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COMMUNICATIONS

FAMILIAL HYALINE DYSTROPHY IN THE FUNDUS
OCULI OR DOYNE'S FAMILY HONEYCOMB
"CHOROIDITIS"

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THE object of this paper is to deal particularly with a familial affection of the central and paracentral areas of the fundus, characterised by hyaline infiltration of the retina, and known as Doyme's honeycomb "Choroiditis," or Doyme's family "Choroiditis." The appearance of hyaline exudates in relation to the choroid and retina, occurs during the progress of various pathological conditions in the eye. An imperfect appreciation of the specific characteristics of widely different clinical entities, has led to difficulty in recognising and differentiating the fundamentally separate aetiological types.

Hyaline degeneration is very common in the eye, and is a product of altered local metabolism. It occurs as a degenerative change in inflammatory exudates; as a result of chronic toxaemias; as a local effect of vascular sclerosis; as one of the manifestations of senile change; and as an example of premature senescence in

ocular tissues, induced by abiotrophies of congenital and familial origin.

It is hoped that the characteristics of the familial affection, seen by the writer at different stages of its development, and here more fully dealt with than heretofore, will help in its early recognition, and to distinguish it from other types, and also assist in the elucidation of the tangled causes of related conditions.

In 1876, Jonathan Hutchinson published a report on 10 cases of "Symmetrical central choroido-retinal disease occurring in senile persons." These cases, though under the care of Hutchinson, had been observed by Waren Tay, who had also contributed the case-notes. Three of these cases reported were sisters, aged 60, 50, and 40 years. The family history revealed that their father and one of his brothers also had bad sight, and other members had nervous defects. The rest of the cases reported were obviously an heterogeneous collection of different types, and included such conditions as:—

(1) Old standing choroiditis with discrete large round atrophic patches in the periphery of the fundi (case 6).

(2) Doubtful renal retinitis with albuminuria and oedema (case 8).

(3) Tropical infections with traumatic rupture of the choroid and vascular changes (case 9).

(4) Probable syphilitic infection (case 10).

All these cases have gone down in the literature as examples of "Tay's central senile guttate choroiditis."

Apart from the fact that the report by Hutchinson and Tay contains the earliest reference to this type of familial degeneration in three sisters, the inclusion of seven other cases of central fundus changes with widely different causes, at once emphasises the need for appreciation of all the factors involved. Not only does this imply an insistence on the important fact that pathological processes of most diverse origin produce an ophthalmoscopic picture of whitish spots in the fundus, but also a reiteration of the fact that a particular degeneration, namely hyaline change, may be the end result of different diseases. A perusal of many communications published subsequently to that of Hutchinson and Tay, at once makes it evident that the above statement on the importance of careful differential diagnosis is not excessively laboured.

The first definite dissociation of a familial type of "central choroiditis," as a distinct clinical entity, was due to the painstaking observations of R. W. Doyne.

Doyne's observations were communicated to the Ophthalmological Society of the United Kingdom, with the demonstration

of affected cases (1899-1910); and he showed further cases at the British Medical Association's meeting in July, 1914. In 1913, Treacher Collins had the opportunity of preparing sections, and studying the histology in one of Doyne's cases. [case (4)].

The following may be taken as a summary of Doyne's observations on the condition :—

It first appears in early adult life (in one case observed at 20 years of age), but much more commonly later (*i.e.*, at about 40 years of age). It may either affect the disc neighbourhood, the macular area, or the "disc-macula" area. It consists of circular whitish patches of exudation. These increase in size and number during middle age, and set up some irritation and pigmentary disturbance, for though gross pigmentation is not a usual appearance, in some cases there is a good deal to be seen. The spots of exudate become agglomerated in the "disc-macula" area, and present a closely grouped distinctive, honeycomb appearance. In old age the condition passes into atrophy, the discrete outlines of the spots forming the honeycomb disappear, merging to form a more or less homogeneously atrophic area. In early cases there is no disturbance of vision. Even when gross lesions are found, the vision does not appear to be proportionately affected. When the lesions become confluent and atrophy ensues, there is a steady failure of sight. Both eyes are always involved, and the condition is steadily though very slowly progressive. It is hereditary, affecting several members of the same family in different generations. No special family diathesis can be detected.

Treacher Collins's report on the eye submitted to section, was in substance, as follows :—

A new formation of hyaline substance is found situated between the retina and choroid, the inner surface of which presents nodular elevations, with depressions between them. In places the hyaline material is covered by a single layer of pigment epithelial cells, in others it is devoid of any such covering, whilst here and there in the recesses between the nodules, heaped up masses of pigment cells are seen. The only changes to be found in the retina and choroid are such as can be accounted for by the pressure of the hyaline mass, which is evidently of primary formation, and attributable to some hereditary degeneration in the pigment epithelium.

At this stage it is useful to recall the normal histology of the parts, and to recognise that the membrane of Bruch is normally a glass-like hyaline structure, and that it is upon the surface of the membrane of Bruch that these new exudates are formed. In considering the origin of the exudates, it is helpful to deal further with the formation of Bruch's membrane. This membrane when stained with Weigert's Elastin is seen to be composed of two

layers, an inner which is homogeneous and becomes diffusely blue, and an outer which is finely reticular, and takes on the deep colour characteristic of elastic tissue. The difference between the two layers shows best where the membrane is thickest, *i.e.*, in the vicinity of the optic disc. At the margin of the papilla they end at different levels, the inner one ceasing with the pigment epithelium, and the outer continuing on for some little distance into the substance of the papilla. These two layers have different origins, the inner homogeneous layer, like the capsule of the lens and Descemet's membrane, which it closely resembles, being formed as a kind of secretion from the pigment epithelial cells which line it; and the outer reticular elastic fibre layer being continuous with the elastic fibre network of the choroid. The inner homogeneous part of the membrane of Bruch, though it becomes slightly thickened with the advance of years, is mainly a product of the pigment epithelial cells during embryonic life. Under normal conditions the capacity which the cells have of producing hyaline tissue ceases in the fully developed eye. Under pathological conditions the pigment epithelium re-acquires this hyaline-producing capacity, and nodules of it may be formed on the inner surface of Bruch's membrane. While recent, small, hyaline nodules remain homogeneous, the larger and older ones may show lamination and calcareous granules in the centre.

In other cases similar hyaline nodules form in connection with the termination of the elastic lamina at the margin of the optic disc, giving rise to elevations on the margin. These latter have to be distinguished from the *Drusen* bodies arising from the centre of the disc, and are demarcated by their marginal site, and their continuity with the elastic lamina. The central *Drusen* bodies in the papilla are usually grouped around the retinal vessels in front of the lamina cribrosa, are not connected with the membrane of Bruch, and are presumably not produced by the pigment epithelium. It is possible that the central *Drusen* bodies are derived from the neuroglia of the optic nerve, for the cells of the neuroglia are derived from the pedicle of the developing optic vesicle, and the pigment epithelial cells from the outer layer of the vesicle. The presence of *Drusen* bodies with retinitis pigmentosa has been described, and it has been pointed out that overgrowth of neuroglia occurs in this latter condition. However, the aetiology is unsettled, and the following theories have been advanced. Lauber believed them to be due to misplaced pigment epithelium. Parsons believed they originated from degeneration following an earlier inflammatory process; while Fuchs favoured the origin from neuroglia. Their interest from the point of view of this paper lies in the fact that the large central *Drusen* bodies (which have been described in some cases as causing a swelling on the

disc of as much as 12 or 14 dioptries), have never been noted in any cases of Doyme's family "choroiditis."

Hyaline degenerations in other conditions are to be distinguished by the size, texture, situation, and arrangement of the exudates; and by a recognition of the characteristic progress, and associated clinical conditions. Thus the exudates in arterio-sclerotic retinitis are composed of hyaline spots in the outer molecular layer. The "cotton-wool patches" and the star figure of renal retinitis are similarly hyaline exudates, and stain strongly in the usual manner, with eosin, acid fuchsin, and picro-carmin; while they do not give the amyloid mahogany-brown reaction with iodine.

According to Leber the exudates in retinitis punctata albescens are also hyaline, while the same is probably true of retinitis circinata.

These related degenerations have here all been lightly touched upon, with the object of drawing attention to their associations. It is proposed to discuss them further after the main subject of this paper has been dealt with.

Reverting to the immediate details of the main subject matter, the writer collected the case notes of all likely patients who attended the Oxford Eye Hospital since 1896, and was able to find records of 25 cases. In six of these cases there was no record to be found of investigation of the family history, and therefore these are excluded. Of the 19 remaining cases a large number were dead, including the original case whose eye was submitted to section by Mr. Treacher Collins. The writer was, however, able to examine 7 of the 19 cases personally, on repeated occasions. During the process of examining other members of the families he was also able to discover one new unrecorded case [case (15)], and in addition 12 unaffected descendants were also examined. The records used thus consist of 20 affected cases and 12 unaffected descendants. Of the 20 affected cases—

- 9 can be grouped into one family tree [the High Wycombe family, cases (1)-(9)];
- 6 into another family tree [the Eynsham family, cases (10)-(15)];
- 2 other cases were sisters [cases (17) and (18)];
- 2 other cases were brothers [cases (19) and (20)], and
- 1 case [case (16)] gave a family history of six members with bad sight out of seven, and one sister was said to have been seen with the same condition as the patient, but no notes were found.

THE HIGH WYCOMBE FAMILY.

*—Affected cases examined at hospital.

S—Seen by writer.

(1), (2), etc.—Refer to case histories.

(a), (b), etc.—Case reference of unaffected descendants.

D—Known to be dead.

THE EYNHAM FAMILY.

*—Affected cases examined at hospital.

S—Seen by writer.

(10), (11), etc.—Refer to case histories.

(J), (K), etc.—Case reference of unaffected descendants.

D—Known to be dead.

Of the 8 affected cases, and 12 unaffected descendants who the writer examined, the examinations where possible consisted of :—

A record of visual acuity, state of refraction; record of visual fields and charting of scotomata. A rapid clinical estimation of the light sense and colour thresholds (using Dr. George Young's album of threshold tests), and examination of the fundi and media under mydriatic. In addition in some cases, urine and blood-pressure examinations were made, and in a few cases of blood-sigma test for syphilis was performed. The records submitted also include fundus paintings, which serve to emphasise the various stages in the development of the fundus appearances. Two genealogical trees have been constructed of the two main families referred to, and are appended.

The general conclusions to be drawn from the series investigated are as follows :—

State of Refraction

(i) In affected cases the refractive condition varied from emmetropia to small degrees of ametropia, extending from +2.0 D. to -1.50 D. In addition, in one case myopia of -3.50 D. was present, and in another case myopic astigmatism in both eyes occurred [-6.50 D.cyl. ↓, case (6)].

(ii) In the 12 unaffected descendants [cases (a) to (l)], one had myopia of extreme degree [Rt. $\frac{-13.0 \text{ D.sph.}}{-3.0 \text{ D.cyl.}} \rightarrow$ L. $\frac{-13.0 \text{ D.sph.}}{-3.50 \text{ D.cyl.}} \rightarrow$ case (h)].

Three had myopia of from -3.0 D. to -4.0 D.; two very small amounts of myopia -0.50 and -0.75 D.; and five of the younger members were emmetropic.

From this it may justifiably be deduced that the affected eyes showed no marked tendency to hypermetropia and under development, but that the eyes of unaffected members were inclined to be more myopic.

Visual Fields

In affected cases the visual fields remained uncontracted, even when the fundus changes were advanced, and the exudates had become confluent. The advanced cases showed the presence of an irregular, paracentral scotoma in the temporal part of the fields, continuous with the blind spot area and extending towards the fixation point. In one case the scotomata were crescent-shaped and concentrically disposed around the central area [case (9)]. In the case newly discovered by the writer,

although the fundi showed marked lesions, no scotomata were observed and the vision was 6/5. [case (15)].

From this one must conclude that the visual fields show no abnormality in early cases; that the peripheral fields remain full throughout, and that gross disturbance of the retinal function is only manifested in advanced cases by the appearance of a para-central scotoma.

The Light Sense and Colour Thresholds

These were rapidly estimated for each eye separately using Dr. George Young's Album of Threshold tests, with the patients wearing their appropriate lens corrections for reading distance.

Using these tests on the early case discovered by the writer with 6/5 vision, both light sense and colour thresholds were normal in both eyes (1st class). In grossly affected cases, both light and colour thresholds were equally reduced. It therefore seems that the estimation of light sense and colour thresholds, as by the method described, yields no additional useful information.

General Systemic Examination

No other obvious defects or associated systemic disturbances were found common to this condition. In a few cases the blood-sigma test for syphilis was performed and found negative. Similarly blood pressure estimations and urine examinations revealed no abnormalities. No associated mental disturbances were noted in the patients examined, nor was there any family history of mental trouble. In this latter respect the cases differ from the three sisters described by Hutchinson and Tay.

Examination of Fundi and Media

Except for the occasional occurrence of the usual senile lens changes in a few of the more aged patients, no abnormalities were noted in the media of affected patients. No posterior polar or complicated cataracts were noted. No instance of vitreous disturbance was seen; and no constantly associated developmental ocular defects were found. The retinal vessels and the colour of the optic discs were normal for patients at that age.

The average fundus appearance of a typical fully developed honeycomb "choroiditis," consists in the presence of a horizontally disposed oval area of closely packed rounded exudates, enclosed above and below by the upper and lower temporal branches of the retinal vessels. Laterally the area extends from the margins of the optic disc towards the temporal side of the fundus for about five disc diameters, while the vertical extent is

about three disc diameters. The exudate are greyish-white or putty-coloured, and are for the most part of uniform size and rounded. The average diameter of the spots is about 2-3 times that of the main retinal vessels at the disc. At this stage the spots are discrete and surrounded by shadow-rings suggesting a honeycomb or circular mosaic appearance. A few round exudates

CASE 3.

are present on the disc margin deep to the retinal vessels, and a few isolated spots occur to the nasal side of the optic disc. A small amount of irregularly disposed pigment may be present, mostly in interlacing linear formation in the macular area, with some fainter cloudy pigmentation. As a temporary phenomenon a small amount of haemorrhage may be seen at the macular region, and this occurrence coincides with a decrease in visual acuity [cases (6) and (14)]. The whole condition is bilateral, though the appearances may vary in the two eyes and may be slightly more advanced in one than in the other.

The fundus in early cases.—The earliest exudates are rather smaller than in the fully developed case, and are somewhat yellower in colour. The latter may presumably be due to their greater depth from the surface of the retina, and the accompanying diffusion of colour from adjacent vascular layers. The exudates occur first in the paramacular region especially between the disc and the macula. They spread in very close proximity around the macula and often encroach upon it at a very early

CASE 4.

stage [case (15)]. In some cases the spots are disposed all round the macula in a ring [cases (3), (7), (10) and (11)], and the condition has then been likened to that in retinitis circinata. However, apart from the circinate disposure of exudates, there is little similarity between the conditions, as later discussion will show. Accompanying the para-macular exudates in early cases, there are also often a few spots resting on the margins of the optic discs, and occasionally a few tiny spots of exudate in the fundi to the nasal side of the discs. If any pigment is visible it is superficial to, and overlaps the exudates in the macular area.

CASE 6.

CASE 7.

CASE 15.

The fundus in advanced cases.—The advancement of fully developed cases, is characterised by the confluence of exudates, beginning in the centre of the affected area of the fundus, to form a pale yellowish plaque [cases (8), (12), (14)]. Before this process is completed the more peripheral discrete exudates are seen encircling the confluent zone. With further extension the plaque of exudate shows a definite raised margin delimiting the affected area [case (13)]. Meanwhile atrophic changes commence in the centre of the plaque, exposing the choroidal vessels to view [case (4)]. Extensive central atrophy may occur while the more peripheral encircling exudates still remain discrete. Such an appearance will give rise to yet another type of circinate phenomenon [case (4)].

Differential Diagnosis

An earlier reference has been made in this paper to the three sisters described by Hutchinson and Tay. The description given of these three cases conforms to that associated with Doyne's family "choroiditis," with the exception that there was also a history of serious mental defect in two other members of the family. Other cases without the element of family mental defect have since been described. Nettleship, in discussing some hereditary diseases of the eye (1906), referred to the condition des-

cribed by Doyne, and stated that he had seen several examples of "Tay's choroiditis" and sometimes more or less characteristic retinitis circinata,—in brothers or sisters. He described a man aged 72 years seen in 1896 with bilateral retinitis circinata. One of his sisters showed "a mixture of Tay's choroiditis and epithelial denudation." Another sister was said to have partly lost her sight from "disease at the back," while the mother of these three became blind as an old woman, "not from cataract." Nettleship described another case in an affected family where there were Tay's dots of larger size than usual.

In December, 1909, during a discussion initiated by R. W. Doyne, Harrison Butler described two cases he had seen in a mother and son. In May, 1910, G. T. Mould described a case where three members of the family were known to be affected. These additional cases, above referred to, serve to accentuate features of the condition previously described, and point especially to the necessity for differential diagnosis from retinitis circinata, and the non-familial type of Tay's choroiditis.

The non-familial type of Tay's choroiditis.—This condition presents minute pale yellowish spots, which are always small, with little evidence of fusion, occurring in old persons, as one of the manifestations of senility, and without any particular family proclivity.

Retinitis circinata.—Typical cases of retinitis circinata occur in elderly women with degeneration of the retinal vessels. The condition is associated with and follows retinal haemorrhage, and is unilateral in 50 per cent. of cases. Recovery with absorption of exudates is also known to occur. The appearance is that of a girdle of bright white patches of irregular shape and size, becoming confluent to form map-like figures, and disposed in a wide ring around the macular area. The latter shows yellowish-white areas, slight pigmentation, and often haemorrhages. In such cases, therefore, the girdle of exudates is whiter, larger, less regular, and more readily confluent, than in the family condition under consideration. The white exudates are limited to the girdle area, and in addition, no typical exudates occur on the margins of the optic disc. Further, the retinal vessels are diseased, both eyes are not always affected, the patient is old, and there is no family history.

As previously described, two stages in the evolution of Doyne's "choroiditis" may present a circinata appearance. In the early stages, the exudates may for a time be disposed around the macula in the form of a ring: but of course such exudates are discrete, round, and regular, and are disposed in more than one row, like a portion of a honeycomb. In the very late stage associated with atrophy, there will be a prolonged history of progressive loss of

vision, the affected eye will show exposure of the choroidal vessels in the macular area, and the other eye will be extensively involved.

Apart from retinitis circinata and Tay's choroiditis, perusal of the literature on this subject reveals that the following additional conditions have sometimes given rise to confusion, and therefore must be briefly considered:—

1. Cerebro-macular degenerations.
2. Retinitis punctata albescens.
3. Early mild disseminated choroiditis.
4. Arteriosclerotic retinitis.
5. Renal retinitis.

While late cases may also have to be distinguished from:—

6. Diffuse choroiditis; and
7. Exudative retinitis. (Coats).

Cerebro-macular degeneration may for convenience be considered in association with other heterogeneous cases, some of which may possibly be grouped with Batten-Mayou disease, and congenital day blindness. The exudates are usually limited to the macular area, the condition is noted at an earlier age, and mental defect in the patient is a prominent feature.

Retinitis punctata albescens presents the clinical signs of retinitis pigmentosa in addition to the presence of exudates. There is therefore night blindness with contracted visual fields or broad ring scotoma. The exudates are uniformly distributed over the entire fundus, with the exception of the immediate macular area. They are white and very small. Their average diameter is about half that of a medium sized retinal artery. In some of these cases, symptoms of cerebral degeneration also occur. In addition, some cases are seen intermediate in character between cerebro-macular degeneration and retinitis punctata albescens. Thus Nettleship refers to a case reported by R. D. Batten and E. H. Holthouse in 1897. The patient was a woman aged 25 years, with abundant choroidal exudates. She was the youngest of a family of 24, of whom 19 died early of some obscure brain disease. Dr. Guthrie who examined the patient believed her to be epileptic, and suggested that the condition was akin to retinitis pigmentosa. In addition, examination of the plate accompanying the case report, showed a different picture from that given by Doyne's family choroiditis. The exudates were mostly irregular in shape and this applied even to isolated discrete spots. Further the exudates were more widely distributed into the peripheral parts of the fundus.

Early mild disseminated choroiditis may involve the central part of the fundus. During the active stage, the edges of the exudates are hazy and not sharply defined; there are dust-like opacities in the vitreous, and precipitates may be seen on the

posterior surface of the cornea. The Wassermann reaction (W.R.) is often positive. These additional features, and the subsequent progress in affected areas, should make differential diagnosis fairly easy. In spite of which, this condition was obviously present among the cases originally described by Hutchinson and Tay.

Arteriosclerotic and renal retinitis may here be considered together. Case 8 of Hutchinson and Tay's series was probably of renal origin. The presence of cotton-wool patches and star figure at the macula, would of course easily distinguish the atypical nature of the condition. The small round exudates present in both renal and arteriosclerotic cases, are usually whiter and smaller than in the cases under consideration. In addition, the vascular changes, the possible unilateral distribution in arteriosclerotic cases, and albuminuria in renal cases, should facilitate differential diagnosis.

Diffuse choroiditis during the stage of exudation may here have to be distinguished from the advanced cases of Doyne's condition showing confluent exudates. However, in the former condition, as in true choroiditis, the exudate has no definite edge but shades off into the normal fundus; the affected area is irregular, and may finally involve the greater part of the fundus.

Exudative retinitis (Coats) may have to be considered especially in elderly patients where the affection is bilateral, or where the other eye shows the presence of retinitis circinata. It is possible that the latter condition is merely a less severe form of exudative retinitis, and both are probably due to retinal haemorrhage. Disease of the retinal vessels is a prominent feature, and there is a history of sudden onset of visual defect associated with retinal haemorrhage. The exudation may be typically massive, in which case the extensive continuous sheet of exudate covers the whole macular region. There is variation from place to place in colour, depth, and texture. No isolated discrete exudates will be seen along the margins; and if the second eye is involved there is not necessarily any relation between the extent of the lesions in both.

It is hoped that the preceding consideration of differential diagnosis will be helpful in eliminating doubtful cases. The appreciation of the stages of development of the fundus condition may be aided by reference to the accompanying fundus paintings. The case notes of the affected patients, and of unaffected descendants who were examined, are appended together with charts of visual fields and the genealogical trees of the two main families.

In conclusion I have pleasure in acknowledging my indebtedness to Mr. P. H. Adams for permission to use the records of cases at the Oxford Eye Hospital; and to Mr. A. Sorsby who suggested that these investigations might be usefully undertaken.

CASE 1. Mrs. ELIZABETH L.

Seen *March* 13, 1901, aged 52 years, with defective vision.

R.v. = 6/36 not improved. } Pupils equal and react to light and accommo-
L.v. = 6/60 not improved. } dation.

Condition of Fundi.—Both fundi show much the same appearance, but with more marked change in the right. Starting from the temporal side of the optic disc and extending somewhat beyond the macula, and vertically above and below the macula, is an area formed by closely aggregated circular white spots (like circular mosaic). There is a little pigment here and there, especially at the macula and around the optic disc—but speaking broadly, pigment is remarkable by its absence. The nasal side of the retina and the periphery generally seem normal. The pigment where it is seen is well forward in the retina, in some places masking the vessels.

March 27, 1901. The fine pigment patches seem to be far in front of the choroid, with evident parallax between the two.

March 26, 1902. R.v. = 6/60 } Fundi unchanged.
L.v. = 6/60 }

July 16, 1903. *In status quo.*

July 13, 1904, and *July* 23, 1906. Given Mist. pot. iodide.

CASE 2. Mrs. MARTHA S.

June 14, 1899, aged 53 years.

R.v. = 6/36 with +2.0 D. sph. = 6/12.

L.v. = 6/60 with +2.0 D. sph. = 6/12.

R. fundus.—Large pigmented crescentic patch at disc outwards numerous patches of choroidal exudate and black pigment in macular region and also down and out.

L. fundus.—Patch of black pigment at disc outwards. The macula normal but there are small white patches of choroidal exudate, scattered irregularly around the macula, with fine lines of pigment.

July 13, 1904. R.v. 6/60 with +2.0 D. sph. = 6/24 one letter.

L.v. 6/60 with +2.0 D. sph. = 6/36.

Refraction under H. and C.

	+2.0	
—	+2.50	R. and L.

CASE 3 with fundus painting. Mrs. ANNA B.

March 27, 1901, aged 60 years. Complains of failing sight two years, especially during the last year.

Pupils equal and react.

R.v. = 6/60 with +1.0 D. sph. = 6/24.

L.v. = 6/36 with +1.50 D. sph. = 6/36.

Fundi.—Honeycomb choroiditis.

Ordered Mist. pot. iodide.

January 22, 1902. Rep. Mist.

Note added by the writer on appearance of fundus painting of R. eye.—The optic disc shows slight abnormality in its formation, below and to the nasal side. There is an irregular area of closely aggregated whitish exudates disposed around the macular region. The area of exudates is enclosed between the temporal branches of the retinal vessels. The immediate macular region is free of exudates. There is a small amount of fine cloudy pigment scattered around.

CASE 4 with fundus painting. Mr. EDWIN B.

February 9, 1910, aged 80 years. Complaining of bad vision for over 30 years and unable to read for over 20 years.

Vision both eyes = Counts fingers at 6 inches.

His mother had bad sight, and his two brothers were similarly affected.

Fundi.—The disc and macular areas are affected and show some scattered pigment. The affected area in each case is sharply defined from the surrounding normal choroid. In the area extending below the disc it is typically honeycomb.

Note added by writer on the appearance of fundus painting of R. eye.—The condition portrayed shows advanced change. There is central atrophy with absorption of exudate and exposure of choroidal vessels. On the temporal side there is still a band of raised confluent exudate with a definite margin. Above and

below in the course of the temporal vessels, discrete round exudate are still present around the margin of the affected area. Below and to the nasal side of the disc there is a larger patch of agglomerated discrete exudates.

Case (4) provided the eye for section and histological examination by Mr. Treacher Collins in 1913, which was the date of this patient's death at the age of 83 years.

CASE 5. Mrs. PRISCILLA H.

July 21, 1904, aged 47 years.

? Guttate choroiditis present (no further notes on this date).

April 14, 1909. Honeycomb choroiditis.

R.v. 6/60 with -3.50 D. sph. = 6/24 one letter.

L.v. 6/60 no improvement.

July 4, 1910. Vision 4/60 R. and L.

CASE 6 with fundus painting. Mrs. ELIZA P.

July 21, 1904, aged 49 years. Complains of gradual failure of sight for 9 years.

R.v. = $\frac{6}{60}$ with $\frac{-0.50 \text{ D. sph.}}{-6.50 \text{ D. cyl.}} = \frac{6}{60}$

L.v. = $\frac{6}{60}$ with -6.50 D. cyl. = $\frac{6}{18}$ one letter.

Visual fields fairly full.

Fundi show honeycomb choroiditis.

Writer's note on appearance of fundus painting of L. eye.—There is an extensive oval area of closely packed discrete exudates, extending laterally from the margins of the optic discs. There is a line of raised exudates along the margin of the temporal half of the optic disc. The macular region and an adjacent area together totalling more than two discs in size are free from exudates, but show the presence of haemorrhage and much local pigmentation.

CASE 7 with fundus painting. Mrs. MARY T.

November 19, 1913, aged 54 years, desires glasses for close work.

R.v. = 6/12. Pupils equal and active.

L.v. = 6/60 with -1.0 D. sph. = 6/18.

Fundi.—Honeycomb choroiditis in both eyes, more advanced in the left, where the macular region is more markedly involved.

Painting made of R. fundus.

September 2, 1914. R.v. = 6/12 one letter.

L.v. = 6/60.

Writer's note on appearance of fundus painting of R. eye.—The exudates are discrete and are mostly disposed in a small ring around and close to the macular area. The rows of exudates in the lower half of the ring, cover a wider area than those above. Some of the exudates are more clearly demarcated by pigment disposed around their margins. There is a larger irregular bunch of pigment close to the macula. Exudates are also present all round the margin of the optic disc.

Notes on writer's own examination.

December 10, 1935. (Patient is now 76 years.)

R.v. = 6/60 prefers +1.0 D. sph. but there is little improvement.

L.v. = counts fingers at 1 metre—not improved.

R. with +4.0 D. sph. = J.14. L. with +1.50 D. sph. = J.16.

Very early lens opacities in both eyes.

Fundi.—There is much exudate present in both fundi with branching and coarse pigment scattered around the disc and macular areas. There are yellowish exudates on the disc margins, but practically no choroidal change on the nasal side of the discs.

Blood pressure 162 mm. Hg systolic.

Urine, acid, S.G. 1020, no sugar or albumen.

January 8, 1936. Visual fields taken. Fields full.

Does not appreciate blind-spot areas.

CASE 8. Mr. ARCHIE H.

July 4, 1928, aged 45 years, Baker. Noticed slight defect in vision for 10 years.

R.v. = 6/12. L.v. = 6/18.

Fundi.—R. eye. Large area at macular region forming honeycomb type of choroiditis with some horizontal streaks of pigmentation. Similar collection of

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circular white areas on upper edge of disc and some areas scattered about on the nasal side. *L. eye* very similar area. Spots above the disc and down and in from it with large area over macular region, and fine lines of pigmentation.

Notes on writer's examination.

March 1, 1936. R.v. = $\frac{6}{12}$ part with $\frac{-0.50 \text{ D. sph.}}{-0.75 \text{ D. cyl. } \sqrt{105^\circ}} = \frac{6}{9}$ part.

L.v. = 6/24.

L. with $\frac{-1.50 \text{ D. sph.}}{+0.75 \text{ D. cyl. } \rightarrow 180^\circ} = \frac{6}{12}$ part.

with added +1.50 D. sph. R.v. = J.8. L.v. = J.10

Fundi.—Very typical honeycomb exudates in both, with some exudates on nasal side of discs. The exudates are becoming confluent in the central region. There is very little fine linear pigmentation present.

The fields are full. There is a small *paracentral scotoma* in each visual field continuous with the blind spot and extending towards, but not quite up to, the central fixation point.

CASE 8. ARCHIE H. 1° White. March 1, 1936.

CASE 9. FRANK H. 1° White. March 1, 1936.

CASE 9. Mr. FRANK H.

February 27, 1907, aged 29 years, Chairmaker. Complains of failing vision for one year especially in right eye.

R.v. = 6/60 not improved. Pupils equal and active.

L.v. = 6/24.

R. fundus.—Large black patch of interlacing pigment at right macula with numerous white spots below, and some also under cover of the pigment.

L. fundus.—The left macula is similar to the right but shows much less pigment, and the white dots beneath are more visible, and more numerous around the pigment area. There are also a few on the nasal side of each disc. The vessels are fairly normal and run over the pigment.

Urine, nil abnormal found. Ordered Mist. pot. iodide.

July 26, 1907. Rep. Mist.

Notes on writer's examination.

March 1, 1936. Patient now 58 years.

R.v. = $\frac{6}{60}$ with difficulty with $\frac{-0.25 \text{ D. sph.}}{+1.50 \text{ D. cyl.}} = \frac{6}{60}$
 $\downarrow 65^\circ$

L.v. = $\frac{6}{60}$ with $\frac{-1.0 \text{ D. sph.}}{-1.50 \text{ D. cyl.}} = \frac{6}{36}$
 $\downarrow 120^\circ$

With addition of +2.0 D. sphs. = J.12.

There are early senile lens changes in both eyes especially in the right.

The fundi show changes very similar to the description of February 27, 1907, with a small amount of exudation on the nasal side of the discs.

The visual fields are full. There is a crescentic paracentral scotoma in each field, more marked in the left, and consisting of extensions above and below from the blind spot areas.

CASE 10. Mr. TOM B.

February 17, 1904. There are well marked fundus changes. Both optic discs are surrounded with a "honeycomb frame" with some pigment present. The maculae appear involved. In the right eye the changes at the macula almost assume a circinate ring.

R. and L. vision = 6/9.

CASE 11. Mr. FREDERICK B. Died in March, 1934.

January 27, 1904, aet. 38 years. Blacksmith.

R.v. = 6/9.

L.v. = 6/6. There are distinct white spots in both macular areas.

April 4, 1906. R.v. = 6/12 with +1.0 D. sph. = 6/9.

L.v. = 6/12 with +1.0 D. sph. = 6/9.

L. fundus shows spots around the L. macula especially down and in, arranged in a broken circle.

R. fundus shows spots around the R. macula more extensive than in the left.

December 31, 1931. Complains that vision in right eye has become poor for several months.

R.v. = 6/60 with +1.50 D. sph. = 6/24 two letters.

L.v. = 6/12 one letter with +1.0 D. sph. = 6/9 one letter.

R. fundus. At the upper edge of the disc is a small collection of honeycomb spots. In the macular region there is a very gross area of honeycomb choroiditis, and what looks like some very fine haemorrhages in the centre of the area, and also on the lower and inner edge.

L. fundus shows a white hyaline area down and in from the edge of the disc and some more on the outer margin. There is a gross area at the macula but no typical honeycomb appearance.

January 27, 1932. R.v. = 6/60 with +1.50 D. sph. = 6/36.

L.v. with +1.0 D. sph. = 6/9 part.

February 24, 1932 } *In statu quo.*

February 9, 1933 }

March 14, 1934. Died.

CASE 12. Mrs. ROSE J. Widow—no children.

July 10, 1903, aged 45 years.

R.v. = 6/12. L.v. = 6/24 one letter.

Honeycomb choroiditis both eyes.

Notes on writer's examination.

March 4, 1936. Patient is now 78 years.

R.v. = counts fingers at 2 metres.

L.v. = variable P.L. Poor projection. Calcareous opaque lens.

R. lens shows a few peripheral opaque sectors.

Under H. and C. refraction + 2.50

+ 2.50

R. fundus shows a large area of confluent exudates in the disc and macular region, with very little pigmentation at all. The plaque of confluent exudation shows a central irregular area free from all exudate. There is marked sclerosis of choroidal vessels to be seen in this central free area, and also around the margins of the plaque of exudate, especially below.

CASE 13. MRS. MARIA B.

March 25, 1909, aged 61 years. Complains that sight in right eye has been failing for one year.

R.v. = 6/60 with - 1.0 D. sph. = 6/36. R. lens changes are present.

L.v. = 6/12 with + 1.0 D. sph. = 6/6 two letters.

The fundi show much choroiditis on the outer side of the discs of the honeycomb type with superficial pigmentation.

November 1, 1911. R. lens becoming more opaque, with barely any view of fundus.

July 31, 1912. R.v. = 6/60. L.v. = 6/24 one letter.

There is recent haemorrhage at the left macula.

June 10, 1914. R.v. = 6/60. L.v. = 6/18 with + 0.75 D. sph. = 6/12.

August 16, 1916. L.v. 6/36 with + 2.0 D. sph. = 6/12 part.

April 23, 1919. R.v. = shadow perception. L.v. = 6/60 with gl. 6/12 one letter.

L. fundus shows fine exudates on the outer side of the disc. The honeycomb condition extends to the disc margin. There are some spots on the upper disc margin and on the inner side.

March 12, 1930. L. eye shows some peripheral lens changes.

R.v. = P.L. L.v. = appreciation of hand movements.

L. fundus. The area of exudation is now surrounding the disc and sends finger-like processes onto the nasal side.

Notes on writer's examination.

April 18, 1936. Patient is now 88 years.

R. eye. Mature cataract. L. eye counts fingers at 1 metre eccentrically.

There is a small lenticular opacity on the nasal side.

L. fundus shows a large confluent area of exudation on both sides of the optic disc with a little scattered pigment.

There is a definite margin to the area of exudation on the nasal side of the disc. On the temporal side the exudates form a yellowish plaque extending between the temporal vessels.

CASE 14. GERTRUDE A. 1° White. January 22, 1936.

CASE 14. Mrs. GERTRUDE A. Widow—no children.

August 15, 1923, aged 30 years. Complaints of aching top of head, worse on close work.

R.v. = $\frac{6}{12}$ with $\frac{+0.50 \text{ D. sph.}}{-0.75 \text{ D. cyl.}}$ v. = $\frac{6}{180^\circ 6}$ Pupils equal and active.
 L.v. = 6/5 with +0.50 D. cyl. = 6/5.
 $\downarrow 90^\circ$

Fundi.—Choroidal exudates in both fundi especially around optic discs.

May 21, 1930. R.v. = 6/60. L.v. = 6/9.

Gross central changes in both eyes especially affecting the right macular region.

Blood *Sigma* for syphilis—Negative

Ordered French iodine minims iii in milk b.d.

August 24, 1932. R.v. = 6/60

L.v. = 6/12 two letters with glasses } Fundi unchanged.

March 7, 1934. L.v. with glasses 6/36 one letter.

L. fundus shows recent additional exudate between disc and macula with small haemorrhage on it.

June 27, 1934. *L. fundus* unchanged, one old haemorrhage seen.

October 31, 1934. L.v. with glasses = 6/24 two letters.

No haemorrhage seen to-day.

Notes on writer's examinations.

January 22, 1936. R.v. = $\frac{6}{36}$ with $\frac{-1.0 \text{ D. sph.}}{+1.25 \text{ D. cyl.}}$ = $\frac{6}{36}$
 $\downarrow 115^\circ$

L.v. = 6/60. Not improved.

For reading with R. $\frac{+1.0 \text{ D. sph.}}{+1.25 \text{ D. cyl.}}$ and L. $\frac{+1.0 \text{ D. sph.}}{+1.0 \text{ D. cyl.}}$
 $\downarrow 115^\circ$ $\downarrow 90^\circ$

Binocularly J.12.

R. light sense with George Young's Album = 4th class (1/16).

L. " " " " " " = 5th class (1/8).

Fundi.—No recent haemorrhages are seen.

There are gross confluent exudates in both macular regions, extending between the upper and lower temporal vessels, with coarse pigmentation. There are some discrete spots of exudate on the nasal side of the optic discs especially in the right fundus, and also on the margins of the discs.

April 1, 1936. Systolic blood pressure 135 mm. Hg.

June 3, 1936. Condition unchanged. The visual fields are full. The right field shows a paracentral scotoma surrounding the temporal side of the fixation point. The left field shows enlargement of the blind spot.

CASE 15. New case discovered. Mr. JOHN B. With fundus painting.

April 23, 1936, aged 44 years. Engineer.

R.v. = 6/5 part | Light sense 1st class in both eyes. Colour thresholds, 1st

L.v. = 6/5 full | class in both, but more readily in left eye.

Visual fields full, no scotomata.

The fundi.—There are round colloid bodies along the margins of both optic discs, deep to the retinal vessels, and mostly grouped along the upper edge of the optic discs. There is some scattered irregular coarse pigment between the discs and the maculae, small in amount, but more marked in the right fundus. Around the deeply pigmented spots is a fainter cloudy pigmentation interspersed with discrete hyaloid dots. In the right fundus the hyaloid spots and faint cloudy pigment seem to extend almost up to the macula. In the left fundus the macula is less encroached upon. There are about three tiny pale yellowish spots on the nasal side of each optic disc.

No peripheral disturbance is visible in either fundus.

CASE 16. Mrs. JULIA V.

August 5, 1896, aged 46 years. Complaints of impaired vision 2-3 years.

R.v. = 6/60. L.v. = 6/6.

There are seven members in her family of whom six have affected sight, and one sister seen has the same condition as this patient.

Own note.—Fundus paintings of this case seen at the Oxford Eye Hospital show a well developed "honeycomb" picture.

CASE 15. JOHN B. 1° White, April 23, 1936.

CASE 17. LAVINIA N. 1° White. January 8, 1936.

CASE 17. Mrs. LAVINIA N.

Sister of case (18). No other brothers or sisters, and both parents are dead.

October 4, 1933, aet. 50 years. Difficulty in reading for one year. States that vision in left eye has always been poorer than the right.

R.v. = 6/12. L.v. = 6/60.

Both fundi show Doyne's choroiditis.

February 13, 1935. R.v. = 6/36. L.v. = 6/60.

Notes on writer's own examinations.

October 2, 1935. R.v. with $+1.25$ D. sph. = $\frac{6}{36}$ one letter.
 $+0.25$ D. cyl. $\downarrow 70^\circ$

L.v. with $+1.25$ D. cyl. = 6/36 one letter.
 $\downarrow 90^\circ$

With addition of $+1.25$ D. sph. J.16 both eyes.

Media are clear.

Fundi.—Deep physiological cups. There are aggregated, discrete, putty-coloured, exudates in both fundi giving a typical honeycomb appearance, especially in the right eye. The exudates are surrounded by shadow rings, with additional scattered, large, coarse, pigment patches. The exudates also extend all round the disc margins.

January 8, 1936. Visual fields are full. The right field shows a large pentagonal paracentral scotoma, extending from the blind spot area, above and in close proximity to the fixation point.

February 5, 1936. Light and colour thresholds with correction for reading distance.

R. light sense 3rd class (1/32)—Colours	Red	3rd class	(1/16).
	Blue	2nd	„ (1/64).
	Yellow	1st	„ (1/32).
L. light sense 4th class (1/10)—Colours	Red	4th class	(1/8).
	Green	3rd	„ (1/16).
	Blue	3rd	„ (1/32).
	Yellow	4th	„ (1/4).

CASE 18. Mrs. SARAH M. Sister of case (17).

October 4, 1933. Has less marked but similar condition to her sister.

Died in May, 1935.

CASE 19. Mr. CHARLES S. Brother of case (20).

May 24, 1916, aged 51 years. Boot repairer. Complains of impaired vision for distance and near.

R.v. = 6/36. L.v. = 6/36.

L. fundus.—There are two round white spots on the inner edge and one on the outer edge of the optic disc; and a rather extensive area of choroidal disturbance with pigmentation in the macular region. The honeycomb condition here is not so well marked as in his brother's case.

R. fundus shows one spot on the inner edge and three on the outer edge of the disc. The macula is also similarly affected.

June 5, 1918. R.v. = 6/36 with -1.0 D. sph. = 6/24.

L.v. = 6/60 with -1.0 D. sph. = 6/60.

Patient died in April, 1919.

CASE 20. Mr. HENRY S.

May 24, 1916, aged 49 years. Farm labourer. Complains of difficulty in reading.

R.v. = 6/24. L.v. = 6/12.

R. fundus extensive area of honeycomb choroiditis with some pigmentation, and a little haemorrhage just above the macula.

L. fundus is similar but not so extensive and also shows some spots on the nasal side of the disc.

November 28, 1917. There is now an extensive area on the nasal side of the disc in both fundi.

November 26, 1919. R.v. = 6/18 part with -1.0 D. sph. = 6/12 part.

L.v. = 6/18 part with -0.75 D. sph. = 6/12.

R. fundus.—The honeycomb area extends about 2 discs breadth to the inner side, and from the superior to the inferior temporal vessels on the outer side; and about 1½ discs breadth beyond the macula.

L. fundus is very nearly as extensive as in the right eye.

March 7, 1924. Condition unchanged.

July 25, 1928. R.v. = 6/60 with -1.0 D. sph.

L.v. = 6/24 with -1.0 D. sph. = 6/18.

Very extensive and typical honeycomb choroiditis in both eyes. The right fundus shows a good deal of pigmentation.

October 22, 1930. R.v. = 6/60. L.v. = 6/60 with -1.0 D. sph. = 6/24.

Gross honeycomb choroiditis.

R. eye very heavily pigmented.

L. eye more atrophic looking.

CASE (A). WILFRED H.

March 8, 1936, aged 13½ years.

R.v. = 6/5. L.v. = 6/5.

Threshold tests, 1st class both eyes for light and colours.

H. and C. instilled.—Nil abnormal seen in fundi. Discs, vessels, and maculae normal.

CASE (B). VERA H.

March 8, 1936, aged 10 years.

R.v. = 6/5. L.v. = 6/5.

Threshold tests, 1st class both eyes for light and colours.

H. and C. instilled.—Nil abnormal seen in fundi. Discs, vessels, and maculae normal.

CASE (C). Mr. GORDON H.

March 8, 1936, aged 31 years. Chairmaker. Married two years. No children.
R.v. = 6/5. L.v. = 6/5.

Fields full. Does not appreciate blind spots.

H. and C. instilled.—Nil abnormal seen in fundi. Discs, vessels and maculae normal.

CASE (D). Mrs. LOUISA MARY E.

March 6, 1936, aged 58 years. Wearing for two years -0.75 D. sphs. R. and L.
R.v. = 6/24 two letters, with gl. = 6/9. L.v. = 6/18 three letters, with gl. = 6/6.
Nil abnormal seen in fundi.

CASE (E). Mrs. ANNIE H.,
September 15, 1920, aged 39 years.

$$\text{R.v.} = \frac{6}{60} \text{ with } \begin{array}{l} -3.25 \text{ D. sph.} \\ -1.50 \text{ D. cyl.} \end{array} = \frac{6}{9} \quad \downarrow 80^\circ$$

$$\text{L.v.} = \frac{6}{60} \text{ with } \begin{array}{l} -3.50 \text{ D. sph.} \\ -1.25 \text{ D. cyl.} \end{array} = \frac{6}{9} \quad \downarrow$$

Notes on writer's examination.

	-2.50		-2.25
Refraction	-5.75		-4.0

March 6, 1936. R. with $\begin{array}{l} -4.0 \text{ D. sph.} \\ -3.0 \text{ D. cyl.} \end{array} = \frac{6}{9} \text{ one letter.} \quad \downarrow 100^\circ$

L. with $\begin{array}{l} -3.25 \text{ D. sph.} \\ -2.0 \text{ D. cyl.} \end{array} = \frac{6}{6} \text{ part} \quad \downarrow 90^\circ$

Binocularly 6/5 with addition of +0.25 D. sph. +1.75 D. sphs. added for reading. Nil abnormal seen in fundi.

CASE (F). Mrs. MARTHA W.

March 6, 1936, aged 46 years.

Wearing R. -3.50 D. sph. L. -3.75 D. sph.

R.v. = 6/60 with gl. = 6/6 one letter. L.v. = 6/60 with gl. = 6/6 one letter. Nil abnormal seen in fundi.

CASE (G). Mr. JAMES WILLIAM E.

April 26, 1936, aged 26 years. Gardener.

Wearing glasses for about six years.

$$\text{R. } \begin{array}{l} -3.25 \text{ D. sph.} \\ -1.75 \text{ D. cyl.} \end{array} \rightarrow \text{L. } \begin{array}{l} -3.0 \text{ D. sph.} \\ -1.25 \text{ D. cyl.} \end{array} \quad \downarrow$$

R.v. = 6/60 with gl. = 6/6 one letter. L.v. = 6/60 with gl. = 6/6 four letters.

Visual fields full, no scotomata. Nil abnormal seen in fundi.

Light and colour thresholds.

R. with glasses. Light threshold, 3rd class. Colours, green, 2nd class.

Other colours, 1st class.

L. with glasses. Light threshold, 3rd class. Colours, blue and green, 2nd class. Other colours, 1st class.

CASE (H). Mr. CEDRIC H.

April 26, 1936, aged 28 years. Clerk.

Wearing glasses since about 10 years of age. Present glasses since November, 1935.

$$\text{R. } \begin{array}{l} -13.0 \text{ D. sph.} \\ -3.0 \text{ D. cyl.} \end{array} \rightarrow \text{L. } \begin{array}{l} -13.0 \text{ D. sph.} \\ -3.50 \text{ D. cyl.} \end{array} \rightarrow$$

R.v. 6/60, with gl. = 6/9 part. L.v. 6/60, with gl. = 6/9 part.
Fundi show myopic crescents, but nil else abnormal seen.
Visual fields are full, with ? slight enlargement of blind spots.
Threshold tests with glasses.
 R. White, 2nd class. Colours, all 1st class.
 L. White and colours, all 1st class.

CASE (g). WILLIAM E. 1° White. April 26, 1936.

CASE (h). CEDRIC H. 1° White. April 26, 1936.

CASE (i). MR. TERENCE GILBERT W.
 January 18, 1933, aged 19 years.
 Seen with conjunctivitis and purulent rhinitis.
 Nil else recorded.

CASE (j). MR. HARRY B.
 April 30, 1936, aged 78 years.
 States R. vision failing 9-10 years. L. vision failing 2-3 years.
 R.v. = no P.L. L.v. = shadow perception.
 Both optic discs show deep glaucomatous cupping.
 R. eye myopic, with vitreous haze. L. eye shows a few lens opacities.
 No choroidal exudates in either eye. Tension + in both eyes.
 Shallow anterior chambers, pupils slightly dilated and fixed, corneae clear,
 no pain.

CASE (K). HARRY B.

April 30, 1936, aged 14 years.

R.v. = $\frac{6}{9}$ two letters, with $\frac{-0.50 \text{ D. sph.}}{-0.50 \text{ D. cyl.}} = \frac{6}{5}$
 $\downarrow 30^\circ$

L.v. = $\frac{6}{12}$ with $\frac{-0.25 \text{ D. sph.}}{-0.25 \text{ D. cyl.} \rightarrow 180^\circ} = \frac{6}{5}$

Thresholds, 1st class for white and colours in both eyes.

Nil abnormal seen *in fundi*. Visual fields full and normal.

CASE (K). HARRY B. 1° White. April 30, 1936.

CASE (L). ALAN B.

April 30, 1936, aged 8 years.

Vision 6/5 each eye. Nil abnormal seen *in fundi*.

Thresholds, 1st class for white and colours in both eyes.

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