in men and steadily progresses to complete destruction.

Migraine is another obscure recurrent affliction of the nervous system which may start in childhood or adolescence and commonly ceases after middle life. The occurrence of headaches in acute attacks of solitary retino-choroiditis and the association of migraine in Friedenwald's and Blessig's cases are superficially attractive. Again the sex incidence is at variance and migraine is often familial. In any case our ignorance of the pathology of either disease makes any comparison between the two futile.

That the condition may be an allergic phenomenon, again one of the few processes with which the age-group can be correlated, is a possibility with, as yet, no reasonable support.

Case Reports

CASE I.—A motor mechanic, first seen in June, 1933, aged 28 years, complaining that his right eye was misty and ached. He was well at the time and maintained that he had always had good health. Closer enquiry elicited the fact that he had had a discharge from his left ear for ten or twelve years but had no treatment for it as it never bothered him. The discharge had ceased altogether about a month before the eye became affected.

On examination the tension was raised in the right eye. The vitreous was hazy but a large patch of "old choroiditis" was seen in the upper temporal region of the fundus. Four days later the tension was still raised and now a patch of fresh exudate was seen on the lower half of the disc and extending downwards between the inferior nasal and temporal branches of the central retinal vessels, which it partly obscured. Vision was fully recovered within three months. Since that time his left ear has discharged occasionally for a short period when he has had a cold. He has had no further trouble with his eye.

The patient was re-examined recently. In the centre of the disc is a small, indefinite, transparent mass from which a narrow "veil" extends forwards and upwards into the vitreous. About six disc-diameters up and out from the disc is a typical, bilobed, oval scar about three disc-diameters in length. The rest of the fundus appears normal. The field of vision shows an absolute sector-defect in the lower nasal quadrant, having its apex 25° from the fixation-point and extending to the periphery. The blind-spot is normal in size and shape (Fig. 4).

From the double scar now present there can be little doubt that during the acute attack there was renewed activity at the site of the old lesion. The changes at the disc appear to have been quite unlike the usual papillitis, but also unlike a primary site of exudation in the absence of any scar.

CASE II.—A photographer, first seen in June, 1936, aged 32 years, having a typical acute attack in the left eye. The fundus showed the scar of a presumed previous attack. He has had another similar attack in February, 1938. In March, 1938, the left fundus showed two typical, contiguous scars less than one disc-diameter up and out from the disc. At the lower end of the scar was a patch of fresh inflammation extending along the normal superior temporal branch of the central retinal artery on to the disc. The field of vision showed an absolute sector-defect extending from the blind-spot outwards and downwards; it was unusually narrow, being about 15° in breadth.

CASE III.—A typist, first seen in January, 1935, aged 32 years, complaining that the sight of the left eye had been blurred for a week. Examination at that time showed "mutton-fat" keratic precipitates. Below the disc was seen a hazy area of retinal oedema extending between the inferior nasal and temporal branches of the central retinal artery. There are unfortunately no further notes of the patient's condition at that time as she did not return again but was treated by her own doctor.

The patient was re-examined in March this year. The left fundus appeared normal apart from a typical large oval scar about three disc-diameters in length and situated with its proximal border about three disc-diameters lateral to the macula. The field of vision showed a full peripheral extent with a 5/330 white object, but with a 1/330 object there appeared a sector-defect extending nasally with its apex 20° from the fixation-point. The blind-spot was normal in size and shape.

A week after the latter examination the patient had a recurrent attack in the same eye. She stated that on this occasion the sight was much more misty than during the previous attack. She was not seen until six weeks later, when the condition was settling down. At that time there was considerable vitreous haze over the centre of the fundus but the periphery was almost clear and appeared normal. A large patch of exudate could be seen on the upper and outer border of the old scar. The disc appeared indistinct and red through the haze, but its outline was clear and the veins were not engorged; adjacent to the disc, however, but this time between the superior nasal and temporal branches of the central retinal artery, was a markedly paler area of fundus due, presumably, to oedema. The vessels appeared normal.

The field of vision showed a sector-defect exactly similar in extent to that previously described, but now absolute (Fig. 5).

There can be little doubt that the peripheral lesion was present during the first attack either as a scar or in active inflammation. With each attack there was some disturbance at the disc unlike the usual papillitis and separated by a wide area of normal fundus from the peripheral lesion.

CASE IV.—A male checker, first seen in June, 1936, aged 20 years, having an acute attack in the left eye. There is now a typical scar about two disc-diameters in extent situated with its proximal border about two disc-diameters out and down from the macula. The field of vision shows an absolute sector-defect in the upper nasal quadrant having its apex 15° from the fixation-point.

CASE V.—A chemist's assistant, aged 20 years, first seen in January, 1938, having an acute attack in the right eye. He remembers having had a "blind patch" in his right eye for many years and thinks that this eye was treated for some complaint when he was about ten years old. The recent attack started with aching made worse on movement of the eye and was followed by blurring of vision a week later. Three weeks later there was some ciliary injection and keratic precipitates appeared for the first time. The fundus showed marked papillitis and three separate lesions. Situated at the upper margin of the disc and slightly towards the outer side was an old, hemispherical, atrophic scar, pigmented at its upper border, giving the disc the appearance of being jauntily "crowned." Immediately below the disc was a similar "crown" but of smaller size and without pigment. The two scars were not continuous around the edge of the disc. In and slightly up from the disc was a patch of fresh exudate about one and a half disc-diameters in length and about half that distance from the disc. Five months later the exudate had almost disappeared revealing a characteristic scar although almost devoid of pigment.

The field of vision showed, in March, three separate defects corresponding to these lesions. There were two large sector-defects extending from the blind-spot, one into the lower nasal and one into the lower temporal quadrant, the former reaching up to but not beyond the horizontal meridian. A small arcuate scotoma curved upwards from the blind-spot. Three months later the field showed considerable improvement (Fig. 6). The arcuate scotoma and the lower nasal quadrant defect remain unchanged, being due to the old scars. The sector-defect in the lower temporal quadrant, corresponding to the recent exudate, has diminished in breadth from 45° to 20°. The borders of the defect are relative.

CASE VI.—A married woman, first seen in June, 1937, aged 22 years, having an acute attack in the left eye. At that time there was a patch of exudate

touching the disc on its medial side, situated between the upper and lower temporal vessels and extending for about one disc-diameter. A small haemorrhage was seen beside the exudate.

The fundus now shows only the slightest trace of this inflammation. On the medial side of the disc is a hemispherical scar of about half its size where the retinal pigment appears to be uniformly denser, giving a slightly greyer colour to the fundus. There is apparently no disturbance of the choroid at all and no disappearance or "clumping" of the retinal pigment. The field of vision shows an absolute, arcuate scotoma extending from the blind-spot around below the fixation-point up to the horizontal meridian on the nasal side.

CASE VII.—A typist, first seen in March, 1931, aged 16 years, complaining of pain and loss of sight in the left eye. The pain was made worse by movement. She seemed in good general health but that her left ear was discharging. On examination the vision was 6/6 in the right eye and counting fingers in the left. The left fundus was obscured by a vitreous haze, but just below the macula could be seen a patch of exudate. In two months the vitreous haze had almost cleared and the exudate was diminishing. Vision had reached 6/60. At this stage she was admitted to hospital and a radical mastoidectomy performed. In six months from the onset of the disease vision had reached 6/18 and was still improving. The patient did not return again for observation so that it is not known whether the vision after that attack became normal.

By 1935 the patient had had three operations performed on the left mastoid in an endeavour to stop the discharge. At that time the ear, nose and throat surgeon considered that the discharge was due only to a patent Eustachian tube and that nothing further should be done. The ear is still discharging. Since the first attack subsided she has had a succession of recurrences of the disease. She has attended hospital rather sporadically but says that she has had not less than six fresh attacks. With each attack there has been pain in the eye made worse by movement. Each acute attack that has been seen has been accompanied by keratic precipitates, vitreous haze, a fresh patch of exudate on the edge of the old scar and reduction of vision to counting fingers. The best vision recorded during this period is 6/18, but it is probable that it has improved beyond this at times.

The patient was last examined in March, 1938, when she was found to be recovering from a fresh attack which had started two months earlier. Below the macula and extending up to it was seen a typical oval, pigmented, atrophic scar about two and a half disc-diameters in length. At the nasal end of this scar was a patch of recent exudate about the size of the disc and extending to within one and a half disc-diameters of it. The rest of the fundus appeared normal. Owing to her low visual acuity it was impracticable to define the field of vision with any accuracy. With a 10/330 white object, however, it was possible to show that there was an absolute scotoma extending from the fixation-point upwards for about ten degrees.

CASE VIII.—A female machinist, first seen in February, 1928, aged 19 years, complaining of aching and blurred sight in the right eye. On that occasion there was papillitis and the scar of a presumed previous attack nasal to the disc. A week later a fresh patch of exudate had appeared just below the scar. Vision returned to normal in about seven months.

A similar attack occurred four years later with slight aching in the eye and cleared in six months.

In October, 1937, at the age of 29 years, the patient was in bed with "influenza" for a fortnight and had a recurrence in the same eye. There was no aching and the sight was only slightly misty. She did not attend hospital.

When examined in March, 1938, there were no signs of activity in the eye. The disc appeared normal. Just nasal to the disc and slightly above it was a group of four typical, round, atrophic scars. The scars were contiguous but did not run into one another. Two were about half the size of the disc and two rather larger. The nearest scar was about half a disc-diameter from the disc and the fundus between showed a greyer colour than normal. Extending forwards, apparently from the disc, into the vitreous was a filmy, transparent, floating "veil" of irregular outline. The field of vision showed an absolute sector-defect extending outwards and downwards from the blind-spot.

CASE IX.—A secretary, first seen in September, 1932, aged 24 years, complaining of loss of vision in the right eye and severe aching in the right frontal region. On examination vision was 6/5 in the left eye and counting fingers in the right. In the right eye there was iritis and a posterior synechia. There were massive vitreous opacities. The fundus showed papillitis and generalised retinal ischaemia. Up and out from the disc, but not touching it, was a large patch of exudate, the macula being involved in its outskirts. Vision recovered to 6/12 in about six months but has not improved beyond this. There has been no recurrence.

On examination recently the iris appeared normal. The right fundus shows an extensive area of scarring starting less than one disc-diameter from the disc and extending up and out for a distance of about three disc-diameters. There are two clear, atrophic scars in this area, a large one at the proximal and a smaller one at the peripheral end. Between and around these are several smaller, less distinct scars and pigmentary disturbances. The macula and the rest of the fundus appear normal (Fig. 1).

Extending forwards from the fundus into the vitreous is the most bizarre "veil" seen in this series. It is in two distinguishable parts. The smaller part takes a broad origin from the disc and extends forwards almost to the lens in a scimitar-like curve, narrowing as it proceeds; it appears that its origin is not linear but oval and that the "veil" is tubular. The larger part seems to take a narrow origin from the disc and arch as a graceful frond over the nearer part of the scar, to be attached again farther out. From this second attachment and from the outer part of the arch a broad "veil" sweeps forwards and outwards into the vitreous like a sail.

Even more remarkable is the appearance of aberrant vessels. A large branch of the central retinal artery leaves the disc up and out and appears to enter the arched part of the "veil," ascending with it into the vitreous and then curving down again on to the scar; from there the main vessel is distributed normally over the fundus but several small branches ascend again into the vitreous in the foot of the "sail." Several veins approaching from the periphery seem to disappear into the scar, one in particular running at first horizontally above the macula bends upwards through more than a right angle to enter the scar. Owing to the interposition of the "veil" it is difficult to see the exact termination of these vessels.

The field of vision shows an absolute sector-defect extending from the blind-spot throughout the lower nasal quadrant, its upper border skirting around 5° below the fixation-point.

CASE X.—A female clerk, first seen in January, 1936, aged 33 years, having an acute attack in the left eye. She remembered that ten years before she had had a similar but milder attack, but thought that it was in the other eye. She remembers no other earlier attack and has had none since. On examination the patch of fresh exudate was seen to be on the edge of an old scar up and in from the disc. Definite spasm was seen just beyond the exudate in an artery which passed through it. There is no note on the condition of the right eye at that time. The left field of vision showed a large, absolute sector-defect out and down from the blind-spot. It extended from 15° above the horizontal throughout the entire lower temporal quadrant, its inner limit curving round 40° below the fixation-point.

When re-examined in March, 1938, the left fundus showed two scars. The larger one was about two disc-diameters in length and extended from the disc inwards and slightly upwards; the fundus surrounding it was rather greyer than normal and this greyness extended on to the nasal side of the disc. The smaller scar was about half the size of the disc and situated three disc-diameters medial to it. The fundus intervening between the two scars appeared normal. The left field of vision showed considerable improvement from the previous condition. There was absolute peripheral loss only beyond 60° from the fixation point and confined entirely to the lower temporal quadrant. With a 1/330 white object the defect reached to within 30° of the fixation-point and curved round below it just into the lower nasal quadrant. With the Bjerrum screen it could be shown that a relative sector-defect extended outwards from the blind-spot. It thus appears that there has been a progressive recovery from the blind-spot outwards, although acuity has nowhere returned to normal.

The right fundus showed a typical scar about two disc-diameters in length situated two disc-diameters in and slightly up from the disc. In addition there were four jet-black spots of pigment unconnected with the scar. Three were flecks, one situated just lateral to the scar, one about two disc-diameters below the disc and one just below the macula. The fourth was larger and shaped like a banana almost one disc-diameter in length; it was situated about one disc-diameter below the macula. Each spot was surrounded by a very narrow ring of paler fundus. The right field of vision showed no sector-defect and no scotomata corresponding to the pigment-spots were found.

CASE XI.—A typist, first seen in October, 1936, aged 39 years, having come up for spectacles. She never remembered having anything wrong with her eyes. Touching the disc and lying beneath the superior temporal vessels was a typical round scar about the size of the disc. Between the disc and the macula was an island of jet-black pigment just like those described in Case X. Two smaller spots were present on the outer edge of the disc. Extending forwards into the vitreous was a long, broad, floating strand. It was difficult to be sure from what part of the fundus it originated, but it appeared to come from just beyond the scar.

The field of vision shows a sector-defect curving around from the blind-spot 15° below the fixation-point and extending throughout most of the lower nasal quadrant. Only the most peripheral part of this sector is absolute, the arcuate part and a wide border along each arm being shown only by a 1/330 white object.

CASE XII.—A metal-polisher, first seen in April, 1934, aged 22 years, having an acute attack in the right eye. Below the exudate could be seen the scar of a presumed earlier attack. Haemorrhages were present near the exudate and fresh ones appeared two months after the onset.

The right fundus now shows a bilobed scar as though produced by two confluent areas of inflammation. The scar is about three disc-diameters up and out from the disc and two disc-diameters in length. The field of vision in 1934 showed a sector-defect in the lower nasal quadrant extending throughout an angle of 115° , having its apex 20° from the fixation point. The field now shows narrowing of this sector to 70° , the apex remaining in the same position.

CASE XIII.—An engineer, aged 32 years, first seen in April, 1938, having an acute attack in the right eye. The attack started with severe headache, from which he very rarely suffers, and profuse epistaxis, to which he is occasionally liable. The headache decreased rapidly in severity although present each morning for the first ten days. He remembered no previous attack. The right fundus showed two scars, presumably from a previous attack. The larger was situated two and a half disc-diameters above, and was about the same size as the disc. The smaller was just up and out from the larger and about one-third of its size. One disc-diameter up and in from these scars was a narrow, oval patch of exudate about one and a half disc-diameters in length. Crossing the lower end of this patch was an arterial twig which could be clearly seen and showed a distinct spasm just beyond the edge of the exudate. About two disc-diameters above this large patch were two very small, round islands of exudate. The disc showed definite papillitis. A small hyaloid remnant was present on the lens. A week later several arteries and veins in the neighbourhood of the inflammation showed one or more points of spasm. A fortnight later the vessels appeared normal again; the vitreous haze was less but the rest of the fundus remained as when first seen. The field of vision showed a narrow, absolute sector-defect downwards, having its apex 30° below the fixation-point.

CASE XIV.—A cabinet-maker, first seen in December, 1937, aged 24 years, having an acute attack in the left eye. He does not remember having anything wrong with either eye before. In March, 1938, the left fundus showed traces of exudate still present and the scar not yet clearly defined. The changes in the scar-area were very slight and there were no pigment masses. The area was rather less than that of the disc, which it adjoined beneath the inferior temporal branches of the central retinal vessels, where the artery showed slight but definite narrowing. Oedema of the macula was still present. The left field of vision showed an absolute arcuate scotoma arching from the blind-spot above the fixation-point.

The right fundus showed a hemispherical scar adjoining the disc on its nasal side. This scar also showed little change, having a greyer colour than the surrounding fundus and a few small areas of deeper pigmentation. No other scar was seen; the rest of the fundus appeared normal. From the disc a small strand-like bucket-handle stood out into the vitreous. The right field of vision showed an absolute scotoma extending outwards from the blind-spot (Fig. 7). The arms of this scotoma diverged at an angle of about 35° as though to produce a typical sector but the defect ended abruptly 40° from the fixation-point. There was a corresponding peripheral defect in the superior temporal quadrant.

CASE XV.—A married woman, aged 26 years, first seen in March, 1938, having an acute attack in the right eye, which had started a fortnight before. The tension in that eye was full and there was slight bedewing of the cornea. A patch of exudate two disc-diameters in extent was present about three and a half disc-diameters nasal to the disc. When first seen there were already two masses of pigment visible through the exudate and these did not appear to belong to a previous scar. The field of vision showed slight peripheral contraction of the temporal border. There was an oval scotoma, its long axis horizontal, beginning at 30° and ending at 55° lateral to the fixation-point. The centre of this scotoma was absolute and the periphery relative, being due presumably to oedema.

CASE XVI.—A typist, aged 25 years, first seen in April, 1938, having an acute attack in the right eye, which had started two months earlier. She did not remember having had anything wrong with the eye before. The details of the fundus could not be seen on account of the very thick vitreous haze in which large "floaters" were present. There appeared to be papillitis. About four and a half disc-diameters out and up from the disc was a typical scar rather larger than the disc and on its upper edge was a patch of exudate of slightly greater extent. The field of vision showed general peripheral contraction, presumably due to the dense vitreous haze and the papillitis, and a sector-defect in the lower nasal quadrant, having its apex 15° from the fixation-point and extending up to the horizontal. The blind-spot was normal in size and shape.

CASE XVII.—A cook, aged 21 years, first seen in April, 1938, having an acute attack in the right eye. She did not remember having had anything wrong with the eye before. The attack started with aching which was made worse by movement of the eye. On examination of the fundus papillitis was present. Two and a half disc-diameters up and in from the disc was a typical scar. On its outer side was a mass of fresh exudate, the whole area being rather more than twice that of the disc. About one and a half disc-diameters above this patch was a small island of exudate with a heap of pigment at its outer edge. It could not be determined whether this pigment also marked a previous scar. The arteries and veins near the lesion and for some distance around showed spasm at frequent intervals so that they appeared to be entirely irregular in lumen. Moreover many of them showed a marked, fluffy, white "perivasculitis." The vessels so affected were particularly numerous between the disc and the lesion and for a considerable area on either side of this zone. The field of vision showed an absolute sector-defect in the lower temporal quadrant covering an area of 15° and having its apex 20° from the fixation-point.

CASE XVIII.—A joiner, first seen in January, 1937, aged 29 years, having an acute attack in the left eye. He could remember no previous trouble with his eyes but had been aware for at least ten years of a "blind spot" in the left eye in the situation of the present sector-defect. This, together with the presence of two scars, would seem fairly reliable evidence of a previous attack. He was found to have several septic teeth. The vision returned to normal in two months. Towards the end of his recovery the bad teeth were extracted. A few weeks later he had a severe attack of acute bronchitis and his left ear discharged for about two months. He had no recurrence of eye-trouble.

When examined in April, 1938, he still had several septic teeth. The left fundus shows two scars, typical in appearance except for the absence of pigment heaping, the edges being marked out sharply by sclera showing abruptly through (Fig. 2). There is no pigment in the scars except for a very fine sprinkling at one corner of each. One scar is just above the disc with a clear area of less

than one disc-diameter between them; the other is just up and out from the first and contiguous with it, with a small satellite scar on its upper border. Both are about twice the size of the disc. The rest of the fundus is normal. A large hyaloid remnant extends backwards from the lens.

The field of vision shows two separate sector-defects, one in the lower nasal and one in the lower temporal quadrant, having a common origin at their apices but separated by an island of normal vision extending to the periphery (Fig. 8).

CASE XIX.—A roundsman, first seen in January, 1937, aged 26 years, having an acute attack in the right eye, when the scar of a previous unrecognised attack was seen. The field of vision showed a sector-defect of about 135° starting from the blind-spot and extending throughout the lower temporal quadrant and part of the lower nasal quadrant. The vision became normal in one month.

The right fundus now shows a scar about two-thirds of the size of the disc and "crowning" it on its upper nasal edge. There is a much smaller, round scar four disc-diameters above this and a similar small one half way between the disc and the macula but on a slightly higher plane. Extending forwards into the vitreous from around the disc is a cone-shaped "veil" reaching perhaps half-way to the lens. The field of vision shows a sector-defect almost identical with the previous one; its extent is remarkable in comparison with the small area covered by the scar. On the Bjerrum screen can be shown a small absolute scotoma corresponding to the scar between disc and macula.

CASE XX.—A hospital nurse, first seen in December, 1937, aged 20 years, complaining of misty sight in the right eye and severe headache. In the right fundus there was marked papillitis and a typical oval patch of exudate two disc-diameters in length and about eight disc-diameters up and out from the disc. Just below this patch was a small, outlying island of exudate. The rest of the fundus was normal. The headaches persisted intermittently for five weeks. Vision returned to normal in about three months.

In April, 1938, the patch of exudate had not quite disappeared and in its centre could be seen a pigmented scar. The disc was normal. The field of vision showed only a scotoma and the blind-spot was normal in size and shape.

CASE XXI.—A married woman, first seen in March, 1936, aged 25 years, having an acute attack in the left eye. There was iritis with synechiae but no ciliary injection or pain.

On re-examination the iris appears normal. The left fundus shows an oval scar two and a half disc-diameters medial to the disc and about two disc-diameters in length. From the papilla a short, sturdy strand with a knobbed free end extends forwards into the vitreous. The field of vision shows an absolute scotoma corresponding to the scar.

CASE XXII.—A printer, first seen in February, 1930, aged 23 years, having an acute attack in the left eye. The left pupil was found to be oval and larger than the right but reacted normally. There was marked papillitis with about three and a half dioptres swelling of the disc. Seven months later there was a typical scar down and in from the disc with choroidal vessels throughout its base.

In January, 1931, the patient had a bad cold and another acute attack developed in the same situation; this time there was pain with the onset.

In December, 1937, he had another bad cold and a further acute attack accompanied by pain. He had had several bad colds in the mean time. On this occasion there was slight ciliary injection but the iris was normal. A fresh patch of exudate developed at the outer end of the previous scar.

On examination in March, 1938, the right fundus showed a large, composite scar about two and a half disc-diameters in length and about one disc-diameter down and in from the disc. The proximal part was composed of the two old scars, from the base of which the choroidal vessels had disappeared except around the edges. The distal part was a fresh scar still slightly blurred by exudate. In the anterior part of the vitreous was a bow-shaped strand with its ends directed backwards, but they could not be traced to the fundus. The field of vision shows a scotoma corresponding to the scar.

CASE XXIII.—A female metal-worker, seen first in September, 1933, aged 20 years. She complained that two days before some benzene had splashed into her left eye and made it very sore and that since then she had been blind in

that eye. She had seen perfectly well before and had had her eyes tested less than a year ago. There was probably no pain apart from the smarting caused by the benzene and apparently there was no injection of the eye when she was first seen.

On examination vision was 6/5 in the right eye and less than 6/60 in the left. The left pupil was slightly larger than the right and was thought not to react so briskly. The tension was full. There was a large number of keratic precipitates. The fundus was obscured by a dense vitreous haze and "floaters." The disc appeared normal. Just temporal to the disc was a large patch of exudate covering the macula. The right eye was normal. Pigment could be seen in the exudate nineteen days after the onset. The inflammation gradually subsided leaving a typical scar. Vision did not improve.

When re-examined recently the vision was 6/5 in the right eye and counting fingers at one yard in the left eye. Central vision was entirely lost. The right eye was normal. In the left fundus, lying lateral to the disc, its edge three quarters of a disc-diameter from it, was a perfectly circular scar of rather more than twice the diameter of the disc with a small satellite scar on its upper border (Fig. 3). It had a pigmented rim, not very heavily marked, and some patches of retinal pigment within the edge. In the centre of the scar there was complete atrophy of the choroid. Around the clear area were normal choroidal vessels, many of them smaller than are commonly seen in these scars. Some choroidal pigment was present towards the edges. The scar appeared to be typical of this disease. The temporal half of the disc was white. The rest of the fundus was normal. A large wedge-shaped opacity was present in the anterior part of the vitreous, attached to the posterior surface of the lens. The field of vision was full peripherally. There was an absolute, circular, central scotoma extending to 8° from the "fixation-point." The blind-spot was normal in size and shape and did not join the scotoma.

CASE XXIV.—A girl, learning to become a typist, first seen in June, 1938, aged 14 years, having an acute attack in the left eye which had started six weeks before. There had been aching in the eye with the onset of mistiness. A patch of exudate rather larger than the disc was present less than one disc-diameter from it and surrounding the inferior temporal branch of the central retinal vein. There was marked papillitis. There was oedema of the macula and just above it was a group of shining yellow dots. The field of vision showed an absolute, arcuate scotoma extending from the blind-spot to the horizontal meridian on the nasal side and arching just above the fixation-point.

CASE A.—A youth of 18 years, first seen in January, 1938, complaining of blurred vision in the left eye for one week. He was in good health at the time. The only illness he could remember was diphtheria seven or eight years before, when he was very ill and had "paralysis" of both eyes. On examination vision was 6/5 in the right eye and 6/12 in the left. The anterior segment of the left eye appeared normal; there were no keratic precipitates. There was vitreous haze, through which a large patch of exudate was seen in the lower nasal quadrant extending to within about five disc-diameters of the disc. The rest of the fundus was normal. The patient was found to have septic tonsils, which were considered to be the cause of the inflammation. The condition settled down quickly and within one month vision had returned to 6/6. He was then admitted to hospital and tonsillectomy performed. Three weeks later his vision was restored to 6/5, an achievement attributed to the elimination of focal sepsis.

Examination in April, 1938, three months after the onset of the attack, showed a quiescent scar quite unlike those usually associated with solitary retino-choroiditis. About five disc-diameters down and in from the disc was the sharply defined proximal border of an oval scar which had no peripheral limit but increased in width while it decreased in intensity as it faded gradually away towards the periphery. There was some heaping of retinal pigment at the proximal border of the scar, but the striking difference from the scars previously described was that it looked "untidy." There were no clear, atrophic areas through which sclera showed; there was none of the characteristic "black and white" appearance. The whole area was obscured by choroidal pigment, giving a dirty brown and yellow picture. A few choroidal vessels could be dimly seen

and appeared to be normal. A normal retinal vessel crossed the scar. Two long, floating "veils" extended forwards from the scar into the vitreous. No normal fundus could be seen beyond the scar but some pigmentary disturbance seemed to radiate from it to the periphery. The field of vision showed a small, absolute, peripheral sector-defect in the upper temporal quadrant, having its apex 40° from the fixation-point.

CASE B.—A woman, aged 53 years, who has had two attacks of blurred vision in the right eye. The first attack occurred at the age of 42 and took three months to clear, the second, ten years later, was exactly similar. At the time of both attacks she was in good health. She had always had bad teeth, but they were all extracted two years before the second attack. Unfortunately the patient was not seen during either of these attacks.

On examination in March, 1938, the vision was 6/24 in the right eye and 6/12 in the left. The right iris showed a few fine posterior synechiae. Both fundi showed general mild arterio-sclerotic changes but the left was otherwise normal. The right fundus showed a large scar beginning at the nasal margin of the disc and extending over a wedge-shaped area medially to the periphery. The disc appeared to be normal. The proximal border of the scar was clearly marked by heaped retinal pigment but the edges had become progressively less distinct towards the periphery. The base of the whole area had an extremely "untidy" appearance, there being irregular patches of retinal and choroidal pigment and a few clear atrophied areas. The choroidal vessels were visible throughout almost the whole extent of the scar and showed a striking degree of sclerosis, some being completely thrombosed and many having only a very narrow lumen. The field of vision showed a relative scotoma extending outwards from the blind-spot to the periphery. It differed from a typical sector-defect in that the outline was very irregular and in that there was marked irregular contraction of the temporal field, which nowhere extended beyond 60° from the fixation-point for a 1/330 object or 70° for a 5/330 object.

CASE C.—A woman, aged 29 years, whose vision in the right eye is reduced to counting fingers at one foot. The left eye is normal. She cannot remember ever having noticed anything wrong with her right eye but the sight of it was discovered to be defective when she was only fifteen years old.

The right fundus showed a normal disc with a well-marked pigment-ring. On the outer side the pigment-ring merged into a dark, slate-grey triangular area about one and a half disc-diameters in length with its apex towards the macula. This area did not in the least resemble the scars previously described, even those of cases VI and XIV, the greyness appearing much deeper than in these; it was darker in some places than others but this was not due to heaping of pigment but to a gradual increase in density. The borders were not sharply defined. From the apex of this triangle there extended a vague area of abnormal fundus covering a roughly circular area about two disc-diameters in extent, including the site of the macula. There did not appear to be any destruction of choroid but the area was generally of a browner colour than the rest of the fundus and had irregular patches of deeper brown. The choroidal vessels were not visible. The retinal vessels were normal. Only peripheral vision remained.

Summary

The history of solitary retino-choroiditis is outlined and it is pointed out that Griffith's and Friedenwald's descriptions preceded Jensen's.

Twenty-four cases have been investigated and the findings compared with those recorded in the literature. An attempt has been made to define criteria for diagnosis.

Points of particular interest which emerge are:—

1. The apparent high incidence of the disease in childhood.
2. The frequent presence of "veils" in the vitreous.

3. One case in which the development of anomalous retinal vessels was preceded by retinal ischaemia.

4. The defects in the field of vision may be wholly or partly relative and may diminish with healing.

5. One case in which a defect in the visual field was relative throughout after the first attack but became absolute after the second.

6. One case in which two separate sector-defects in the visual field were produced by two apparently confluent scars.

7. Defects originating in the nasal visual field appear to be incompatible with the conventional conception of the distribution of the nerve-fibres of the retina.

Many cases recorded in the literature have not been included in the bibliography because they appear to add nothing of interest to the discussion. Unfortunately I have not been able to read the Italian authors intelligently; they are included to bring the references up to date.

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A NEW NEEDLE-HOLDER

BY

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LONDON

THE needle-holder illustrated below in Fig. 1 shows certain modifications of the Silcock pattern. The handle, made of bakelite, is bulbous and shaped like that of an awl and other small carpentry

FIG. 1.