CHAPTER I

GENERAL: AETIOLOGY

GENERAL

DEFINITION. By Detachment of the Retina we mean a separation or cleavage of the two primitive retinal layers so that the pigmented epithelium remains adherent to the choroid, and the inner retinal strata of cells and fibres are separated from it. The retina is not completely detached, therefore this is an inaccurate title. It was the original title, and is universally adopted, therefore the continuation of its use appears to be desirable. Various pathological processes may account for detachment, such as an effusion of serum, blood, or pus, the growth of a tumour, the contraction of fibrous bands, the presence of a foreign body, or an entozoon.

With further investigation it is hoped that the terms "spontaneous" and "idiopathic" as applied to detachment of the retina will become less prominent. It is found that in many of these there is a history of trauma. In an eye predisposed to detachment of the retina by one of several different forms of degeneration, a surprisingly slight trauma can play the rôle of the exciting agent.

The importance of any detachment of the retina depends largely on the effect it will have on the total sight of the individual. It may be a mere incident of little value, as, *e.g.*, in endophthalmitis. Here the underlying pathological basis is so potent as to destroy vision regardless of the additional retinal separation. Its importance however can hardly be overestimated when it results from a preretinal or subretinal haemorrhage which may clear up, leaving the detachment, or if when it occurs in the course of the degenerative changes in a myopic eye, the possibility of useful vision then depends on the end result of our treatment of the detachment. An understanding of the exact mechanism of its production is essential for successful treatment. No matter how overwhelming the causative lesion, it is wise to study and con-

sider its effects, for only by so doing can one gain insight into the pathogenesis of detachment of the retina. On this depends the differential diagnosis and the success of treatment. Detachment due to such destructive disorders as Coats' disease, nephritis, sarcoma, etc., will be referred to and briefly discussed.

If one attempts to divide detachment of the retina into primary and secondary, it is only an artificial division based on the permanent effect and prominence of the underlying cause. If this is latent, the detachment appears to be primary, but if it is obvious and manifest, it is secondary. So all detachments of the retina will be considered as secondary lesions, and therefore as physical signs of some ocular or general disorder.

RETROSPECT. It is fitting before we proceed further to look back to the earliest recognition of detachment of the retina in the history of ophthalmology. The important books on morbid anatomy by Morgagni (1740) and de Krzowitz (1781) do not mention it. Earlier writers have mentioned animals' eyes in which it occurred, and the possibility of its occurrence in man. It is mentioned by St Yves (1722), but we find that he confuses it with "mouches volantes". It was not until much later that reliable histological observations were made by Ware (1805), Wardrop (1818) and Panizza (1826). It was called "hydrops subchoroidalis" to distinguish it from detachment of the choroid (hydrops subscleroticalis) which was in those days considered to be so much more common.

It was not seen *in situ* until expert observers like Chelius (1839), J. Sichel (1841), and Desmarres (1847) saw through the dilated pupil a white and at times vascularised membrane.

It was this appearance that Beer (1817) originally referred to as "amaurotic cat's eye". Invention of the ophthalmoscope was necessary before it could be studied clinically or any idea gained of its frequency. The first descriptions date back to Coccius, van Trigt, Arlt (1853) and A. von Graefe (1854). Certainly by his accurate clinical descriptions it appears that von Graefe fully proved the truth of the state-

ment made by him, when first seeing the fundus with an ophthalmoscope, "Helmholtz has unfolded to us a new world". Because of their sudden onset von Graefe considered most detachments of the retina due to a subretinal haemorrhage. Stellwag (1856), as a result of his anatomical investigations, was able to disprove this idea. Arlt (1853) originated the theory of a choroidal effusion as the causative force, and five years later Müller showed that the organisation and contraction of connective tissue in the vitreous was the cause in certain eyes.

When Magnus (1883) was studying the various causes of complete blindness, he considered that 4·74 per cent. were due to retinal detachment. Evans (1929), when classifying 700 patients certified as blind, found retinal detachment in 4 per cent. and myopia the cause in approximately 3 per cent. More recently Cords (1930) has shown that myopia and bilateral detachment cause a higher percentage (19 per cent.) of blindness in the city of Cologne than any other disease.

AETIOLOGY

INFLUENCE OF AGE, SEX AND OCCUPATION. There are certain factors worthy of consideration. The first deals with the relation of age to the occurrence of detachment of the retina. Leber found that it was rare before the twentieth year, and that it gradually increased in frequency with increasing years. It appeared most frequent between the ages of fifty and sixty. Poncet (1887) and Walter (1884) found that about 66 per cent. of cases occurred after the age of forty. Stallard (1930) found 61 per cent. after this age. This preponderance becomes greater when we recall the obvious fact that there are considerably fewer patients alive after forty than before forty. Poncet (1897) found sixty to be the most affected age. Sattler (1905) found that males were more frequently affected than females; the proportion being 66 : 34. This disproportion is not due to the greater exposure of the male sex to injury, exclusion of non-traumatic cases from the statistics not affecting it. In Stallard's and Poncet's series, 62 per cent. were in males. Occupation does not appear to affect the incidence of retinal detachment. Magnus, who

analysed a series of 243 patients, found that in half of them no special demands on the eyes were made. Leber and Poncet concurred with this view.

BILATERAL OCCURRENCE. The most marked variation occurs in the estimation of this point. Naturally if patients were followed over a longer period, and if detachments due to nephritis were included, a greater number of bilateral cases would be reported than in others which excluded these. Galezowski (1883) reported 9 per cent. of 551 patients and (1895) 2·5 per cent. of 1129. Deutschmann (1907), whose series included a very large proportion of severe cases, reported bilateral detachment in 32 per cent. of 220 patients. But Elschnig's figures (1914) are probably the most valuable. In 99 patients with non-traumatic detachment both eyes were affected in 25 per cent.

INFLUENCE OF HEREDITY. Treacher Collins (1892) and Clarke (1898) reported a family in which a brother and two sisters were affected. The condition was present in early life, and the parents were related by blood. In Arlt's family (1888), a woman, her son and granddaughter were affected. But here as in many other instances the patients were myopic, and it was this tendency that was inherited (Salzmann, 1921). In Kennon's family (1920), two brothers and one sister with otherwise normal eyes were found to have detachment of the retina. Previous injury excited the lesion in the males. Schreiber (1920) reported a mother and her son with bilateral detachment in non-myopic eyes. Pagenstecher's family (1913) was exceptional. Here the condition appeared to be transmitted from an affected male, through an unaffected female to two males. However, other ocular lesions were found in each patient as well. In three of the five members of a myopic family reported by Schmelzer (1929) a detachment was found. Pigmented areas in the retina and choroid of each affected eye were also seen. These were considered to be myopic in origin and the probable basis necessary for the detachment. Isolated cases associated with congenital coloboma of the iris and the choroid have been observed (Komoto, 1926; Wagener and Gipner, 1925).

CONGENITAL CASES. Numerous examples of retinal detachment found during the first few weeks of life have been reported. Some of these have been reported as pseudo-glioma. In most cases foetal inflammation, with or without delayed involution of the hyaloid artery, appeared to be the most satisfactory explanation. Injury at birth explains some of these cases (Fernandez, 1905; Lachman, 1927; Marshall, 1897; Rockliffe, 1898; Fleischer, 1907). Onken (1928) has discussed the possible connection between naevus flammeus and retinal detachment.

THE RECOGNITION OF RETINAL DETACHMENT. The signs of an impending detachment are those due either to retinal irritation, choroidal inflammation, or some vitreous degeneration. Patients therefore frequently describe flashes of light, distortion of objects looked at, or an increasing number of black specks in the visual field. Once the separation has occurred, a cloud or a veil is often the way in which the field loss is described.

In Stallard's series (1930) a sudden onset is described in 52 per cent., and in 42 per cent. the onset was gradual.

Central vision need not be affected, even though a detachment has been present for many years. A patient reported by Lawson (1924), maintained 6/5 for at least fourteen months, even though there was a bilateral detachment present.

Two signs of interference with the rods and cones, or the visual purple, are an increase in light minimum and colour-blindness for blue. This is a combination which is due either to congestion of the chorio-capillaris, or to some other upset in the functioning of the rods and cones. A loss of transparency of the fundus, a lack of brilliancy, and a very red fundus are further evidence of capillary congestion. Though at times green or blue vision is associated with retinal and choroidal diseases, coloured vision appears to be very rare in detachment of the retina. Beaumont found that the field for red demonstrated the extent of a tumour more accurately than the field for white.

The symptom which chiefly helps us in the recognition of a detachment of the retina is a loss of one area of the visual

field, especially if the onset is sudden. The presence of myopia, the previous history of transient visual diminution, or the occurrence of some injury, are further evidence. Often the patient states that there has been for some time an appearance of flashing lights or sparks, or that objects looked at appeared distorted. Once the detachment is established, monocular diplopia may be found. The diagnosis is clinched if, through the pupil, either with or without the ophthalmoscope, a greyish membrane is seen showing the characteristic ramification of retinal vessels. If, however, the media are not transparent, the difficulties of diagnosis may be great. The main signs then are reduced tension and a loss in the visual field. For a consideration of the difficulties in diagnosis see the section on "Differential Diagnosis".

As the pigmented layer of epithelium remains in contact with the choroid, the retina is at first transparent. Often when first seen it has become dull and grey, because it has been separated for some time from the choroid which nourishes so much of it. Later, as a result of atrophy, it becomes transparent again.

One characteristic appearance is the manner in which the vessels run over the undulating folds of the retina. They soon appear black in colour, because of the increased proportion of light reflected from the choroid. It is this dark appearance of the vessels and a tendency to tortuosity that gives one the greatest assistance in recognising a shallow detachment. The vessels later may show patchy infiltration of the lymph sheaths.

SLIT-LAMP FINDINGS. If an eye with a retinal detachment is studied with the slit-lamp one may find numerous dot-like deposits and well-defined and twisted fibres, due to coagulation of albumen in the vitreous. If myopia is present, the marked degeneration of the vitreous will be shown by the presence of prominent white fibres, narrow bands with transverse striation, fine white and brown dots and large masses which are fixed to the fibres and bands. Vogt (1921) finds a "dissolution" of the framework in high myopia. "Large areas appear optically empty, and on bulbar move-

ment masses of fibre-like framework rapidly move." They do not gravitate, but usually return to their original location, so that we must presume that they are attached to a definite vitreous supporting structure—the "Glaskörperbasis" (Salzmann). Figure 353 in Vogt's atlas shows changes which he considered characteristic of detachment in myopia. Brownish red dots, evidently containing pigment, were attached to the framework, which itself in part was dissolved into fibres. The framework was more freely movable than usual and delicate white punctate changes and larger pigmented deposits were found. The retro-lental space was absent. If the tension is very low, the vitreous may assume a greyish yellow colour due to serous infiltration. When the retina has become atrophic its surface may show white masses or ramified lines over which run degenerate vessels ending in little coils. One may be able to discern dots of pigment epithelium adherent to its outer surface, and the subretinal exudate often appears stiff and of a dull yellow colour. A detachment due to inflammation with vitreous exudate may show numerous minute new vessels on the retinal surface and very delicate strands which radiate from its surface into the vitreous (Meesmann, 1927). Koby (1925) states that the vitreous framework is completely broken up in old myopic detachments. There are irregular and twisted fibres or large coloured platelets but no sign of a regular scaffolding.

THE RETINA AFTER DETACHMENT. The external layers of the retina normally receive their nourishment by osmosis, and it is possible that this process may continue even though they are separated from the pigment epithelium as in retinal detachment.

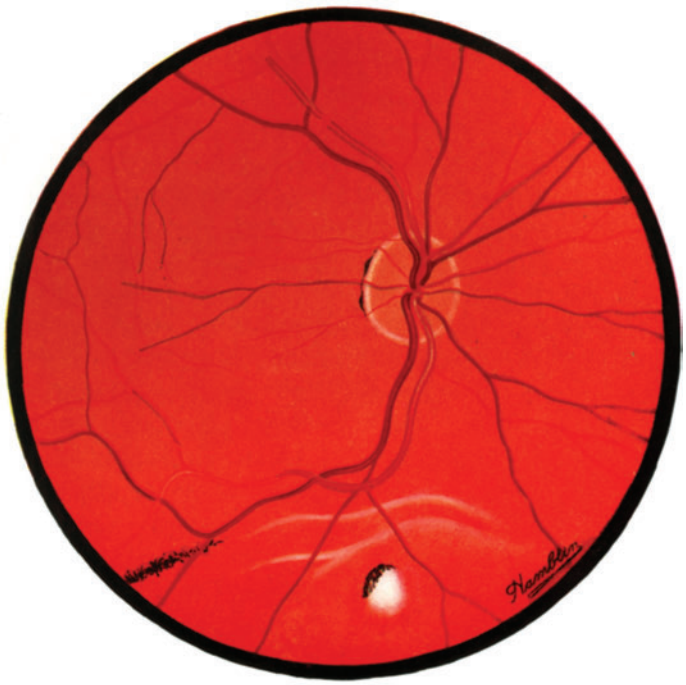
It appears that, even though the retina has been separated from its epithelial layer for some time, recovery of vision can occur. This is probably made possible by the filaments of the pigment cells growing in between the visual elements again. In an èye with typical detachment the retina was found only slightly raised from the choroid in two areas. The filaments were still present in a few places, but in most places they were broken off and formed a layer outside the rods and

cones. As long as sufficient filaments remain intact, no changes in the rods and cones are noted, and vision may be as high as one-tenth with full perception of colour. If these connections are destroyed the rods and cones swell and become degenerate, the latter being more affected than the former. In old cases no trace of rods and cones may be found, and all the other cells appear degenerate (Speciale-Cirincione, 1925). The pigment epithelium shows an irregularity in the arrangement of its cells, and a tendency to proliferate through the subretinal space. The tendency for excrescences from the lamina vitrea (*drusen*) to form is marked. Areas of pigmentation and of fatty degeneration are found, and appear black, brown or yellow. White lines, due to cholesterin crystal formation, are further signs of degeneration. The yellow colour of the macula persists for a surprisingly long time after the development of a detachment (Vogt).

THE FUNDUS AFTER RE-ATTACHMENT. If a detachment has not been present too long, on re-attachment the fundus may assume its former appearance completely. As a rule, however, certain alterations are visible. These may be in the form of an irregular and dark pigmentation or a spotted appearance as in old choroiditis. This is due to the partial disappearance and the migration of pigment. But the most striking appearance is that described as "retinitis striata". The unsuitability of this term is obvious. It is wiser to adopt Schilling's classification (1903). He included under the title of "retinitis striata" any changes, due to inflammation, which assumed a striate arrangement; but the white lines following detachment he called "striae retinae". As far back as 1869, v. Jäger had recognised them and designated them "retinal cords".

After re-attachment one may observe these white retinal striae extending over a considerable area of the fundus (*v.* Plate 1). They are at times edged with pigment, and then may be raised, leading to undulation of the vessels which cross them. The striae are often about the same width as the larger retinal vessels. At times they branch. They lie deep

PLATE I



"Retinitis striata", and a healed hole. (Note that the vessels are not deflected as they pass over the streaks.)