To the Dean of the Faculty of Medicine

Dear Sir,

I herewith enclose my thesis for the degree of Doctor of Medicine, in accordance with your kind permission to send it in one week after the 30th of April.

I beg to request that any communications may be forwarded to me at the above address. I am

yours faithfully,

J. H. Batchelor

104 Guilford St.
May 6th, 1895.
Thesis for the Degree of Doctor of Medicine

Actinidia pigmentosa:

with notes on twenty-one cases.

by

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Blindness. Those members of the family who had passed the age of puberty without being affected, remaining free from the disease.

The second case is that recorded by Dr. Samuel Linnier, in a pamphlet published at Ghent in 1838. He relates the history of a family in which night-blindness, followed by slowly progressive failure of sight, existed for one or two centuries, the disease being confined to the descendants of one ancestor.

Retinitis pigmentosa is essentially a family disease; that is, it is a disease which very frequently affects more than one member of a family in the same generation. Like leprosy, hemophilia, Vincent's disease, and colour blindness, it may go on manifesting itself in a few members of each successive generation for several centuries. It is not a very rare disease. Half of Boston reports 29 cases out of 12,000 eye patients giving a proportion of 1 in 411.4 of patients. Bowen gives the proportion as about 1 in 1200.

Poincy states that from over 100,000 recorded hospital cases he found the proportion of retinitis pigmentosa to be 1 in 1300. Lee found, out of 520 new cases attending the Liverpool Eye Infirmary in 1881, only 3 cases of this disease, giving 1 in 2080. If we take an average from these cases we find that it occurs in about 1 in 1200 cases of eye disease equal to 0.084 per cent. It is much more common among Aranologists, and in Turkey and India than elsewhere.
This disease is almost invariably found in both eyes, though cases have been recorded in which only one eye has been affected; with regard to these unilateral cases, Hutchinson recommends always to enquire for a history of injury. He states that he has seen the retina almost black from the confluence of stellar pigment accumulations resulting from a blow.

The disease is extremely chronic in its course. Its most characteristic symptom begins in childhood; it becomes troublesome about puberty and slowly advances to almost total blindness late in life. As may be supposed, it is usually neither suspected nor discovered until some time after birth. Occasionally the symptoms are very late in manifesting themselves. Hollester records a case in which failure of sight did not become apparent until the age of 35, whilst Proctor records another in which the first symptoms occurred in the 50th year.

The disease may extend over many years from the commencement, until total blindness supervenes. Bradenell states that disorders give the duration of the disease as about 20 years, whilst Hutchinson and also Proctor as from 20 to 50 years. Siebert states that total blindness supervenes after the 30th or 40th year. Facts that it usually occurs late in the fifth decade, whilst de Blickeher & Sandell assert that poor patients seldom retain
much vision after 40, whereas in the rich we
often find patients of 50 or 60 years of age will
fairly good central vision.
In the number of cases of complete blindness that
are that condition to this disease, Wagner of
Puebla gives the percentage as 1.26 in a table
published on 2528 cases, whilst Oppenheim
obtained nearly the same result after the terminus
of 3.72 of the totally blind of the City of New York.
The found that 1.39 per cent of cases were due to
retinitis pigmentosa.
This disease is much more frequent in males than
in females. Out of 185 cases reported by Lieben,
111 were those of males and 74 those of females,
the proportion being 2:1 to 1. Hand states
that in his experience it occurs three times more
frequently in boys than in girls.

Symptoms:—The most characteristic symptom of
this disease is night blindness, the patient to
appreciate differences of intensity of illumination being
impaired, and the vision in subdued light being
diminished. There appears to be some uncertainty
as to the correct technical term which should be
applied to this condition. The choice lies between
hemeralopia and nyctalopia. The generally
accepted derivation of the terms is as follows:—
hemeralopia from Ἕμηρα, day, & ὕπατος, right eye, equivalent to
right blindness, and nyctalopia from νύκτα, night and
Up, right, equivalent to dry blindness.

So Greenhill's, in a learned and critical dissertation on the meaning of these terms, asserts that the true meaning of *aetetalopia* is right blindness and not right—right, that the word is distinctly used in this sense by Galen, as also by Posthum, Aristotle, Pliny, and nearly all the old medical writers. He admits that the use of the term by Hippocrates is somewhat ambiguous, and that in two of his passages, though there are no doubtful authorities, he uses the word in the uppermost sense.

Fralow upheld this opinion of Dr. Greenhill's, in an essay on the meaning of the terms as disclosed by the diseases described under those headings by the ancient and modern writers. He actually goes so far as to assert that the *aetetalopia* of Hippocrates was really right blindness, the negative having been omitted. The further states that up to the end of the 18th century, writers were unanimous in using *aetetalopia* to mean right blindness; but from then on to the beginning of the 19th century was a transition stage, left terms being used indiscriminately, to describe the same condition, right blindness; since then, however, hemeralopia has come into almost universal acceptance, nearly every writer on ophthalmology for the last century and a half using this term instead of *aetetalopia*, to signify right blindness.

Taking this into consideration, and also the facts that
The published books by Hippocrates, and the definition of the terms in nearly every medical and scientific source agree with this; there appears to be sufficient grounds for adhering to the more popular term. Perhaps the simplest method of solving the difficulty and of avoiding discussion would be to abandon the Greek terms altogether and to use instead the English equivalents, "night-blindness" or "day-blindness" which are sufficiently clear and expressive for all purposes.

Patients suffering from this condition of night-blindness complain that they cannot see well in the dark or in twilight, nor when the illumination in a room is reduced. This symptom develops early in life, but increases with age until patients are unable to trust themselves out alone at night, although in the daytime and in a good light they are not much inconvenienced, and are able to find their way about without difficulty.

With regard to the cause of this condition, Keilh" showed by his investigations that it is not due to a disturbance of the sense of light, but rather to a defective power of retinal adaptation, the pathological changes lying not in the nerve centers of vision but probably in the retinal epithelium from which the visual purple is derived. "Ehren" agrees with this view, but on the other hand "Belling" attributes it to defective light sense, due to changes in the pigment epithelium layer.
Leber's is a disease that in some cases the opposite condition of hypoaesthesia is present, in which the patient sees better at night time than in the day, owing to some hypoaesthesia of the retina which makes it extremely sensitive to strong light. Cases of this nature have been recorded by Hutchinson, Reese, and others.

Another almost constant symptom is that of progressive diminution of the visual field. At first, taken in a good light it may be nearly normal, whereas if the light be reduced it becomes very much contracted, the peripheral portion of the retina, owing to atrophy of the nerve elements, loses its function, becomes so to speak under-sensitive, and does not respond to feebly illuminated images, admitting the conduction of strong impressions only. As the atrophy advances the retina becomes less and less sensitive, until ultimately it is incapable of being stimulated by even the strongest light. The field then becomes permanently contracted, so that with relatively good central vision the patient may be able to read but not able to find his way about alone, even in the daytime.

On account of this loss of lateral perception, patients are obliged to bring their glotic areas to bear directly upon an object in order to see it. They thus develop a continual restless movement of the eyes, turning them to the right and to the left, upwards and downwards, in order to examine every
individual object and to take in as much as possible of what is going on around them. This movement is continuous and cumulative as in aphylaxis.

As the disease progresses, central vision becomes involved also, and total blindness supervenes.

Being records a case in which a small temporal portion of the field retained light perception after the function of the rest of the retina was lost.

The disease may be unattended by any other symptom though sometimes subjective luminous sensations manifest themselves; also a feeling of weight and tension deep in the orbit, which seems to almost forcibly close the eyelids.

Colour vision may remain good even in cases where the visual fields are greatly contracted, though according to de Blicot et Landolt it is always impaired and never perfect.

As regards refraction, the disease occurs in myopes, hypermetropes, and in astigmatismis in variable proportion. Myopes suffer more than hypermetropes, for the myope requires to bring the object close to his eyes to see it clearly, whereas with a contracted visual field the further off the object is held the larger the extent of surface seen. There is thus a continual struggle between the accommodating powers of the eye, while many result in the development of a hypermetropic posture.
Ophthalmoscopic appearances:— These consist in a deposit of pigment in the inner layers of the retina, in a telangiectatic and atrophic state of the retina with obliteration of vessels, and in secondary atrophy of the optic nerve.

The deposit is of a coal black colour and is arranged in branching delicate streaks usually around the periphery of the retina and gradually advancing towards the macula and disc. The deposits usually commence on the inner side of the retina and it is here that the accumulation is always greatest. It occasionally commences in a narrow annular near the equator of the eyeball and advances inwards towards the disc. There may thus be left a small eccentric portion of the field in form of a narrow peripheral zone or belt which retains some perception of light after central vision is destroyed.

In some cases the pigment is arranged in a broad zone midway between the centre and periphery of the fundus, giving a ring shaped appearance, as described by von Grafe, Michel, and Binder. In it may be grouped chiefly in the macular region, forming a central dejection with good peripheral vision, as described by Belin and Hefzi. Rarely however, deposits of these cases in which annular and central dejections have been described, are really true cases of retinitis pigmentosa at all, he is inclined to think that there is some error in diagnosis.

The pigment is deposited in the inner layers of the retina. This is favored by the fact that the retinal
vessels are frequently seen crowded by the pigment spots. The spots being in front of the vessels, not underneath them. The pigment spots are usually also large, in shape with branching processes which in advanced cases assume a more or less like arrangement forming a kind of network or lace-like arrangement stimulating the appearance of the lacuna and canaliculi in the Descrimin system of bone as was first remarked by Dietl. In some cases the spots are round or irregular, or they may be punctiform, coarsely granular, or arranged in streaks; the pigment is most frequently deposited along the course of the retinal vessels. Often clinging to the vessel walls like litter to the branches of a tree. A very favourite spot is just at the bifurcation of a vessel. Furtwängler is of opinion that the spherical form of the pigment merely results from its deposit in the rounded vessels or vessels, and that it is as peculiar to retinitis pigmentosa.

The degree of blindness cannot be inferred from the extent of pigmentation. Dietl asserts that the pigmentation actually diminishes in the later stages of the disease. He states that as the disease progresses the affected area becomes more extensive, but the pigmentation increases in the area of the fundus first affected. Furtwängler, however, does not hold with this at all. He says that up to a certain point an increase does take place in the pigmentation of the area affected,
and he also considers that the rule is for the amount of pigment to increase rather than to diminish with the duration of the disease.

The presence of pigment is not absolutely essential for the diagnosis of the disease. Some cases occur in which it is quite absent, and many in which it is very small in amount. The sclerosis and atrophy of the retina taking place in a otherwise typical manner. Seitz first described cases of this nature, terming them anomalous forms of retinitis pigmentosa. Later have also been recorded by Heitzig, Altman, Buell, Germinati, and many others.

Pratt, in a microscopic examination of a case of this nature, found the retinal tissue to be infiltrated with a small quantity of finely divided pigment in its deepest layers only, where it could not be recognized with the ordinary microscope.

Certain peculiar cases of retinitis punctata albinens, described by Gage and Dot, in 1893 agree in all respects with retinitis pigmentosa, but show instead a pigmentation of the retina, a number of minute, discrete, pinprick dots, distributed near the fovea in a fairly regular manner, though chiefly between the equator and the macula region.

Prance first described cases under this title, but his differed essentially from the above in as much as there was no night blindness, and the cases improved under treatment. Lobeski records five cases, and Belcher, one, of a precisely similar nature.
to time of gazet. and Doc. In Adie's case, he
observed that the dot eventually became pigmented and
atypical retinitis pigmentosa resulted.
In some cases of aphakic choroid-retinitis, pigment
can wander from the pigment epithelium layer into the
retina, and may be seen occasionally with a
retinal vessel running beneath it. The pigment
can even assume the true corpuscular shape, rendering
the diagnosis very difficult, though usually it is
not so equally distributed as in the true disease,
and there is generally some further change associated
with it, such as spots of choroidal atrophy, old
clots, 

The optic disc becomes atrophied, flat, grayish-yellow
or grayish-white in color, but never pure white. It
is described as a waxy dot by Rokitansky, and as a
dead leaf color (pallide mortis) by Panum.
The vessels, arteries as well as veins, become diminished
in size and narrowed. They may be represented
by fine lines only. Often a progressive diminution
in the calibre of the vessels is one of the earliest
signs revealed by the ophthalmoscope.

The retina shows a grayish light everywhere from telereosin
of the connective tissue elements. In some cases
irregular patches of a brighter aspect are seen in
the fundus. This is due to the electrolyte shining
through the choroid, owing to the defective
pigmentation of the hexagonal cell layer. The
choroidal vessels with their pigmented intercrosses are
more easily seen, and appears of a deeper red, while the fundus has a biviscidated, wrinkled, or mottled appearance.

Retinal vascular abnormality is a slowly progressive growth, usually develops after the disease has lasted some time. Landolt assigns its formation to the abnormal state of nutrition of the vitreous. As to its frequency, one great says that it occurs in about a third of the cases, and Brown found it present in 12 cases out of 64.

Vitreous opacities may occur owing to the escape of pigment into the vitreous, carried there by leucocytes from the pigment epithelium layer, its cells becoming disintegrated and the pigment escaping. The presence of flakes or threads strongly colored with pigment are thus explained by Russe, who states that the superficial layers of the vitreous condense and adhere in places to the retina, following on which occurs an infiltration of pigment cells. Jude accepts that in true retinal pigmentation there should be an obvious specificity.

Brown found only 3 cases with opacities out of 64 of the disease.

Pathology:—The layers of the retina chiefly affected are the perceptive and pigmentary layers, namely, the layer of rods and cones, and the hexagonal pigment layer. There is a hypertrophy or thickening of the connective tissue elements with a simultaneous
destruction of the nerve elements, chiefly in the outer layers of the retina, the nerve fibres being comparatively little affected. The retina thus becomes thinned out in its whole extent and closely adherent in places to the choroid. On a microscopic examination of a typical case, found that at three or four places where the retina was most deeply pigmented, it was firmly united to the choroid by melanin.

Sagittal sections of the inner surface of the retina, a definite layer of thickened connective tissue with numerous small aneurysms and detachments forming spaces which were filled with amorphous albuminous material. The basilar layer was wanting and replaced by newly formed fibrillated tissue.

The layer of rods and cones becomes atrophied and may almost entirely disappear, but in places where they persist such as in the immediate neighborhood of the vessels, they are seen to be short and clubbed at their ends. In a case examined by Leber, the rod layer had entirely disappeared, its place being occupied by pigment cells and large mucous processes from the elastic lamina.

The pigment in the retina is derived from that contained in the pigment epithelium layers, which layer consequently becomes much altered, and in places is quite devoid of pigment. The pigment migrates from the hemispherical cells into the nerve fibre layer of the retina, being usually deposited in the perivascular connective tissue, where it probably increases by proliferation.
Donder's found a delicate, irregular, network of pigment penetrating all the layers of the retina which contain vessels, the pigment generally surrounding the vessels. But the deeper layers of the retina appeared normal and free from pigment, and the roth layer was mostly wanting, or seemed to be lost in a thickish layer of exudation. He considered the pigment to be developed in the retina from a chronic process of inflammation, a chronic retinitis in fact.

Junge observed that whenever pigment was deposited around a retinal vessel, all the layers of the retina which should exist between the vessels and the choroidal epithelium were destroyed.

Schweigger found pigment in all the layers of the retina, and formed the opinion that the deposit around the vessels was only met with in those places where the sclerosis of the retinal connective tissue, which is the main factor in the disease, had already taken place. He concluded that the entire amount of pigment originated in the pigment epithelium layer, and that its presence in the retina was dependent on a choroiditis. He found no constant relation between the quantity of the pigment and the degree of retinal atrophy. He appeared to be coincident consequences of the choroidal process. A considerable thickening of the interior of the vessel walls takes place, with a consequent narrowing of their lumens, leading to their obliteration. In advanced cases the vessels are converted into mere strands of connective tissue.
Kirschner, found the walls of nearly all the larger bloodvessels, as well as their finer ramifications, infiltrated with pigment cells. A few non-pigmented, yellowish lymphoid vessels were seen as well.

In Bagean's case, the cells of the iris' inferior retina were much pigmented, and had undergone great pigmentation. On their outer surface, between the iris and the choroid, was a thick layer of connective tissue extending backwards as far as the ora serrata.

The choroid was universally thickened, owing to an increase in the intervascular stroma, the vessel walls, particularly the outer and inner coats, were much hypertrophied, and the lumen of the vessels more or less obliterated. The choroid capillaries as a definite layer had disappeared.

The optic nerve became atrophied secondarily, owing to the thickening of the inter-papillary trabeculae and branching of the nerve fiber bundles. The smaller vessels in the nerve show hypertrophied walls and narrowed lumen.

The vitreous humor to the retina became altered in structure, infiltrated with colorless blood cells and with pigment granules. Occasionally it becomes fluid. The lens may show degeneration of its fibers at the posterior pole.

Dandie, in 1898, described the disease to a very chronic form: insidious, of the retinal vessels, comparing the eruptive process to that which takes place in tuberculos of the liver and kidneys. How common it is...
considered that the disease is essentially one of the choroid, and that it is the change in it which primarily causes the pigment to wander into the retina from the epithelial layer. This was clearly demonstrated by Hagenmuller in his experimental researches on the nutrition of the retina in rabbits by section of the ciliary vessels, anterior central retinal vein, and optic nerve, both separately and in combination. Previous experiments had been made by Berlina, Seilhe, Vries, etc., in dividing the optic nerve, but in every case the ciliary vessels were divided at the same time as the nerve.

Hagenmuller found that section of the long and short posterior ciliary arteries on one side, caused a grayish-white opacity of the retina on that side, with migration of pigment cells into the inner fiber layer. The pigmentation occurred five days after section, in form of small pigment spots which rapidly increased and became aggregated together, forming clusters. The pigment in the first instance being carried by lymph cells from the pigment epithelium layer and afterwards increasing by proliferation.

The appearance was like that of old choroid-retinitis, with absence of pigment in the pigment epithelium layer, so that the choroidal vessels became very distinct. They appeared pale and contracted.

After the fourth week the affected part of the retina began to show signs of atrophy.

Microscopically, the choroidal vessels were seen to be
enlarged or only partially filled, the blood columns being interrupted or subdivided. There was a fine granular opacity over the retina. The rods and cones were further affected, and between them were many fine granules of pigment. Neutrophils were seen in the retina, filled with eosinophilic material. In some pigment granules, pigment cells could be seen. Neutrophils are like the pigment epithelium into the retina. Eosinophilic filaments were observed floating in the vitreous.

Sections of the optic nerve with the ciliary vessels show the retina and the connective tissue of the retina, and no emigration of pigment.

In cases of embolism of the central artery, where the ciliary vessels are not affected, no emigration of pigment occurs, but retina of the retina taken place from circulation through the optic. Buhleboi reports a case where after 2 months embolism of the central artery, the layers of the retina remained quite unchanged except for the dense density.

These experiments seem to point to retinitis pigmentosa being a vascular disease, from the fact that the pigment epithelium layer being supplied by the ciliary vessels, and any interference with the blood supply causing emigration of pigment into the retina.

Hagmaner et al. have found that atrophy of the retina, and the changes in pigmentation, are to be accounted for by the disturbance of the circulation in the choroid.
A rare condition associated with retinoblastoma is that of glaucoma, which Dr. Belkinn kept to relate a case in the Chobbinic Clinic of the Army Medical School at St. Petersburg. In this condition, it has been thought to be iatrogenic, for owing to the interstitial process in the retina and the increasing and obliteration of the vessels, there is a decreased blood supply to the globe and the filtration of fluids is diminished. Belkinn attributes the increased pressure in his case to a growth of connective tissue in the choroid, compressing the veins contained.

In other instances of this associated condition are recorded by Brandelstrom.

CAUSES:—In every case first pointed out by Dr. Seager, undoubtedly plays a great part in the production of this disease. In the case of Dr. Hahnen mentioned above, he distinctly states that on no occasion did the disease affect any member of a family whose both parents were free. Dr. Webster relates an interesting case of Dr. Brackham of Louisville, in which he concludes from the very complete family history, that retinoblastoma has been present in the family, handed down from father to son, for over 200 years. Out of 22 cases of his, he found 9 with a hereditary history, equivalent to 59.10 per cent. This shows that out of 66 cases, in only 1 was hereditary present, making a percentage of 1.5. While the Western & Randell report that direct transmissibility...
in essentially rare, and can only refer to the one case reported by Sebrell, and to another reported by Debeyre.

Constitution may be the parents or grandparents, has been held to be an important factor in the production of retinitis pigmentosa, but in regard to this there is great difference of opinion.

Debeyre, who first described this as a cause, stated that almost one half (from 40 to 50 per cent) of these patients were the offspring of union of consanguineous, while nine years later he says that this opinion has been confirmed by a constantly increasing number of cases.

In his first series of 53 cases, 44, or 41.8 per cent had a consanguineous history.

On Grafe's observed, out of 25 cases, 11, or 44 per cent, had a history of consanguinity. Of the two series (Debeyre's and Grafe's) five cases were identical, 15.5 taking the two we get out of 65 cases, 28, or 41.8 per cent, had this history.

Schröder states that in almost a third of the cases, 33 per cent, the disease occurs in individuals descended from consanguineous parents.

Debeyre and Breidholt get a history of consanguinity in 28 out of 103 cases, 27.3 per cent.

Sebrell in 12 out of 66 cases, or 17.9 per cent.

Reardon in 16 out of 60 cases, or 26.6 per cent.

Farrington in 9 out of 50 cases, or 18.0 per cent.

Debeyre in 8 out of 23 cases, or 34.7 per cent.

Igoe in 5 out of 25 cases, or 20.0 per cent.
We find 5 out of 22 cases, or 23.6 per cent. of reductio pigmentosa among the natives of India. It cannot be a cause in these cases. Froebeke considers that it is independent of consanguinity, though it may be present. He does not believe that the relationship of parents has anything to do with age lesions.

In the report of the Hôpital Marie Jeanne for 1861-2, 12 cases are recorded, in 11 of which there is any consanguinity traceable.

Oppenheim reports 8 cases, and Dardix 6, in which no consanguinity occurred. The preponderance of juvenescent cases proves that consanguinity is the parents is probably a cause, though not the only cause, of the disease. Activities pigmentosa and desquamation are closely connected and frequently associated together in the same patient. In a family in which both conditions prevail it is found that the children also suffer.
This suffer from the one complaint can also affected with the other. While those free from the one are free from the other. It is probable that they depend on similar structural changes and may be due to the same cause. In both constitution and heredity play an important part.

Lieberich found out of 241 deaf-mutes, 14 cases of retinitis pigmentosa, equivalent to 5.8 per cent.

Lieberich stated at this time that at least more than 20 other cases of retinitis pigmentosa were to be found among the entire population of Berlin. It is noteworthy that of these 14 cases, 9 occurred in Jews.

He examined 110 deaf-mutes with the opthalmoscope and found 5 cases of this disease, giving a percentage 4.5.

Bergmann found 5 cases in 200 deaf-mutes or 2.5 per cent.

Hence I am inclined to think there is some relation either between retinitis pigmentosa and certain mental conditions of the nervous system or as it frequently occurs in families in which there is a history of nervous disease. It is frequently associated with defective intelligence epilepsy, ataxy, etc.

Having in an examination of 31 retinias at Stettin, found 4 cases of this disease, or 12.9 per cent.

Lieberich, out of 50 idiots found 3 cases or 6 per cent.

Opinions, both congenital and acquired, have frequently been advanced as a cause. "Macnamara a Cambridge, Epler, specialized, and Douglas", attributing it to this. "Tong". "Blairfield", etc. and others have
reported cases. As a rule in these cases the disease is asymptomatic and there distinct differences from the true disease, as associated choroiditis, etc. Dr. Hutchinson says that any marked deviation from symmetry should make the suspicion of syphilis as a cause. James considers the connection between true retinitis pigmentosa and syphilis to be very doubtful, while Lister has never been able to connect the disease with syphilis either inherited or acquired.

Marked shock may also be a cause, as in the case recorded by Riebling, in which four children were subsequently to a severe fright suffered from deaf mutism and retinitis pigmentosa, whilst the two children born previously were quite healthy. Riggs records a case in which one eye was affected with the disease as the result of a blow, and the other eye became affected secondarily from sympathetic inflammation.

Schlesinger reports several cases in which he found the disease to be present in both lost eyes, it having developed secondarily to some injury.

Prolonged residence in a hot country has been ascribed as a cause by Berman, and also by Irwin, whilst Lewis calls attention to the connection between the disease and intermittent fever. Immuong states that out of 17 cases, 14 inhabited hot climate countries. For reasons for cases,

23
(infection and edema) which developed after an attack of
varicella.

The disease is frequently associated with certain congenital
anomalies, as supernumerary fingers or toes, of which
cases are recorded by Livingstone, Hector. The "posterior
acetabulum - Lawrence and Horsley," giving of cases
occurring in the same family, all of whom were of
enlarged stature, and of arrested sexual development;
lesser or posterior polar cataract - when this occurs,
vision becomes worse in a very bright light, for then
the opacity of the lens coincides with the newly
enlarged pupillary margin. In the case cited, the existence
of posterior polar cataract in both eyes should always
excite suspicion of the presence of retinoblastoma.

Or it may be associated with "necrolysis," deafness,
encephalitis, microphthalmos, persistent hyoid arches,
encephaloschisis, or aniridia, any of which may be present.

Such explains the occurrence of these anomalies
with retinoblastoma by attributing all to
the same cause, namely to the congenitality of
the parents.

Treatment:— The majority of writers agree that
treatment is of very little use, and that it is
prudent to resent the disease.

Cadaver states that pigmentary degeneration of the retina
is as a rule incurable, leading despite all efforts
in the way of treatment to a steady decrease in vision.
and to entire blindness.

Gehardt"" states that slight temporary improvement in central fixation has been observed in a few cases, but in no case was the improvement long maintained, and in no case and through no remedy has there resulted any enlargement of the contracted visual field. "Gehardt"" states that even temporary improvement is produced by no known remedy."" Whilst "Hubel"" is of opinion that treatment is useless except in syphilitic cases, temporary benefit has however been obtained by the use of the continuous current as recommended by Dr. Gurne."" The reports of cases, in all of which improvement took place, are explains the rationale of the treatment as follows: the defect of pigment prevents the rays of light from reaching the rods and cones, and consequently the nerve fibres in connection with these retinal end elements are thrown out of employment. The optic nerve becomes stretched from disease and it is this which the electrical current tends to prevent.

It also has a beneficial effect in dilating the retinal vessels, and thus improving the circulation of the tissues. Though Gurne admits that this has proved only temporary and not permanent in his cases.

""He recommends that the sponges be placed, one on the closed eyelid and the other on the supraorbital ridge on the same side, and that both be moved along the skin within their limits.

The sponges may occasionally be placed on both upper
gyrals with advantage. The advised commence with a weak current, say 0.5 to 0.7 volts, and cautiously increasing to 25, employing them daily for about two minutes.

Istert reports one case of improvement after galvanism, and 14 other cases. Benefit has been temporary.

Benefit frequently accrues from its use, as also have Bier of Philadelphia, and Deitz and Snyder, Hardie, of Boston. The latter records a case in which widening of the visual fields and improved central vision resulted from use of the current.

Fosterbridge, on the other hand reports no benefit whatever from its use, in some cases getting worse resulted.

Little reports one case of improvement under the use of the interrupted or bradic current.

Subcutaneous injections of thymol to stimulate the conjunctival retina has been recommended, and Chisholm has obtained good results from its use. One patient who could not see even a gas light at night, was able after a few injections to read a newspaper by the same light.

Riley of Philadelphia got marked improvement in the vision of light, and in extent of visual fields, by this method of treatment, but the improvement was not maintained.

Cincinnati reports a case where improvement in the visual fields was noticed three days after commencing injections of argyria (1/4 gr.), and after 10 days the fields were more than doubled.
central vision also improving greatly.

Grandolmest" is said to have obtained beneficial results from the hypodermic injection of Diphysine. He records three cases, in which he got retention of the visual fields with improved visual acuity and lesional sight blindness. Two of the cases he reports as cured, and one as improved.

Hypodermic treatment has many advocates though as cases of permanent improvement after its use are recorded.

Carmel states that he has the most confidence in hypodermic injections, whilst the Selden and Sandall

injections of the sublimate, they state that in three cases it does good but in the "typical achintia pigmentosa it is of no use whatever.

Hornet records a case where although visual power improved from 5/10 to 5/9 after the use of the sublimate the contraction of the visual field became one third greater.

Herseum in a hereditary" whilst advocating the administration of mercury admit that the disease does not seem to be influenced by it in the least.

Of the remedies, iodide of potash, the prolonged administration of iron, diaphoresis, hydro-

therapeutics, camphor, absorberts, abstraction of blood, and various derivatives have been recommended, with little or no benefit.

Blue glasses may be used to cut off the range
coloured rays, and to give the eye a certain amount of rest, and Hasley reports improvement after the continuous use of etetrine.

The prognosis is very bad. In spite of treatment the disease continues to progress, and complete blindness inevitably follows.

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References


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Notes on twenty-one cases of retinitis pigmentosa.
Cases I and II

Two cases of retinitis pigmentosa occurring in deaf mute.

Patients, brother and sister. History of consanguinity in parents, also history of maternal stroke. Father doubtfully affected. Hystagmus in one case.

I. Frank Russell, age 55, single, formerly a Rat Dealer.
II. Sarah Russell, age 48, single, formerly a Needlewoman.

Patients were both born deaf and dumb; they live with their received sister from whom the following history was obtained:

Frank never had good sight but at 21 it began to grow worse and he found he had difficulty in seeing in the dark. His sight gradually deteriorated, until at 30 he could not see to read and dare not trust himself out alone in the evenings. It was not until 8 years later that he had his eyes examined. He was taken to the Charing Cross Hospital where he was told that there was something the matter with the back of his eyes and that the condition was incurable. In the last year he has been totally blind, and for 2 or 3 years previously could only distinguish light from darkness.

Sarah had good sight up to the age of 30, when she began to suffer from right blindness. A few years later she attended St James's Eye Hospital but has lost her vision and does not know what was said to be the matter. Her sight has gradually got worse from that time. She can still see to get about in the daytime, and brings her brother home to the Middlesex Hospital by herself. She carries about with her a pocket blackboard and a lead pencil. She communicates with her one sister in
fairly large type, then the, by inclining the head until the light shines on the pencil marks, can read very much better than if the writer is in ink or white figured.

Family History: Father & Mother were 1st Cousins.

Father died at 82;of phthisis, he had very bad sight 40 years before his death could only guess his way about. He came to London to consult an oculist but it is not known that was the matter. Does not remember whether he suffered from sight blindness or not. Mother died of ascension, she always had good sight.

No history of grandparents, are 1 collateral branch.

Both parents of parents were born in a small village in Cambridgeshire in the England, their ancestors have always lived there as far as the bones, it is an agricultural district & the inhabitants have usually intermarried among themselves.

There were 8 brothers & sisters in the family of whom 3 are living:

1. Son, died of phthisis at 40, he was a doctor, he wore glasses but never complained of sight blindness.

2. Frank, born deaf & dumb.

3. Mr. Harris, married his 1st cousin, had children, all with good sight.

4. Son died in childhood.

5. Frank, born deaf & dumb.

6a & 6b. Died in infancy.

It is worthy of note that Mr. Harris has himself a son by a consanguineous marriage, has married his 1st cousin, a son of healthy children, all of whom were carefully examined. They are all fine, healthy, rather
precocious children. Of these, one, being hypochondriacal, gave her a sense of being unwell or unwell, the others being myopiacal, there were positive changes. The former were no congenital anomalies of any kind in the family.

The authors always attributed the condition of the two patients, Frank and Fred, to disorders which the experienced during their pregnancies.

In the case of Frank, during her second pregnancy, when the subjects of the children fell into an empty flower bin and being of a very serious excitable temperament, the occurred a loss of consciousness. Frank was thought to be an idiot up to 3 years of age, but when sent to school, he proved to be intelligent, quick at understanding and picking up things. As he grew older, he showed himself very acute in business matters.

In the case of Fred, during her second pregnancy, one of her children struck down by a horse, a cart, she was severely frightened. I always attributed the child's condition to the shock she sustained. Fred was a particularly intelligent child, getting on very quickly at school and easily learning everything she was taught.

Present Condition:

Frank, V. Rs. E. No perception of light.
Anterior chambers very shallow.
Posterior synechiae in both.
Anterior & posterior plan cubets.
Is tender, reflex.
Hydrophthalmia; slight lateral twitching of the eyes.
Deep - V. 

Cases III and IV

The cases of retinitis pigmentosa occurring in deaf-mutes.


Stereoscopic facilities in both.


Documental letter writer.

Patients are sister and brother, and both are congenital deaf-mutes.

They are of Jewish extraction and were educated at the Jewish School for the deaf a short time before the patient became blind. The following history was obtained from their mother:

Bessie was discovered to be deaf a dumb when 2 years old, as a child she had good sight and nothing was noticed amiss until she, during the time she was at school, that is up to 16 years of age. For after she came home from school she began to complain of not being to see well in the dark. Her mother thought nothing of it until one evening when patient went out of doors she was knocked down by a passing cart, she told her
under the sense of its coming. As time progressed, he noticed that if anything was held out to
patient. He failed to see it, he stumbled over potsheds
and chairs, and if anyone wished to take leave of him,
he failed to see the outstretched hand, all these
symptoms were much aggravated in a bad light.
Patient was never allowed to go out by himself in the
evning, after the accident. He was thought to
be simply short-sighted and was not taken to any
specialist until 6 months ago when she was taken to
Hawfields with her brother. Patient can see to
read small print easily, also to sew, thread needles
etc. The disease is not nearly so far advanced in her case
as in that of her brother.

Israel was thought to have good sight up to the age
of 17 when he was sent to school. He remained
at school until he was 16, and it was during this time
that his sight began to fail. His mother noticed
that he was continually knocking against people in the
passages, without seeing them; thinking he required
glasses he was taken to Hawfields, he could then
be about 12 years of age, the diagnosis Dr. Retin's
pigmentation was made at that time. The patient
returned home from school his sight was found to be
very bad, he apparently did not see anything unless
it was held directly in front of his eyes or unless his
eye-piece was particularly drawn to it. He could not
be trusted to go out alone; in a bad light his sight
is so very much worse that he tells his mother he could
see anything. He was again taken to Hornfield 6 months ago and put under treatment but was not obtained any benefit.

Family History: Father died at 70 of heart disease, he was an artist, he wore glasses but always had good sight. Mother being aged 47, hypermetropic -1.5 D, needs glasses for reading. Father a brother were not related before marriage. Father was a Russian Pole and Mother a native of Friesia. No consanguinity in the grandparents. No other members of the family have suffered from this disease, mother cannot account for it in any way. There is no history of retinal disease, in fact the mother was particularly well and in prosperous circumstances during the time she was pregnant with these patients.

Three were 8 brothers and sisters in the family, of these 5 are living:
1. Son, my brother, wears glasses, no sight blindness.
2. Brother, one deaf a mute.
3. In good sight.
4. Daughter, good sight.
5. Died, one deaf a mute.
6. 4x. 3x. 2x. 1x. 1x. and daughters, all in good sight.
7. 3 children, died in infancy.

The mother has brothers, a sister living with large families but none were born deaf a mute, at time complaint of this condition of the eyewight.

There are no signs of congenital syphilis in either parent, they are both of good physique, no itching? hectic or jaundice eminences. No history of acquired syphilis to be obtained from the patient.
Present Condition:

Posterior v. 1/2 Raka. 5-5.1. 31. There is posterior pole of left lens. Cause is unclear. Speculum is fully seen with glass in place. Date of surgery is unclear. Beautifully defined pigmentation in the form of retinal Laminae with branching processes in some places, charging to the vessel. Retinal changes to be observed.

V. 1/2 Raka 1/5. Conclusion insufficient.

Cilioretinal vessels sluggish to light. Cause is unclear. Speculum is fully seen with glass in place. Vessels are seen to their limits.

Arterial vessels are distinct. Extensive pigmentation of retina is present. A thickest chiefly projected around the vessels.

V. 1/2 Raka 5-5.1. Patients being kept under constant medical supervision.
Parasitic pigmento affecting 6 out of a family of 10, all males or 6 cases. Associated severe oedemae and deafness. In one case, no consanguinity, no heredity, and no specific disease. Symptoms commencing late in 1 case, probably congenital in 2. Whereo specifics in one case.

It will be more convenient in this instance to give the family history before going into each case individually.

Family History: All are born in Anerhau, Buckinghamshire. He died at 56 of diabetes. Father was born in Buckinghamshire. He died at 69 of cancer. They were no relatives whatever before marriage. He had good sight all their lives.

Father suffered from his nerves a good deal. Grandparents were not related. No history of collateral branches.

There were 10 children in the family. A list the following is a list in order of birth:

1. Elizabeth, a patient, see case V.
2. John James, died at 63, complained of night blindness for 30 years or more. Was a patient at Hopefield for many years. He became insane and died in Longthatch Asylum. Fully blind. He was married and left a family of five daughters, all with good sight.
3. William, a patient, see case 77.
4. George, age 63, a rate collector in Crockham. He has good sight. Wears glasses for reading, does
not complain of night blindness.

6. Emma, died at 38. She has no trace of it. Poor and

7. James, died at 57. I good sight. Married, and left

8. Thomas, died at 32. He never slept, but did not

9. Henry, died at 20. He never slept, but did not

10. Henry, a patient, see case VIII.

Here are no signs of inherited syphilis in any of the patients, and there is no history of acquired

All the patients were visited at their

ever carefully examined with the stethoscope, none were found to be suffering from this disease.

Case V. Elizabeth Wilson, age 18. Single. Formerly roasted. This patient was seen at her home in Amsterdam, there she lives with her brother Henry. She is also a patient (see case VIII). There was much difficulty in obtaining a clear history as she suffers from deafness, and her memory is somewhat impaired. The following was elicited with the help of her brothers and
She has lived to 82 years of age, the cause complained of her sight, then gradually increasing right blindness developed. She could see to follow her employment as a dressmaker, and could thread a needle, age 65. With her sight began to fail rapidly, and at 67 she could not see to read even large print. It was at this time that she came to live with her daughter. When her sight failed altogether the corneas were, perhaps, 10 years, or may be longer. It has been ailing for many years, suffers from rhematism, and walks with difficulty. With the aid of a stick, she finds her way about the house and knows the rooms by their voices, so that still though she was able to see a little, the result is that she always had good sight as a girl and as a young woman. The cornea never been under treatment. It has never had her eyes examined before in her life.

Present condition: Patient is totally blind, she has no perception of light in either eye, the corneas feel light from darkness. There is anterior polar cataract in both eyes, also posterior cortical cataract, get red reflex at periphery. If dilated pupil, one can see the fundus individually. Can see much black pigment but cannot make out its distribution. Cannot see disc, and cannot make out any retinal details. There is no cataracts.

His patient was visited at his own home in Hexham. He states that he never had good sight, even as a child he used to complain of not being able to see well after dark, and it was noticed that he would run into furniture without seeing them. When 19 or 19 years of age the sight blindness was so marked that he did not go out alone after dark.

At 40 he found himself unable to read, and at this time he had his eyes examined by Dr. Bower at Morpeth, who informed him that nothing could be done to improve his sight. He has not ceased to attend. He has still the ability to follow his employment but was obliged to give up all fine work, his sight gradually became worse until at 50 he could not see to do anything at all.

Patient is a tall frame man with well marked features. He has a considerable degree of deafness. His wife says she is much for Terrorism, but he is very easily excited, and that if he is worried or ill he gets very nervous. At times he is peculiar in his conversation and actions, making her rather anxious about him. He has had 7 children, 1 boy, died aged respectively 12, 20 (five children), 52 (from cancer), and 65 (from phthisis). All his children have good sight and none complain of sight blindness. One daughter, aged 50, was visiting him at the time these states were taken, she is quite unaffected and free from the disease.
Recent trouble - in R. & L. S,ingers at 7 a.m.
Visual fields - Very much contracted. Asked to look at a certain spot. Patient moves his eyes about in all directions in his endeavour to find it. When he does see it, then moving fingers require to be brought to within 2 inches of the spot before he sees them, noting the vertical and horizontal limits of his fields not more than 4 inches. His patient has been in his own house, so fixation charts could be made.

Optikulometrific Appearance - Anterior polar cataract is little, some cortical opacity or posterior pole.
BH lines are absorbed, and the vessels represented by some strands close in the periphery. They cannot be traced at all. There is a plentiful deposit of black pigment in the periphery of the fundus, and extending almost up to disc, especially on inner side. Near it extends quite up to edge of disc. At whole fundus is pale and there is a great chasma about it.

Cannot detect any retinex facies, and there is no nystagmus.
Colour sense very imperfect, all colours appear dark to him, and he distinguishes one from another only by a difference of shade.

Case VII. Harriet Hitcham, age 52. Paralysed, formerly Dressmaker.
His patient was seen at Tweepeals, and also at his home.
"Centre each chart with 'pointer' at Zero before commencing to use the Automatic Registration."

**LEFT.**

**RIGHT.**

PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal Field of Indirect Vision; the small red circle the position of the blind spot.

The patient stated that she always had good sight until 10 or 15 years ago when she was about 35 years of age. She then began to notice that she could not see well at night. At first, she looked as usual, but over the years she complained of anything before this. The patient has been able to trust herself out alone after dark for the last 2 years. Up to 6 months ago she was able to read ordinary newspaper type in a good light. But since then her sight has not rapidly become, and it was on this account that the letter of Fairfields at Horsham was made.

Patient was married at 20; she has had 2 children and 2 miscarriages. Her first child died at 9 months. Her second is a girl aged 31. She was examined at home, she has good sight, does not complain of night blindness, and shows no further change. Patient afterwards had two miscarriages. Her history ofophthalmia to be obtained. To these charts we read, to the best of our recollection, patient's daughter is healthy in appearance with no signs of inherited disease. Patient has only indifferent health, suffers much from nervousness and from dyspepsia. She has a slight degree of dyspepsia.

Present Condition: Vision R. E 58/20 after Tg. 8 + 0.5 D. Tt. Refraction: Hypermetropic +1.0 D. R. E.

Visual Fields: Constricted concentrically. See charts opposite.


Ophthalmoscopic appearances: coarse floating specks in vitreous, especially in left. Anterior vitreous turbid in both PLR green-white. vessels narrowed to thin
lines. Pigment is not abundant, it is scattered over the periphery in form of dots and patches more in relation to vessels but more lying between them, the spots are chiefly isolated and do not appear to coalesce with one another to any great extent. The pigment is seen more on the nasal side of left side, and also down and inward.

Case VIII. Harry Whiton, age 49, married, formerly Servant to Jane. Patient was seen at his own house in London, he is the youngest brother of the 3 preceding patients. He states that he had perfectly good sight up to 20 years of age, it was then between 25 and 30 that he first noticed anything the matter. He was in the habit of playing cricket in the summer evenings and he found that as soon as it commenced to grow dark he had great difficulty in seeing, he often had to give up playing half an hour or more before his companions. Soon after this he was out driving late one afternoon, when he ran into a tree which was driven up in the road, he said he could not see it. Those who saw the accident laughed at him and said it was plain enough, it was getting dark at the time. It was a week after this accident that he decided to have his eyes examined, he attended Moorfields, he was under treatment for a considerable time and took a medicine which eventually made his feet loose, it was then stopped. Patient got so better he ceased attending.
the right blindness got more checked until 3 or 4 years ago, he found that he could not trust himself out alone after dusk. Up to 2 years ago he could read a newspaper with ease in a good light, but up to 6 months ago could make out large print. Since then because he says he has not been able to read anything. He can find his way about in the daytime and can recognize people he knows. The says he sees best on a clear day with an overcast sky, he cannot see at all in bright sunlight. He was able to follow his employment up to 2 years ago then he was obliged to give it up.

Patient is a strong healthy looking man, he suffers from a slight degree of deafness, but is otherwise in good health. He says he has never had any illness of any kind, he has lived in the country all his life. He has 9 children living all have good light, was able to examine 3 of them with the ophthalmoscope and found the fundus quite normal in each.

Patient's wife stated that he is very nervous about his sight and broods over it a great deal.

Present conditions: — Refraction Orthostatic 0.15D. L12D Visual fields normal, estimated roughly by having patient look at a fixed point and approaching with moving figures, the limits here as follows: — R. Temporally 5 inches, and vertically 6 inches.

L. Temporally 7 inches, and vertically 6 inches. In various directions as before specified.
The right blindness got worse marked until 2 years ago. He found that he could not trust himself out alone after dark. Up to 2 years ago he could read a newspaper with ease in good light and up to 6 months ago could make out large points. Since then because he says he has not been able to read anything. He if can find his way about in the daytime and can recognise people he knows. He says he sees best on a clear day but at night he says he cannot see well in bright twilight. He was able to follow his employment up to 2 years ago when he was obliged to give it up.

This is a strong healthy looking man, he suffers from a slight degree of deafness but is otherwise in good health. He says he has never had any illness of any kind, he has lived in the country all his life. He has 9 children living all born good sight. Was able to examine 8 of them with the tulip lunette and found the fundus quite normal in each.

Patient is quite satisfied that he is very conscious about his sight and broods over it a great deal.

Present condition: Refractive Hypermetropia 1 1/2D, 2 1/2D Visual fields were contracted, estimated roughly by making patient look at a fixed point and approaching with moving fingers, the limits were as follows:--

R. Bregial 1 horizontal 7 inches
1 vertical 7 inches, and vertically 6 inches.
L. Bregial 6 inches, and vertically 6 inches.
In colour sensations and 24 hypermetropes.
Pathological appearance: - Cornea clear. Both discs are of a greyish color and the vessels are diminished in size. There is a deposit of pigment arranged in a lace-like manner in the periphery of both eyes, fairly equally distributed in all directions. In some places it can be seen clinging to the retinal sheaths. In the right eye there is a small patch of pigment to the inner side of the disc, and another immediately below the disc. There are some small, hazy-looking spots of varying size, scattered irregularly in the macula region of both eyes. More marked in the left.

The patient was able, when in his own house, could test his vision with test figures, and could read ordinary charts in his visual fields. Subject could count figures at a distance of 5 meters, - the length of the room. As regards color sense, he could distinguish red from green, and green from yellow, could not apply precise tests.
Case 28.

Retinitis pigmentosa in a patient of dwarfed stature. 
No history of congenital syphilis or of syphilitic disease.

Genitalia normal, Rectum also affected.

Nystagmus. Vitreous Opacities.


Patient states that he always had poor sight, he was formerly a boat-maker but had to give up this work when 25 years of age, owing to increasing difficulty in doing fine work, threading needles, etc. He did not have his sight examined, but changed his occupation and became a labourer. So and a half years ago, he began to notice that his sight was much worse in the evening. In a dark light he found he could discern, see at all, and it was on this account that he went to Moorfields to be tried for glasses.

Retinitis pigmentosa was diagnosed at that time.

Patient is dwarfed in stature. Height 4ft 7in.

His ears are very small, when standing up the tips of his fingers only reach to inches below his great toes.

Family History: Father aged 68, good sight, can see to read without glasses, highly astigmatic.

Mother 66, has very bad sight but does not complain of night blindness, she absolutely refuses to have her eyes examined, but is willing to remain to be seen externally. Sister a suitor named not related before marriage. Patient was the 3rd of 8 children of whom 3 are living, none of his brothers or sisters
complain of their sight. As his sisters' children cannot
see well at night (see case 5).
Patient was born in Bermuda. His parents and
grandparents have always lived in London. Does not
know what his grandparents died of. Never heard that
they complained of their sight. Does not think they were
related before marriage.
No signs of inherited aphakia in patient and no history
of acquired aphakia.

Present condition: - Vision R.E. 2/5, Love's fingers at 4m.
Reads letters of T.9. Reproduction 1/3 R.E.
Visual fields normal. Excluded the charts opposite.
Colour sense - Patient is completely colour blind. Given a
pale green, he selects grey. Brown, pink or orange.
Given a light purple, he selects greens, brown, grey or blue.
Iridoscopy: In left eye no exudate present.
Cornea vitreous:
Patient easily seen with plane mirror.
Uveal vessel appearance - Vessels greyish in colour.
Nests small and staphylini. In the right eye there is
some central choroiditis; in the left there is a large
patch of choroidal atrophy on the temporal side of disc.
The pigmentation of the retina is in this case very
typical of the disease. Numerous spots with branching
processes joining one another being seen in the
periphery of the fundus.
Case X.

Retinitis pigmentosa, probably congenital, occurring in a child aged 9, nephew of previous patient. No congruinity, and no cause.


This child was examined at his mother's house. He is a robust healthy looking lad, and shows no signs of congenital syphilis or frontal enuresis, as noticed by Bell & Co. Mother states that when the child was about 3 years of age, she noticed that he did not see objects when held out to him unless his attention was directly called to them, also he would fall over things apparently not seeing them.

As he grew older this became much more marked, especially in the evenings and in a bad light, she has never taken him to have his sight examined. He goes to school and knows his letters, but his mother says he is very slow at learning anything.

Family history: He is aged 9. Mother 54. Both have good sight. They were not related before marriage. Grandparents on mother's side both living, see previous case. No history to be obtained.

Grandparents on father's side.

Mother is the eldest of 7 living children, 4 died in infancy previous to his birth. The 2 youngest children are twins and are only 2 months old. All the three children appear to have good sight, and mother has not noticed anything wrong with them.
The mother and # of the children were examined with the ophthalmoscope and nothing abnormal detected. Present condition: examination necessitated any improvement as was unable to induce the mother to bring the child to hospital and she would not allow the pupils to be dilated.

Refraction: Hypermetropia about +2.0.

With the ophthalmoscope fine crescents could be seen on the outer side of both discs, the retinal vessels were small and the discs were considerably pale. The choroidal vessels were very distinct. A few small dots and streaks of pigment were seen in the peripheral of the fovea in both eyes.
Case 81.

Anomalous form of retinitis pigmentosa occurring without pigment. Similar to those cases of retinitis punctata albinosa with associated night blindness and contracted visual fields. As consequence, no cause: subjective luminous sensations; late manifestation of symptoms. The vitreous crystals.

Carroll, Andrew, 61 years. Married. Barrington.

Eleven or fourteen years ago, that is about 5 years before her marriage, patient began to notice that she could not see to well in the evenings, she was then 50 years of age. At this time she particularly noticed that the street lamps at night looked like little specks of light, this she thinks was the earliest symptom. As time progressed, she found herself getting worse in this respect, if she went out at night she often ran into people without seeing them, and she could not distinguish objects clearly. She did not suffer much discomfort until 10 years ago when she began to complain of seeing black clouds. Whereupon she looked at any object in a bad light, this symptom has persisted up to the present time. The clouds she describes as being about the size of a breakfast plate, and resembling with her eyes, the only articles it is the streets; occasionally she complains of seeing gold specks in both eyes, especially the left. In winter she always sees better than in summer. She has noticed this early years since the affection commenced. For the last 6 years
She has not been able to trust herself out alone at night.

Family History: Father died at 71, he wore glasses but had good sight. He was able to see up to the day of his death. They were not related before marriage. Father was a native of Hampshire.

Mother was born in Plymouth. Knows very little about her grandparents.

Felt his right eye for the last 10 children of whom 6 are living. Died of pneumonia. One of them, aged 6, died in 1935. One other living.

Patient has been living in Beaufort, Yorkshire. Dr. Halsey of the Beaufort Hospital kindly examined him for me and found him to be suffering from tuberculosis and very much from this disease.

Patient has never heard of any member of his family suffering from anything similar to his complaint.

He is married and has 2 children, one of whom is of unsatisfactory age. The history of his disease is as follows:

Recent observation: R. V. = 4/6 in Snellen test. 2/6 in test for near. L. V. = 1/6 in Snellen test. 2/6 in test for near. Teller's E = +2.25 D. red. 5/3 at 26 cm.

With Bignami's types in dark room V = 4/6, R. Z. is the disease.

Visual fields contracted symmetrically on the chart. Clear sense of a little infradescence, contrast of grey to blue with light green.

Can match juplets in dark.

Fine retinal specacies in left. No astigmatism.

Dilated pupils. No inflammatory reaction.
Case XIII

Isolated case of retinitis pigmentosa. The consanguinity is known. History of insanity and tubercle on father's side.

Alfred King, 18. Gottholden.

At 13 years of age, when patient was an inmate of Dr. Geyger's orphanage, it was noticed that he could not see to read nor to do any work in a dim light. He was taken to Dr. Kohnstamm on account of this but does not know what was said to be the matter.

His symptom gradually increased, and in 2 years time became so troublesome that he was afraid to trust himself out alone in the evenings. He then went to Horsham where he has been under treatment since.

Family History:—Father died at 52 of phthisis. He always had good sight. Father being aged 50 received complaints of his sight. They were not related before marriage. No consanguinity in grandparents.

Father's uncle died in a lunatic asylum, and one of his cousins was always "peculiar."

Patient is the 4th of 8 children. I show 6 have died.

1 in infancy (two twins), and aged 10 from spinal meningitis. 3 others living all have good sight.

-28-
"Centre each chart with 'pointer' at Zero before commencing to use the Automatic Registration."

**LEFT.**

**RIGHT.**

**PERIMETER CHARTS.**

The eccentric continuous red line indicates the average normal Field of Indirect Vision, the small red circle the position of the blind spot.

Patient was born in London, as were his parents and grandparents before him.
Patient's health was doubtfully affected, but he has no deafness and no frontal excrescences, his features are good & well formed. His younger brother also has also been in quite free from the disease.

Recent Evidence:
- V. = 5/6, S. = 10 D, 15 D, 1st inst.
- 51 at 10 cm, 2nd inst.

Visual fields are partly contracted as seen charts.
Claw toes are insignificant.

In Eyegrounds, no vitreous changes.

Fundus Oculi Appearance:
- Ophthloid vessels, retinal vessels, vessels, slightly contracted. Pigment is found in small tiny dots scattered near the periphery of both retina, not very abundant. Both retinal regions have a peculiar brownish dotted appearance. Chorioid vessels are very apparent.

Patient has been taking quinine a long for the last 3 years, the lashes current was ordered but he could not intend regularly to have it applied, and could not afford to buy it himself. Five years ago his vision was R. 6/12, L. 6/10, but it has gradually deteriorated.
Case XIII

Case occurring in a patient of weak intelligence.

History of maternal shock. In consequence:

Venous Glaucia.


Nothing was noticed amiss with patient's sight until
the age of 13, when his mother noticed that he
knocked against objects in the dark, and had
difficulty in seeing. As he grew older, this condition
became more and more evident. For the last
12 months patient has not been allowed to go out by
himself in the evenings.

In appearance, patient is strong and healthy; he looks
like a typical farm labourer. His mother states
that he has always been deficient in intelligence.
She knows his words, and even she is not always
able to understand what he says.

His mother attributes his condition to a severe fright
she had during her pregnancy, owing to a large dog
attacking her.

Family History — Father and mother both living and
enjoy good health. They were not related before marriage.
Grandparents also had good sight as history of
consanguinity. Patient is the eldest of 12 children
of whom 10 are living; none of his brothers or sisters
complain of their sight. Patient was born at
Whitaker Alley in Evesham, as were both his parents
and grandparents before him. There is no history
Of any similar condition occurring in the family before. 

The signs of congenital syphilis i patient. in his 
brothers or aisters. 

Recent Condition: Patient can read letters but that is all. 


Refraction: Hypermetropia +0.75D to +1.0D R.E. 

Visual Fields: and Colour sense it was impossible to 
take owing to patient's lack of intelligence. 

The equotropia. The vitreous gazhes in both. 

With the fundoscopy there is a network of pigment 
to be seen in the periphery of the fuduses in both eyes 
in some cases it can be seen adhering to the 
vascular sheaths. The disc are pale and the 
retinal vessels are narrowed somewhat.
Case XIV


John Powell, 28. Single. Formerly domestic servant. Patient lives with his mother, he has suffered from epileptic fits since 18 years of age, he was obliged to give up his work on this account and has done nothing since. He is now attending the Hospital for Paralysis and Epilepsy, Queens Square. He never enjoyed good health as a lad. He is I tall stature and from physique, he has dark, thin hair and light blue eyes. Patient was always short-sighted but did not complain of anything else until he attained the age of 18. Then he found he had difficulty in reading, letters used to run together, and his eyes to ache. He attributed this to the fits which he began to suffer from, and has always connected the two conditions together. The fits and the failure of sight, as cause and effect.

18 months ago he began to complain of night-blindness which soon became very pronounced, and it was on this account that he went to Westhampnett to have his sight examined. For the last 12 months he has not been able to go out alone after dark, his sight has rapidly got worse during this time up to 12 months ago he could see to read a newspaper but now he can only make out exceptionally large print. Patient was born in Ireland but
"Centre each chart with "pointer" at Zero before commencing to use the Automatic Registration.

**LEFT.**

PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal field of Indirect Vision, the small red circle the position of the blind spot.

Designed for use with Prof. M. Hardy's Registering Perimeter. Published by Messrs. Curry & Paxton, 195, Gt. Portland St., London, W.
has lived in London since 5 years of age. She has never suffered from deafness.

**Family History:**

- Theodore was the 2nd of 3 children.
- His mother states that he was born early without instruments, she never had any trouble or fright during her pregnancy. She has one older brother aged 50, who is 65 years of age and has very good sight.
- One younger sister died in childhood of measles.
- Theodore's father died of typhoid fever before he was 2 months old.
- Before his death, he complained of difficulty in seeing after dark, his sight was never examined.
- Theodore's cousin (his sister), died in a lunatic asylum.
- His history of grandparents on father's side.
- His father, born in good health and good sight.
- Grandfather on father's side died at 80 of old age, grandfather at 85. The does not know what.
- The great grandfather died of consumption.
- Father and mother were 20 related before marriage, they were both natives of the same place, Down, County Down, and their ancestors for generations back have been always lived there. The result shows no signs of inherited aphthous, but was fairly good, well-formed, not darkened, no cavities about mouth, no typical cataract, no...

**Present Condition:**

- R: V. to 2 - +10 D + - . Tib 2 H in.
- L: V. to 2 - +10 D + - . Tib 2 H in. While reading, read slightly to light and accommodation. Eyes fairly active.
- Visual fields fairly full, contracted - little or temporal side.
- Color sense intact and unimpaired.
- Right lateral nystagmus and extreme movements of the eye. No visible fixations.
Ophthalmoscopic appearances: Discs greyish white and angular. Blood vessels much narrowed. Pupils were greyish light all over from retention of the aqueous humour. Vessels has a heaped appearance from the elastic wearing through the retinal border. Very little pigment to be seen in retina, but indistinct greyish area at all, but no dilatation into roundness, three small patches can be seen, adhering to the upper part of a vessel, no extreme jumbling of the right eye; no partial lighting well upwards. The left eye a few fine dots can be seen in the upper and lower jumbling, there are no tellable defects. Sclera are corneal white in the jumbling jule of both lenses.

Case XV.

PATIENT: Pigmentation commencing at 29. Present a fovea.
No constellation. Descent on neither side.


Patient states that she never had good sight even in a child, but it was not until 14 years ago that she began to notice that her sight was much worse then the light was bad. This symptom caused her a good deal of trouble and annoyance and shortly progressed until she found herself unable to go out alone after dark.
This was 4 years ago, 2 years after she was knocked down by a horse & cart, resting in the shade to let being able to see it coming. She felt weak from that to horse until she became unable to see anything at all. For the last 3 years she has been totally blind.

Family History: Father was a tailor, he died at 50. Mother died young. She had good sight as far as she knew, they were not related before marriage. Grandmother on mother's side died insane. Her cousin in grandparents' history.

Father had one brother, and one step-brother, both with good sight. He has not heard of any close family ever suffering from this complaint.

Patient has been married twice, she has two children living, one by her first husband, and the other by her second, a girl aged 29. She is in service away from home. She has good sight, and a boy aged 16. She was examined carefully. He has vision 20/70 & 20/20. Generous healthy no findings, deposits and no contraction of the visual field.

Patient lost a child in infancy, and the two lost at miscarriage. Patient was born in Brandon, Holland. Her parents and grandparents were also natives of that place.

In history of acquired ophthalmia and no signs of inherited. She of miscarriages are suspicious but patient is herself healthy, and has for others no signs of the disease.

Present Complaint: Patient cannot count fingers with either eye, she has no perception of light. Refraction examination was 5/200. Unable to delineate visual fields or color other.

The right eye dilates 2%. Tight lateral straining of the eyeball. A few old blue patches in the whites, seen...
test with a plane mirror, and a +12.0 D spherical
ophthalmoscopic appearance: grey opacity of vitreous.
acuteness small, vision not markedly so. India ink pigment
of stellar character with branching processes. Extending up
to rim of disc on nasal side. Having a particular
relation to the vessels in its deflection. Protrusion of disc
visible in both lenses. It is addition a triangular opacity
in the substance of the left lens with the base to the infer-
site.

Case XVI
Case occurring in girl aged 22. No consanguinity.
No cause of trauma to be ascertained. Right blindness a very

His patient came to Moorfields Hospital on account of a
diagnostic Retinitis of the right eye which he had noticed for
5 years. Retinitis pigmentosa was discovered on ophthalmological
examination.

Isabella Gilchrist, age 22. Height: 4’8” Domestic Servant.
The eyeguard patient states that she always sees worse in the
dusk, and has done so once since she can remember.
In walking in the evenings she requires to exercise great
care as she sometimes runs into objects. She has often
stumbled against chairs and tables in the house at night
before the lamps are lit. Patient is short in stature
and strong, and well built. She shows no signs of inherited
ophthalmitis. She has perfect health, has not lost one and
Centre each chart with "pointer" at Zero before commencing to use the Automatic Registration.

LEFT.

RIGHT.

PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal Field of Indirect Vision; the small red circle the position of the blind spot.

Designed for use with Prof. M'C. Hardy's Registering Perimeter.

Published by Messrs. Curry & Paxton, 195, Gt. Portland St., London, W.
Here is not a single one disabled.

Family History: - Both living, age 56. Both clear.

Mother, clear, suffers from cataract. Uses glasses for reading but has good sight.

Mother, living, age 49, does house work, she is nearly blind from glaucoma, can only distinguish light from darkness, she has undergone 7 operations for this.

Father was born in Newcastle, father comes from Yorkshire.

They were not related before marriage. Grandparents had fair sight, no consanguinity.

Mother is the 2nd of 9 children, two of her brothers have disabled horsemen, and wear glasses, but none complain of sightlessness. Her sister suffered from pole when a child, and also had glaucoma. She has 3 cousins all still good sight, but has lost all her other relations.

Present Condition: - Vision Right to 20/200. Left 20/70. 

Refraction: Hypermetropia Astigmatism R +2.00 sph +0.50 cyl 180 
Left +2.00 sph +0.50 cyl 180

Visual fields extended slightly in temporal sides.

Color sense, impaired, responds blue white green, hesitates a little on yellow, and is difficult to finish.

Ophthalmoscopic appearance: Dyes are a little fainter.

Cornea particularly clear, perhaps a little duller than normal. Pigment is small in amount, not chiefly in periphery. None it appears like a fine delicate lace work. In addition there are a few capillaries like blood vessels along the retinal vessels.
Case XVII


Frederick Drayher, St. George, formerly barrack-sergeant. His earliest symptom patient can remember is not being able to see objects when he looked at them directly. He noticed this particularly when playing at marbles, he could not see to pick them up if he looked straight at them, but required to look to one side when he saw them distinctly. His mother states that when 11 years of age he had a severe illness, a "cholera", and that he lost sight completely for three weeks, he recovered but his sight has gradually got worse from that time.

Right blindness has always been present as long as he can remember, he can get about at night but has to exercise extreme care. Ten years ago he went to Bright-seat to see if glasses would improve him, he was told that his condition was incurable and advised to learn some occupation for the blind. Three years ago he was obliged to give up his occupation as he could not see to do his work in the dim light of the warehouse.

He has not been able to read a newspaper for the last 3 years.

Family history: — Father died of heart disease. He always had good sight. Neither living free from the disease.

Grandparents on mother's side both lived to be over 70, and both had fair sight. No history of grandparents or father's side. No consanguinity in parents or grandparents.
"Centre each chart with 'pointer' at Zero before commencing to use the Automatic Registration.

LEFT.

PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal Field of Indirect Vision, the small red circle the position of the blind spot. Designed for use with Prof. Mr. Hardy's Registering Primeter.

Published by Messrs. Curry & Paxton, 195, Gt. Portland St., London, W.
Patient is the 3rd of 9 children, all living, and all in good health. One of them has been much gassed. None of his family as far as he knows, have ever suffered from this complaint. Patient lives with a married sister and her three children. Have been all examined with the ophthalmoscope and nothing abnormal discovered. Patient was born in Hertford. His father came from Suffolk and his mother was a Londoner.

His signs of inherited affections in patient, he is a very intelligent man, and owns some very good features. There is no congenital abnormality of any kind in the family and nothing to account for his condition.


Refraction: Esophoria + 3.0 D Plue.

Visual fields show central scotoma, six chart fingers.

Color sense much impaired, hesitates a good deal.

Give a pale green he chooses greens, light blue as well as green.

Give a deep jade he selects brown, greens and purples.

Give a dark red he selects dark brown as well.

Eight lateral cycloplegics. Tried especially to patient looking upward to extreme.ckenhaha. To be made out.

Gall-bladder fluoresce appearance. Both discs are partially shadowed and the vessels are small. Retinal pigment is seen running along the vein sheath like lichen in the branches of a tree. There is not a great deal of pigment, and it is chiefly seen along the veins as above. Jimen in nothing in the distribution. The pigment to account for the central scotoma is the fields.

Two is a brownish patch at each yellow spot which simulates
an old hemorrhage, and around the vessels there are numerous little blue dots scattered over the retina.
The retina is thinned and shows a grey tinge everywhere. The choroidal pigment is very evident. There is no choroidal vessel exposure anywhere.

Case XVIII

Case of retinitis pigmentosa occurring in one only in a family. Its consanguinity, closely on father's side. Associated with acquired myopia. Symptoms commencing late in life, at 49.

George Poole, 52. Dulledness. Hocks laboured.

Patient never complained of his sight until 5 years ago, when he began to be troubled with night blindness. He found difficulty in seeing objects after dusk. He noticed this especially in the house when he was working, owing to the lack of light, and it was on this account that he came to Harropfield 18 months ago. He can still find his way about at night but with difficulty. Patient is a short, thick-set, robust looking man, he has red hair and grey moustache. 28 years ago he had an attack of syphilis and was under treatment 6 months. He has been a heavy drinker, and a hard drinker all his life. He drinks 1 oz strong daily but used to drink 2 oz.

Family history: Father and mother both deaf; they were both born in London, father in West Ham, mother in Whitechapel. They were not related before marriage. They had fair sight as far as
he never, and never complained of night-blindness. Does not know anything about his grandparents, except that his father’s mother died insane in St. Bonaventure, where she was an inmate for 9 or 10 months. Child is the youngest, and the survivor of a family of 5. Two sisters died in childhood, one brother at 45 of consumption, and another brother at 42 of blood poisoning, his brothers had good sight up to time of their death.

Child is a widower with one child, a girl; 16 years old she ran away, and he has not seen her since. He had good sight up to that time. His wife died at 48. From change of life, he had no other children, and no miscarriages.

Present Condition: — V. Rail & the 0.20’s 25 c. E. +2.52 51 at 25 cm.

Tension: +1.02 D & 5.

Visual fields: contracted a little on right side, see chart opposite.

Colour sense: Slight chromatic sense, shows some confusion. Littke green confusion Dragon, but matches perfectly a dark red.

Droops Specchio: There are a large number of fine dots, a hanker seen floating in the Retinae with a + 1.2 lens, some have highly refrangible power, like cholesterol crystals, there are especially numerous in the left eye, where they can be seen by focal illumination above.

Pathological appearances: No lesion of lens in either eye. Discs involvement with peculiar glossy appearance, like pale yellow satin.

Iris, especially marked on outer side of each eye, where it forms an irregular crescent, particularly in the right eye.

Periost arranged neatly all around the periphery of pupilia, where it forms a beautiful black lace-work like arrangement, in some places its seen in threads like grains clinging to the supra of its edge.

The vessels are not much altered except in periphery, where they are narrowed to fine threads, here also the retina appears thickened, & the choroid appears an old red.
Case XIX

Case of retinitis pigmentosa, with associated specific disease.

Earliest symptom at 30. "Be convenience.


Rebecca Irish, aged 49. Married. originally Descartes.

This case was seen at the German Hospital, Dalston, and the following history obtained:

Patient was always short sighted, but never noticed anything else serious until she attained the age of 30, when she found that she had difficulty in seeing in a dark light. This symptom gradually increased, and in addition she found that she could not see objects outside the direct line of vision, unless the object was directly in front of her. She has been unable to trust herself out alone for the last 10 years. She has been to many different Hospitals, but nothing has done her any good. Her sight has got worse and worse, she cannot see to read, and cannot find her way about even in the daytime.

Family History:— Father died at 57. Mother at 90, does not know what they died of, both had fair sight up to day of their death. Father was a native of Ireland. Mother was born in Ireland. Neither her parents nor grandparents were related before marriage.

Patient is the 2nd of 8 children. 3 have died, none complained of their sight. One sister suffered from them since a boy. Patient has married at 22, she has only 2 living children, out of 9 confinements, which were as follows:

1. A miscarriage.
PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal field of vision, while the smooth red curves the position of the blind spot.

Left.

Courses each chart with 'posterior' at zero before commencing to use the Automatic Registration.

Published by New 'E' Chappell & Friends, West End, London, W.

Designed for use with Prof. McIlhenny's Registering Perimeter.
2. Boy, living, age 21. Frighten stroke that these two were quite.
4. Seven months old, died in infancy.
5. Girl, died aged 8. Incontinent, had dyspnea at birth.

Miscarriage.

Patient has suffered from frequent accidents of some kind, sometimes losing her voice entirely. The disease came having a rash or losing her hair. The above pains in front of her legs at times. Her husband has left her, he was a story healthy man as far as she knew.

Present Condition:

- R. V < 8/20, T +4.0 D,ylinder of 9. Fig.
- L. V < 8/20, astigmatic, lines figures at 2 m. Cylinder of 9.0 D.

Optic Fundus:
- Right eye: +4.0 D.
- Visual fields extremely contracted, see chart opposite.
- Iris hypoglossus.
- Retinae cilio corneal, anterior in both eyes.
- Note the posterior pole. The optic disc is seen to be atrophied, a grayish color, in both eyes: the retinal vessels are narrowed to thin lines.
- There are irregular, ill-defined crescents on the other side of both discs. There is extensive pigmentation of the retina in spots and lines, chiefly in the periphery of the fundus, in some places adhering to the retinal sheath, some with branching processes anastomosing in a typical manner.
- There are several irregular patches of choroidal atrophy in both eyes, in some cases reticulated by pigment masses.
Case XX.

Case occurring in a boy aged 10. Earliest symptoms noticed when between 4 and 5. No right blindness.
No conjunctivitis. No cause. Vitreous syphillis.

Walter Brown, 10. Att School.

Mother states that when the child was between 4 and 5 years of age, he noticed that whenever he dropped his toys on the floor, he had difficulty in finding them, also he seemed to hold objects very close to his eyes; when he was 6 years old, the doctor told him to Horrofields where he was examined for glasses and -3.0D ordered R.E. spectacle was taken at that time of the palpebral conjunctivitis, but no treatment is made of any pigmentary deposits.

Patient was again brought to Horrofields in November 1974, on account of his teacher having complained of the boy's sight. Retinitis pigmentosa was diagnosed at that time and these notes taken.

His mother has not remarked any right blindness. He can find his way about at dusk, and plays outside in the evenings. Mother states that he was quite healthy up to 3 years of age when he had an attack of coughing cough, soon after this he had measles followed by an enlargement of the glands in the neck, these were opened and there is a cicatrice now to be seen on the left side of the neck. Patient is an intelligent looking lad, he has rather a prominent forehead, but no atrophy of the teeth, cicatrices about mouth.

Family History:—Father is a stoker on the railway, he is in
"Centre each chart with "pointer" at Zero before commencing to use the Automatic Registration."

LEFT.

RIGHT.

PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal field of Indirect Vision, the small red circle the position of the blind spot.

Designed for use with Prof. Mr. Hardy's Registering Perimeter. Published by Messrs. Curry & Paxton, 195, St. Portland St., London, W.
good health and has good sight. Further living, age 74, she has never complained of her sight.

She was born in West Brom, was there in Shropshire, they were not related before marriage. In history of consanguinity in grandparents. She has 7 brothers and sisters living, no one is one of 9, all living, all of them have good sight, and none complain of night blindness. One of 3 sisters comes suffers from jobs.

Patient is the 2nd of 5 living children, one boy aged 7 has an absence in the cheeks, but the others are all healthy, and show no signs of congenital syphilis. Further has a third nothing peculiar about their sight. Further has had no miscarriages, but often patient has been, attributed in each case to lifting heavy weights. There is no congenital anomaly or hereditary disease of any kind in the family.

Present Condition: - Vision R 2/60 F 4 + 0.25 F T + 0.00.

Pupils Round, under achromat R + 0.25 L + 0.25.

Visual fields - uniformly contracted, no see charts opposite.

Small dots? float in the interior in both eyes.

In conjunctivae: Clients bear defective, with light green

Refractive Ig xray, under achromat R + 0.25 L + 0.25.

No visual fields: uniformly contracted, no see charts opposite.

Small dots? float in the interior in both eyes.

In conjunctivae: Clients bear defective, with light green

Refractive Ig xray, under achromat R + 0.25 L + 0.25.

In the peripheral of the fundus in binocular spots with branching processes, anastomosing, some can be seen clinging to the sector MALDON FOR A CONSIDERABLE JOURNEY OF THEIR LENGTH.
Case XXI

Case of retinitis pigmentosa, probably congenital. Father and one sister also affected. No consanguinity.

Night blindness, a very early symptom.


Patient has complained of night blindness all her life, she cannot tell when she noticed it first but it has been present as long as she can remember. She never was able to see well in the twilight or dusk. She is quite unable to find her way about in the evenings, and has not gone out alone at night for many years. She is now in service in London and her mistress has sent her to Bournemouth to see if anything can be done for the symptom. She has never had her eyes examined before.

The patient states that she reads quite well in a good light.

Family History:—Father, aged 62, living in a bad sight.

Mother aged 59, she has very bad sight, and has complained of night blindness for many years. She has never had her sight examined and refuses to come to Hospital.

Father and Mother were not related before marriage. Both were born in Surrey though not in the same village.

Their grandparents are all dead, the niece heard that they suffered from anything of this kind, does not know if they were related before marriage, but does not think they were.

Patient is one of 6 children, all have good sight except one married sister, who complains of symptoms exactly like her own, inability to see in a dim light or at night time. Patient says she cannot persuade her to come.
"Centre each chart with 'pointer' at Zero before commencing to use the Automatic Registration."

LEFT.

RIGHT.

PERIMETER CHARTS.

The eccentric continuous red line indicates the average normal Field of Indirect Vision, the small red circle the position of the blind spot.

In tropical and the will not allow her eyes to be examined, she is separated from her husband and in a situation we considered she has no family. On further the child inflicted pain fits. No signs of congenital syphilis infected.

Visual Fields contracted a little, chiefly on upper and outer aspects. Colour Sense quite normal, with light green chooses all shades of green and no others, with rest chooses red only.

No symptoms, and no anterior synechia.

Ophthalmoscopic appearance: - Iris looks greyish white, vessels narrowed. Some superfiicial choroidal chorioretiny below and to the outer side of the discs, crescentic in outline. There is a plentiful deposit of pigment in streaks and dots, and some most typical brown crescentic like bodies, arranged in many cases alongside a vessel with their processes crossing it and joining one another, this pigment is thickened over the greasing of the fundus, and is very numerous below the discs.

This would explain the contraction of the upper part of the visual fields. The same place the pigment presents the appearance of fine black lacework.

____________________
Remarks.

These cases, 21 in number, occurred in 15 families; in 4 instances, more than one in the family was affected; in 3 as brother and sister, and in 1 as uncle and nephew.

12 cases occurred in males, and 9 in females.

In 8 cases the disease commenced in early childhood, and in 1 case it occurred as late as the 47th year. Of the remaining cases, the average age of commencement was 25 years.

In 3 instances, the patients were Jews.

With regard to cause:—In 6 cases there was a doubtful history of heredity, and in only 2 (father and sister), was there any history of consanguinity.

In 3 cases material shock was given as a cause.

In only 2 instances was associated syphilis proved to be present.

In 9 cases there was a history of insanity, or of some obscure nervous or mental condition occurring either in the patient or in members of the family. This would seem to agree with Dr. Gour's observation associating cutaneous pigmentation with certain nervous conditions of the nervous system.

Of congenital anomalies, 4 cases occurred in deaf-mutes, 1 in a patient of dwarfed stature, 1 in an epileptic, and 1 in a patient with defective intelligence.

In 4 cases deafness was associated.

Asthenia or prostration, joint contract, occurred in 4 cases.

Gyphagnosia in 5 cases. Hallucinations in 2 cases.

As regards refraction, 5 were myopes, and 14
In 9 cases, it was not ascertained.

In 9 cases, 2 cases were noticed.

In 6 cases, posterior cortical localization was.

With regard to colour sense, in 7 cases, it was found to be impaired, and in 2 quite normal; in 9 cases, it was not ascertained.

These cases were obtained from the practice of the Royal London Ophthalmic Hospital, Moorfields, the Middlesex Hospital and from the ophthalmic department of the Glasgow Hospital for Injuries.

I have to thank Dr. Lamb, Dr. Sanford, Dr. Giese, and Dr. Julius Jacobson for kindly allowing me to take notes of the cases and to visit them at their own hospital.