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PLATE I.

FUNDUS SKETCH, CASE II—RIGHT EYE.

The disk is well defined and atrophied. The vessels are very small but well distributed. The veins are dark and without light streaks. The pigmentation is of the classical variety, *i. e.*, feathery and branching, and completely encircles the equator. Although very evenly deposited, the pigment is rather densely packed around the vessels in a number of places.

Both retina and chorioid are markedly sclerosed. The case is thirteen years of age, has good central vision and a fair field.

RETINITIS PIGMENTOSA

WITH AN

ANALYSIS OF SEVENTEEN CASES OCCURRING IN DEAF-MUTES

BEING AN ESSAY FOR WHICH WAS AWARDED
THE ALVARENGA PRIZE OF THE COLLEGE OF
PHYSICIANS OF PHILADELPHIA, JULY, 1908

BY

WILLIAM T. SHOEMAKER, M.D.

PHILADELPHIA

LABORATORY EXAMINATIONS OF THE BLOOD AND URINE

BY

JOHN M. SWAN, M.D.

PHILADELPHIA

WITH ILLUSTRATIONS AND THREE COLORED PLATES

"Valeat Quantum Valere Potest"

PHILADELPHIA

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PREFACE

THE opportunity of having under observation for several years a number of cases of retinitis pigmentosa in deaf-mutes, seemed to the author one which might, if improved, lead to a better understanding of this interesting and important disease.

With this idea in mind, seventeen cases were selected for study from among the pupils of the Pennsylvania Institution for the Deaf and Dumb at Mt. Airy, and from the Home for the Training in Speech of Deaf Children before they are of School Age, at Bala.

The cases selected were all true exponents of the disease, any presenting chorioretinitis of doubtful significance having been rigidly excluded. By including questionable cases, a larger but less reliable series could have been reported.

A careful study and analysis of these cases form the basis of this essay, and the author has endeavored to draw logical conclusions from his observations, to discuss the work of others, and to review as far as possible the vast literature germane to the subject.

To Dr. Mary Buchanan the author wishes to express his indebtedness and appreciation for the most excellent water-color sketches here reproduced, and for

her careful and difficult work in plotting the visual fields.

The elaborate analyses of the blood and urine made by Dr. John M. Swan in the Laboratory of Physiological Chemistry of the University of Pennsylvania, constitute a valuable contribution to the subject, for which appreciation is here acknowledged.

W. T. S.

2031 Chestnut Street.

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RETINITIS PIGMENTOSA

CHAPTER I.

HISTORY AND ANATOMICO-PATHOLOGICAL CHARACTERISTICS.

Retinitis Pigmentosa, Chorioretinitis Pigmentosa, or Pigmentary Degeneration of the Retina, is characterized by anatomical changes and symptoms so pronounced and definite as to make the disease one of the most easily recognized in the domain of ophthalmology. Although many of the cases described and pictured in text-book and atlas as most typical are in reality atypical, the essential changes are such that, be the case typical or atypical, they can scarcely be overlooked or misinterpreted.

History.—Fifteen years before the invention of the ophthalmoscope, by Von Helmholtz in 1851, Max Langenbeck made mention of pigment occurring in the retina, under the name of *melanosis retinæ,* and two years later, or in 1838, Von Ammon published two pictures representing what he termed "*getigerte Netzhaut,*" showing flakes of pigment in the nerve-fibre layer of the retina, situated mostly at the periphery, and diminishing in numbers toward the centre. As there

are now recognized clinically and anatomically a number of chorioido-retinal conditions associated with pigment, the eyes examined by Langenbeck and Von Ammon may have been, or may not have been, the seat of pigmentary degeneration of the retina.

E. Jäger was perhaps the first to note the disease with the ophthalmoscope. He had in his possession in 1853 two pictures, which, however, are not published, portraying the ophthalmoscopic appearances of the affection in a patient with posterior polar cataracts discovered by Van Trigt (A. Wider). The first published drawings of retinitis pigmentosa were by Reute in 1854, who did not, however, designate the condition other than a disease of the human eye.

It remained for Albrecht von Graefe to produce an exact ophthalmoscopic description of the disease in 1856, and this great master did more than any other to establish a true understanding of the process. He early called attention to heredity as an etiological factor, and believed it to be an important one, and he recognized the absence of a true inflammatory process, favoring rather as the cause, a deep-rooted trophic disturbance. Donders made pathologico-anatomical studies, and gave to the disease the name of retinitis pigmentosa in 1857, a name which, although most convenient, does not accurately describe the condition.

The Anatomico-Pathological Characteristics. —The essentials of these are to be found in the retina,

the chorioid, the optic nerve and the blood-vessels, all of which structures may be said to be affected in every case. In a certain number of cases changes are also found in the crystalline lens and in the vitreous. In an important contribution to the pathological anatomy of retinitis pigmentosa, Wagenmann describes the following conditions as found in a typical case of the disease:

The retina was everywhere thickened and degenerated, showing extensive connective-tissue proliferation. In many places it was intimately adherent to the chorioid, but without interchange of fibres. The two structures were at all points differentiated by the basal membrane. The rod and cone layer was completely destroyed, and in its place was found connective tissue in which were single cells with unusually large nuclei. The pigment layer was composed in most part of regular cells, some well pigmented, some poor in pigment, and some devoid of pigment. In some places the pigment epithelium had entirely disappeared, while in other places the cells had proliferated and were superimposed, making several layers, and were pushing into the retinal tissue. In form and size, the pigment epithelial cells showed many variations from the normal.

Degeneration of the nervous structures of the retina was extensive. The nervous elements, with the exception of the fibre layer, were entirely lost to within a zone close to the macula. The retinal vessels were greatly sclerosed. The pigment deposits in the retina

were extensive, and especially marked in the inner layers. Most of the pigment was in cells; some of it, however, was free. The walls of the vessels contained much pigment located partly in the outer sheaths, and partly in the cells within the vessel walls. Cross sections of vessels completely obliterated by pigment packed within the lumen were found. The macula lutea was well differentiated as a distinct excavation. The cells of the pars ciliaris retinæ were deeply pigmented and overgrown. Between the epithelial layer of the pars ciliaris and the chorioid, was a thick layer of newly formed fibro-connective tissue.

In the optic nerve, the interfascicular connective tissue was greatly thickened, the nerve-fibre bundles were thin and distinctly atrophic. The central vessels showed very little change, but the smaller vessels, especially the capillaries, showed a marked hyaline degeneration of their walls. The capillary walls were very thick and their lumina very narrow.

The chorioid was thickened, due to a rich increase of the intervascular stroma, which contained considerable pigment. The walls of the smaller and larger chorioidal vessels were thickened and sclerosed; the structure of the adventitia was indistinct, opaque and granular. In many places, the choriocapillaris could not be made out with certainty; in others it was distinct. Drusen bodies, found by other observers, were absent in Wagenmann's case.

The ciliary processes were poorly developed and without their usual branchings. The ciliary muscle was normal. Endarteritis was found in many of the vessels of the iris. The vitreous also showed changes.

The pathological changes above noted have been confirmed by others, especially by Burstenbinder and by Deutschmann, who report similar examinations of the eyes from cases of retinitis pigmentosa.

These anatomical changes are distinctly degenerative in character, and little evidence can be found of a true inflammatory process having existed at any stage of the disease. Burstenbinder, however, while confirming the pathological findings of Wagenmann and Deutschmann, found in his case leucocytes also, and concludes from this that the case was of inflammatory origin. Thomas Windsor was also of the opinion that the disease is probably inflammatory in nature, and that the altered nutritive processes in the adjacent portions of the chorioid, the inner layers of the retina, the optic nerve, the vitreous, and the lens are secondary.

Ferentinos, on the other hand, holds that the disease is neither a degeneration nor an inflammation, but is essentially a simple atrophy of the retinal cells, brought about by physiological conditions. The involution, he thinks, takes place early; the functionally less active regions are affected sooner than the more highly developed portions of the retina; a gradual loss of function is associated with a gradual diminution in the

calibre of the vessels supplying the retina. Before, however, the vascular contraction has adapted itself to the lessened demands of the retinal cells, the connective tissue, receiving more than the usual supply of blood, proliferates.

Regarding the tissue of origin, whether chorioid or retina, there has been much discussion. It is probably the former, for, as Wagenmann contends, the evidence as to the origin of typical retinitis pigmentosa is overwhelmingly in favor of the chorioid. This, he says, can be demonstrated experimentally in rabbits by interference with the chorioidal circulation. That the chorioid and retina are not simultaneously attacked is, according to him, not proven.

Oeller, in 1879, declared the characteristic vessel changes to be chiefly in the choriocapillaris and the smaller vessels. The degeneration, he thought, could scarcely be considered as *primary,* but rather as secondary to chronic chorioiditis, and resulting perhaps from the pressure of extensive lymphoid cell infiltration. Pagenstecher, Müller, Schweigger, and Rednew describe a *vitrifying* of the vessels of the chorioid in certain instances, completely obliterating their lumina. Definite changes were noted in the endothelium which were thought to be the underlying cause of the characteristic vessel changes.

The Pigment is Retinal.—According to Camill Hirsch, pigment in the retina can come from but four

sources: First, from the pigment layer of the retina; second, from the chorioid; third, from the blood (hematogenous pigment); and fourth, from iron introduced within the eyeball,—siderosis bulbi. Sources one and two only are germane to the subject under discussion. Chorioidal pigment very seldom finds its way into the retina, and then generally after an injury establishing a direct communication between the two membranes. On the other hand, the retinal pigment cells depend entirely for their nutrition and normality upon the chorioidal circulation. When this is sufficiently disturbed, they undergo degeneration with all its consequences. The pigment is then translated; it *wanders,* and in retinitis pigmentosa eventually reaches the nerve-fibre layer of the retina, in advance even of the retinal vessels. The pigment may be free, or contained within the cells which have likewise passed through the retina. Such changes as are here described are never produced or influenced by disturbances in the retinal circulation alone.

The character of the pigmentation, which will be described later, has been much dwelt upon as being of clinical significance in diagnosing retinitis pigmentosa from other conditions associated with pigment deposits in the retina, notably, chorioretinitis syphilitica. It would seem to the author that the fine feathery or branching deposits found in true retinitis pigmentosa, as contrasted with the more or less gross, irregular pig-

ment massing resulting from chorioretinitis, would be further evidence of a slow degenerative non-inflammatory process in the former disease. It is true that post-inflammatory pigment deposit may simulate very closely that secondary to simple degeneration, but there is usually an unevenness or irregularity about it which points to a more rapid and less systematic process.

The method by which the pigment enters and traverses the retina is not certainly known. Hirsch considers four possibilities as follows:

1. Independent movement (ameboid) of the pigment epithelial cells themselves.

2. Direct proliferation, or *pushing* of the cells into the retina.

3. Transportation by leucocytes. Pigment molecules, liberated from the pigment cell, are taken up by leucocytes.

4. Through the lymph spaces within the retina.

He concludes that all four methods may be in evidence, and that none is exclusively a factor.

Retinitis pigmentosa sine pigmento has been frequently seen clinically, but no microscopical examination of an eye so affected is on record (Nettleship). Donders, although he gave to the disease the name of retinitis pigmentosa, thought that pigment deposition was not the essential feature, but was more often secondary. In this belief he was undoubtedly correct.

Schweigger regarded the presence of pigment of

accidental significance, and noted that there was no relationship between the amount of pigment and the grade of retinal atrophy, adding that the process could progress far without the deposit of pigment. Leber, in treating of anomalous forms of retinitis pigmentosa, states that the essential factor in the disease is not the pigmentation, but an interstitial hypertrophy of the retinal elements. He observed several cases in the beginning when there was no pigment, which later took on pigmentation. Pettelsohn, from a study of these and other observations, concludes that retinitis pigmentosa sine pigmento is identical with retinitis pigmentosa, and is only an early stage of the same. He reports a case of his own before and after the deposit of pigment.

Among the author's seventeen cases, one is *sine pigmento,*—Case XI, a girl eleven years old. In this case the degeneration is extreme. It will furthermore be noted in the summary of this series, that the ages of seven cases showing a small amount of pigment range from eight to fifteen years, while the ages of eight cases showing extensive pigmentation range from ten to nineteen years. There can be no doubt that the pigmentation of the retina in this disease is secondary, and little doubt that it will eventually be found in every case.*

---

* Recent examination of Case XI, five years after original record, shows a small amount of pigment around periphery, both eyes.

Cataract, usually posterior polar in variety, is found in some cases of retinitis pigmentosa, and although Graefe thought that it was present in one-third of the cases, this estimate is probably very much too high. In the series here detailed, cataract was found once, the opacity being posterior cortical, and not typically polar. The posterior polar cataract of retinitis pigmentosa is probably congenital, and represents a remnant of the fetal vascular tunic of the lens, or is a variety of retained hyaloid. Posterior polar cataract is more frequently observed in cases of chorioiditis than in retinitis pigmentosa.

The vitreous changes are not very pronounced. It has been noted by Landolt and by Hirschberg that the layer of vitreous next to the retina at times becomes thickened and adherent. The former observer also found fluidity of the vitreous in some cases, which observation was confirmed by Leber. Vitreous opacities have been generally considered to be uncommon in the typical uncomplicated cases, a conclusion not substantiated by our cases, among which four, or 23.5 per cent., showed vitreous opacities. According to Loring, the pigment passing through the retina may even pass the internal limitans and reach the vitreous. Under such circumstances, these minute particles of pigment would be seen as vitreous opacities. Mauthner became convinced that the small pigment fleck often seen in the visual axis in retinitis pigmentosa was not really on the

lens capsule as commonly supposed, but in the vitreous at the centre of motion of the eyeball, a millimetre or so behind the lens.

Small pigment spots are very common on the posterior capsule of the lens, irrespective of disease. Attention has been called to them a number of times, especially by Norris some years ago, and more recently by Zentmayer. Many of them are, doubtless, remains of the fetal vascular tunic of the lens, and if found in association with retinitis pigmentosa, should be recognized as having no pathological connection. Frost noted a case of retinitis pigmentosa in which there was a deposit of pigment on the *anterior* lens capsule.

Persistent hyaloid artery, not an uncommon anomaly, in some of its various forms, has been noted a number of times in cases of retinitis pigmentosa.

Glaucoma, inflammatory and non-inflammatory, has also a number of times been recorded by Schundhaüser, Heinrichdorf, Bellarminoff, Carbone, Goldzieher, Strachow, Mandelstamm, and others. In the case reported by Heinrichdorf, it was bilateral and non-inflammatory, and in conjunction with myopia, in a twenty-two year old subject. The other cases were forty years old or more. An interesting discussion would be the modus operandi of glaucoma in eyes thus degenerated.

CHAPTER II.

SYMPTOMATOLOGY.

The symptoms of retinitis pigmentosa are in close harmony with the anatomical changes, and are for the most part constant, although notable exceptions to the rule are occasionally found. The disease is, with few exceptions, noted mostly by competent observers, always bilateral. The first case of unilateral typical retinitis pigmentosa was recorded by Pedraglia in 1865. Deutschmann examined this eye pathologically nearly thirty years later, and in his account of it mentions but two other unilateral cases (Ancke and Günsberg). Moon reported a case which the author has had the opportunity of examining. In the judgment of the latter, the case, aside from showing a collection of pigment in a localized area of the retina above, did not correspond in any respect with retinitis pigmentosa. That one-sided retinitis pigmentosa can exist is perhaps possible, but from the very nature of the process, the writer thinks it extremely doubtful if such cases could ultimately maintain that distinction.

The essential changes of retinitis pigmentosa, if not congenital, make their appearance at a very early date. True, cases are first noticed perhaps in childhood or about puberty, but when first seen, the degeneration is

almost invariably far advanced, showing a long existence. The earliest symptom is night blindness; a disproportionate reduction of vision under reduced illumination, due to torpor of the retina. The degenerated retina fails to respond to any but a strong light stimulus, and vision fails greatly under a lesser stimulus. An exception is found to this in a few rarely observed cases in which there is retinal hyperesthesia, and in which vision is improved when the illumination is less intense (Leber, Hasse, de Wecker). Hasner (quoted by Stellwag) reported a case of retinitis pigmentosa with day blindness instead of night blindness, and enlargement of the visual fields under reduced illumination.

The visual field is very characteristic. Concentric contraction for form and color is invariably found, and in most cases is very pronounced. Indirect vision, always poor, is often entirely lost. The victim betrays the defect by stumbling, and by collision with surrounding objects when walking. The subjective sensation of a concentrically contracted field has been likened to looking through a long, narrow tube, and the shifting of the eyes and head to take in as much as possible or necessary of the outside world, together with the hesitating and uncertain bearing of the patient when walking, give to him a characteristic appearance which, especially in the case of a young subject, can well excite suspicion.

Central vision, in marked contrast to peripheral

vision, and to the degenerative changes in the retino-neural tract, remains good in most cases, and in many cases normal, despite these changes. Central color perception is likewise persistent as a rule until the disease has reached a very late stage. The color fields suffer greatly, and become extremely narrow. The tendency would seem to be for the form and color field boundaries to approach one another in a common *circle* around the fixation point as the centre. This is shown in the accompanying series. Color blindness, unless congenital, and independent of the disease, is, according to Berry, not a part of retinitis pigmentosa. Eventually, all vision is lost, but so slow is the progress of the disease, that absolute blindness results usually quite late in life. Useful vision, however, may be lost many years before.

It will be noted that in those cases which have come to pathological examination, few or no changes were found at the macula, or in the corresponding chorioid, a circumstance which will account for the preservation of central vision far beyond what would be expected from the nature of the disease, and its ophthalmoscopic appearances.

A ring scotoma, partial or complete, has been found to occupy the visual field in some cases, and has been the subject of much discussion and investigation. It has been interpreted and misinterpreted in various ways, forming as it does, a splendid phenomenon around which to theorize.

In a recent contribution upon *Ring Scotoma,* by W. Ilbert Hancock, the literature has been reviewed, and many important conclusions drawn. Two cases are reported, the one following lightning stroke, the other occurring in a case of retinitis pigmentosa sine pigmento. Quoting from Hancock, ring scotoma might be met with in six clinical conditions as follows:

1. Retinitis pigmentosa (including R. p. sine pigmento).
2. Retinitis, chorioiditis, and chorioretinitis.
3. Lesion of the optic nerve, with or without optic neuritis.
4. Glaucoma.
5. Idiopathic night blindness.
6. Myopia.

Of these conditions, he says, it occurs most frequently in retinitis pigmentosa, is probably one of the earliest symptoms, occurring in the majority, if not in all of the cases, is always progressive, and *has no definite relationship to the area of most marked pigmentation.*

This last observation, together with Hancock's case of ring scotoma in retinitis pigmentosa sine pigmento, is in strong confirmation of the views early expressed by Schweigger and others, that the pigment deposit was accidental or secondary, and not of essential importance to the disease.

In not one of the thirteen cases of this series, in which it was possible to get the visual fields, was a ring

scotoma, or a scotoma of any kind found, and while realizing the sources of error in the perimetric examination of patients, such as those here reported, the author thinks that further confirmation is needed before ring scotoma can be accepted as occurring in the majority, if not all, cases of retinitis pigmentosa, as stated above.

To explain ring scotoma, Hancock gives and analyzes the six existent theories. These are:

a. Attributes the scotoma to a lesion of the retinal vessels, and accounts for its ring-like character by the arched course of the vessels surrounding the macula (Hersing; Ole Bull).

b. The anatomical arrangement of the venæ vorticosæ. The chorioid at the equator is more richly supplied with blood than at other parts, and is therefore more likely to become diseased (Schön).

c. The scotoma is explained as being the result of an exudation. The form is accounted for by assuming that the tension of the chorioid and retina is less at the equator than in the anteroposterior directions, therefore, exudation from the chorioid would begin at the equator, causing a circular defect (Crzellitzer; Wellendorfer).

d. Claims ring scotoma to be an expression of impaired function of the retinal rods, and that it can be demonstrated in every case of night blindness irrespective of causation (Heinrichdorf).

e. The equatorial belt of the fundus is situated at the point of meeting of the anterior and posterior ciliary

systems; it is therefore not so well supplied as other parts of the eye. Nettleship regards the equatorial region as a sort of divide between the two systems, and not efficiently served by either, while the blood current must be slowed or reversed in many of the anastomosing twigs. Given a congenital condition, as in retinitis pigmentosa, in which there is a tendency to atrophy or hardening of the inner layer of the chorioid, changes would, he thinks, show first where the nutrient stream is weakest (Gonin–Nettleship).

f. Ring scotoma is best explained by a retrobulbar lesion of the optic nerve (Graefe, Leber, Burnett, Gallus).

In addition to the above six theories for the production of ring scotoma, Gallus advances still another, and includes in it an explanation of the pathology of retinitis pigmentosa. His theory is, however, according to Hancock, not supported by the facts as found. Gallus advocates a retrobulbar origin for ring scotoma. He assumes the primary cause to be compression of the optic nerve and the ophthalmic artery in the bony optic canal due to chronic periostitis. The pigmentation is explained as due to degeneration and thickening of the ciliary arteries, which he asserts are necessary results of long continued pressure on the parent trunk. All of the theories for ring scotoma are open to objection so forcible that the pathogenesis of this phenomenon must still remain in question.

In summarizing his studies, Hancock, excluding rare cases of idiopathic night blindness, glaucoma and myopia, places all other cases with ring scotoma in one of two groups as follows:

A. Cases in which the chorioid and retina are diseased.

B. Cases in which the fundus is ophthalmoscopically normal.

In class *A*, the diseased fundus corresponds to the scotoma in the field; or there may be no such correspondence.

Two separate hypotheses explaining ring scotoma are recognized as defending an intra-ocular and an extra-ocular causation respectively. In considering those theories based upon the anatomy and circulatory arrangements, around the equator, it is well to remember that there are cases recorded in which the extreme outward limit of the ring does not extend beyond 15 degrees from fixation, and that Hallide and others have proven that the equator is situated 67 degrees from the fixation point. The want of definite relationship between the ring and the retinal area of most marked pigmentation is another obstruction to some of the intra-ocular theories.

With regard to the extra-ocular theories, Parsons has shown that we know definitely of but one specialized bundle in the optic nerve—the macular bundle. It is, however, generally thought that the most peripherally

situated fibres in the optic nerve supply the intermediate zone of the retina; and that the periphery of the retina is supplied by fibres more centrally located in the nerve. Destruction, therefore, of the peripheral fibres would give a field defect in the intermediate zone, or a ring scotoma, but the position of the ring scotoma as found clinically varies so much, that correspondence with the optic nerve bundles becomes very indefinite.

OPHTHALMOSCOPIC APPEARANCES.

A glance at the fundus in retinitis pigmentosa usually determines the diagnosis, and further observation is mostly in the interest of variation and detail. The optic disk, the blood-vessels and the retina itself invariably show unmistakable changes. The disk is atrophied, and presents a dull, opaque surface, the actual color or appearance of which has been variously described as yellowish-gray, yellowish-red, pale gray with a yellowish tinge, or like bee's wax or old parchment. It finally becomes dull white (de Schweinitz). The surface is flat and shows no excavations, even the porus opticus being as a rule obliterated. The margins are not lost, but in most cases are blurred, while in some they remain well defined. A similarly appearing disk can result from old chorioiditis, but when seen in a young subject (with the vessel changes) it is pathognomonic.

The vessels are always small, the veins and arteries

approaching a like diameter. The vessel walls are greatly thickened at the expense of the lumen. The smaller vessels and the terminals are no longer visible, many of them being completely obliterated. Thus there is a reduction in number of vessel units. It is not uncommon to see a portion of a vessel completely ensheathed with pigment (as shown in Plate III). Sometimes fine white lines accompany the vessels for a short distance from the disk, such as result from a moderate perivasculitis. The light streaks are diminished or lost in proportion to the increased thickness of the vessel wall and the narrowed lumen. Loring says he has often noticed changes in the light streaks early in the disease before other characteristic changes were marked.

The pigmentation is characteristic of a sclerotic process as opposed to inflammation. It is of very slow and gradual deposition, and is not therefore seen in large or dense masses. Particles do coalesce to an extent, but the result is quite different from a post-inflammatory accumulation. The pigment wanders into and through the retina in minute particles, appearing first ophthalmoscopically near the equator.

Stellate particles when in sufficient number have their processes approaching or in contact, and the general picture is well described as being bone corpuscle-like, or simulating the Haversian canals. Many feathery deposits are seen, as well as short lines which probably represent the position of vessels too small to

PLATE II.

FUNDUS SKETCH, CASE XII—LEFT EYE.

Shows a sharply defined, atrophied disk, which is flat, there being no trace of the porus opticus. The vessels are very much reduced, the veins relatively more so as they are about equal in diameter to the arteries. The whole chorio-retinal expanse is sclerosed, and the fundus is characteristically dull in color and lacking in normal brilliancy. Toward the periphery the disturbance in the pigment epithelium of the retina is most marked. Where the pigment is absent, and the chorioidal pigment likewise disturbed, the sclera shines through, giving the effect of white patches.

The pigment which has reached the nerve-fibre layer of the retina is small in amount. To the nasal side, it is deposited in an unusual manner, as though sprinkled over the retina.

The case is fourteen years old. Corrected vision is good ; the fields are very much contracted.

be seen, or with the lumen packed with pigment. By preference the temporal fundus seems to have the greatest pigment deposit. The pigment, although freely distributed without regard to the vessels, when in contact with the vessels, becomes rather more conspicuous, because it here loses its feathery and branching character, and settles into a more dense accumulation. As the disease progresses, the pigment is deposited nearer and nearer to the posterior pole. It is rarely found on the disk but may accumulate at the disk margin. Occasionally it has been found heaped at the macula. On account of the removal of the pigment from the retinal epithelium, the chorioidal circulation becomes more or less in evidence. The normal fundus color loses its life or brilliancy; the sclerosed retina overlying the partially visible chorioid is distinctly gray; and there is a lack of evenness, and defective definition, similar to a poorly washed surface in water color painting. At times there are white spots toward the periphery which are probably depigmented cells in the retinal epithelium, which do not happen to be backed by sufficient chorioidal pigment to prevent the sclera from shining through.

The ophthalmoscopic appearances in retinitis pigmentosa sine pigmento are identical with those of retinitis pigmentosa, except that there is no pigment deposit. The evidences of sclerosis and degeneration are the same.

CHAPTER III.

ETIOLOGY.

Notwithstanding the collection and examination of almost innumerable statistics by the most competent observers, the etiology of retinitis pigmentosa remains obscure. The disease is so often found in association with other material conditions, the etiology of which is likewise unknown, that the question becomes from the start most complicated. It might be said that true retinitis pigmentosa is seldom if ever found in an otherwise perfectly normal and standard individual. On the other hand, it is generally found in conjunction with other deep-rooted defects, or at least in the presence of more than the usual number of the stigmata of degeneracy. For example, it is unquestionably associated in many cases with defective hearing or deaf-mutism; with defective intellect (idiocy); is found in a number of the members of the same family; is found in the offspring of consanguineous marriages; and has in association with it, in a certain number of cases, stammering, microcephalus, polydactylism, congenital cataract, nystagmus, glaucoma, and other conditions.

The geographical distribution of the disease would seem to show that no country is without its victims, but, as would be expected, it is particularly prevalent in

Turkey and throughout the East. The Jews are said to be more frequently attacked. Although, according to all previous statistics, the male sex has a distinctly greater predisposition to retinitis pigmentosa than the female sex, our statistics show unequivocally the reverse. According to Leber, about three-fourths of all cases are males. From several series of cases, he gives as an average 72.8 per cent. males and 27.2 per cent. females. Derigs, in 27 cases, found 70 per cent. males and 30 per cent. females. Wider (Tübingen Klinik), in 42 cases, found 66.6 per cent. males and 33.3 per cent. females. Of the 17 cases in our series, 4 were males and 13 were females, or *76.4 per cent. females.*

The most prominent etiological factors and associated conditions which present themselves for consideration, whether or not their influence and relationship can be proven, are:—heredity, consanguinity, syphilis, maternal impression, deaf-mutism and idiocy. With regard to deaf-mutism and idiocy, it is, of course, fair to presume that where one or the other, or both, are found in the same individual, with retinitis pigmentosa, the one is not the cause of the other or another, but both or all are the result of a common cause.

HEREDITY.

Although often or generally attacking several members of the same family, the direct transmission of retinitis pigmentosa from the parents to the offspring

is seldom. Leber found one case of direct heredity in sixty-six cases of retinitis pigmentosa, and Derigs found two in sixty cases. In our seventeen cases of the disease occurring in deaf-mutes, in not a case is there a history of the disease in the parents, or in either branch of the family. There is, however, abundant evidence that the disease has been transmitted through several generations, or that certain symptoms, as night blindness, have been transmitted, appearing in some of the offspring as retinitis pigmentosa, and in others perhaps as idiopathic night blindness, or congenital amaurosis. Heredity would seem, therefore, to have a potent influence. Donders, in 1859, reported a family history in which the father had night blindness without pigment in the retina, and the son had night blindness with pigment in the retina. Mooren, in 1882, reported a mother with retinitis pigmentosa, having two children with night blindness without pigment. In Mooren's case the children might have had retinitis pigmentosa sine pigmento, but in Donders' case, if we regard retinitis pigmentosa sine pigmento as an early stage of retinitis pigmentosa, it cannot be disposed of in this way.

Dumont, reducing to statistics the transmission of blindness (from all causes) from parents to children, thought he proved heredity 68 times in 1168 cases, or in 5.8 per cent. Cunier gives the history of one family in which since 1637, congenital night blindness was indigenous, and in which no less than 125 members of the

family were so affected. The record, however, is one of pre-ophthalmoscopic time, and although it may be presumed that some of the cases suffered with retinitis pigmentosa, there is no positive knowledge of the existing conditions.

Leber found 33.3 per cent. of 66 cases of retinitis pigmentosa in which other members of the family were affected.

Hutchinson found in 23 cases, 43 per cent. in which more than one member of the family were affected.

Nolden established heredity in 2 out of 33 cases, or in 6.6 per cent.

Derigs, analyzing 27 cases, found nine times, brothers and sisters affected, four times parental consanguinity, and twice distant relatives affected.

An interesting family history is reported by Simeon Snell. The father and mother were free from retinitis pigmentosa; there was no history of their ancestors having had it, and no consanguinity. Of eight living children, one daughter was free, and six of the sons had retinitis pigmentosa. The one free was thirteen years old and the youngest. Cases II, III, IV, and V of our series have a somewhat similar family history.

It is practically impossible, however, to establish heredity, or in fact, to learn very much of value concerning the ancestry of children such as those constituting our series. Beyond the generation next preceding, we know nothing definite.

Francis Galton says that the complete heritage of a child, on the average of many cases, might be assigned as follows:—One quarter to the personal characteristics of the father; one quarter to the average of the personal characteristics of the fraternity taken as a whole, of which the father was one of the members; and similarly as regards the mother's side.

James Kerr Love, in applying this principle to the study of deaf-mutism, says, "It is not enough to seek for the cause only in the parents of the affected child, these may hear or be deaf, but that single fact teaches little. Indeed, in the first generation, the tendencies of two congenitally deaf parents may so counteract each other that the result is a hearing child, but reversion will ultimately assert itself. The second generation will probably follow the grandfather or grandmother with greater faithfulness, and a deaf grandson will result; or, the characters of a prepotent progenitor separated by many generations, may crop up, and an unlooked-for outbreak of deafness may take place. In calculating, therefore, the chances of deafness in a family in which it is feared, our view must not only include the immediate progenitors, but the whole family antecedents on the father's and mother's side. This statement is of practical importance; it takes all, or nearly all, the value out of the proposal to prohibit the intermarriage of the congenitally deaf, for, as has been shown, the hearing members of a deaf-mute connection

send down the tendency to deafness with as great certainty as the deaf members."

These principles apply equally well to retinitis pigmentosa when considered from the standpoint of heredity. It is seldom, indeed, that we can obtain any reliable information regarding the antecedents beyond one generation. Genealogy is of little interest to the class of people from which the majority of our material is obtained.

CONSANGUINITY.

Consanguineous marriages and the defects to offspring resulting therefrom, or supposed to result therefrom, have been a fruitful source of discussion and speculation for many years. Opinions still differ widely as to the effects of consanguineous marriages, owing to the complexity of the problem, which becomes apparent at once when we attempt to deduce incontrovertible facts. In the first place, consanguinity is more or less inseparable from heredity, and its results are of necessity, influenced by it. It is, for example, impossible at the present time, to determine whether consanguinity is in itself a cause of congenital defect, or, whether it operates through the intensified transmission of hereditary tendencies prevalent in a family.

The practical importance of consanguinity, in its effect on offspring, is thus thrown into doubt. With good hereditary family tendencies, could they not be accentuated, and a greater refinement of them be pro-

duced, by a consanguineous union? Or, with evil family tendencies, could they not possibly be counteracted and eliminated? It has been claimed by Dr. Arthur Mitchell that the marriage of cousins in higher social states, including hygienic surroundings, greater mental and physical activity, etc., is less often followed by defective offspring, than similar marriages in opposite conditions. Mooren found consanguinity more damaging, the more sedentary the people are, agreeing in this with the views of G. Darwin. In quiet country districts of England, 2.25 per cent. of the cases of retinitis pigmentosa were associated with parental consanguinity, while in the fluctuating population, but 1.05 per cent. According to Mooren, the great fluctuation and activity of the population of North America and Australia render consanguinity of scarcely any importance. Nevertheless, consanguinity would seem to have been a most important factor in the study of retinitis pigmentosa.

Liebreich (1862) first called the attention of ophthalmologists to what he considered the damaging influence of consanguinity as an etiological factor in retinitis pigmentosa. Examining the pupils of the Deaf and Dumb Asylums in Berlin, he found a frequent coincidence of the affection with deaf-mutism, and a number of these cases he found to be of consanguineous parentage. He thought that in retinitis pigmentosa, consanguinity was more frequently found than in deaf-

mutism (without retinitis pigmentosa), idiocy, insanity, etc., *i.e.*, one-third of the cases of retinitis pigmentosa that were associated with deaf-mutism were of consanguineous parentage.

The same author later (1863) found among 100 cases of retinitis pigmentosa, 45 children from consanguineous parents, and he concluded that about 40 per cent. of children with retinitis pigmentosa have related parents. Jews favor consanguineous marriages and are relatively more frequently the subjects of retinitis pigmentosa. In this connection, Mygind says that it is a known fact that the Hebrew race produces a larger number of blind and idiotic individuals than the European races among which it lives. He believes that consanguinity plays an important part. Oakley believes consanguinity to be an important, if not the chief, etiological factor, and he reports three cases. Manhardt (Constantinople) in the examination of many cases, failed to find consanguinity in a single one. In most of his cases syphilis was found.

David DeBeck, in a report of eleven cases of retinitis pigmentosa, and a study of the literature, concludes that consanguinity must be held to be one of the strongest factors in etiology. In 559 cases which he quotes, 157 positively stated that blood relationship existed in the parents, or 28 per cent. His own cases, however, do not show consanguinity, blood relationship of parents existing in but one case.

Hutchinson had in his note-book twenty-three cases of retinitis pigmentosa, in eight of which parental relationship was shown.

The statistics of seventeen observers, elaborately compiled by Wider, show consanguineous parentage in from 13.6 per cent. (Webster) to 60 per cent. (Hocquard) of cases of retinitis pigmentosa, or, an average of 31.8 per cent. The list includes Liebreich, Mooren, Höring, de Wecker and Jäger, Hocquard, Nolden, Leber, Webster, Hirschberg, Hutchinson, Badal, Derigs, and Wider.

Our seventeen cases were the offspring of thirteen marriages, two of which were consanguineous, giving three defective children,—17.6 per cent.

As coming to conclusions not supporting the importance of consanguinity, are Pagenstecher, who, among nine cases of retinitis pigmentosa, found no consanguinity, and Macnamara, who found retinitis pigmentosa very prevalent among Hindus, with whom consanguineous marriages are strictly prohibited by religion.

Ancellon compared the statistics of the offspring of marriages not consanguineous and consanguineous, *to the disadvantage of the first.*

Bourgeois and Seguin published their own genealogies, in which there were frequent marriages of near relations, but were unable to give a single case of deaf-mutism, hydrocephalus, etc., in 200 years.

Child in England, and Bailey in Frankreich, found similar results.

RETINITIS PIGMENTOSA AND SYPHILIS.

No investigation of a pathological condition would be complete until every effort had been made to shift the responsibility therefor upon syphilis. And so it is with retinitis pigmentosa; the relationship has been sought for, but except in the opinion of a few observers, has not been found. Syphilis produces a form of chorio-retinitis, which is sometimes difficult to differentiate from true retinitis pigmentosa, but with the exception of certain cases, such as mentioned by Hutchinson, in which the changes are *degenerative* rather than *inflammatory,* these two methods of attack and destruction can usually be separated, the one from the other. The changes in retinitis pigmentosa are distinctly degenerative, while those in syphilitic chorioretinitis are inflammatory, or inflammatory plus degenerative. Evidences of previous inflammation can usually be seen, yet, no less astute an observer than Galezowski regarded every case of retinitis pigmentosa as of syphilitic origin, either in the parents or the patients themselves. Quaglino likewise regarded syphilis as the most frequent cause of retinitis pigmentosa. Leber, in the old Graefe–Sæmisch, says that in no case in which he could with probability place the cause as syphilis, were all the symptoms of typical retinitis pigmentosa present.

Manhardt found syphilis in most of many cases examined. Wider, among his cases from the Tübingen Klinik, was unable to establish syphilis as a cause in a single typical case, but did find atypical unilateral retinitis pigmentosa of syphilitic origin.

Baumeister holds that in unilateral retinitis pigmentosa, the presumption of a syphilitic cause is justifiable. The writer would suggest that the presumption of a syphilitic cause in unilateral cases is more justifiable than the diagnosis of retinitis pigmentosa.

J. Hutchinson is strong in the belief that syphilis is not a factor in etiology; he says,—" Of the causes of true retinitis pigmentosa we know nothing, but it is certain that it has nothing to do with syphilis."

None of our cases showed the unmistakable outward signs of syphilis, but it was impossible to investigate very thoroughly along this line, so that no positive statement concerning syphilis can be made from this series.

The prevailing opinion among ophthalmologists is, however, that syphilis plays a very doubtful, if any, part in the causation of retinitis pigmentosa.

MATERNAL IMPRESSION AS A POSSIBLE FACTOR IN THE ETIOLOGY OF RETINITIS PIGMENTOSA.

Although strenuously denied and ridiculed as impossible by some writers of the present day, there are, nevertheless, so many authentic and well reported cases

of defects in offspring, mental and physical, following profound shock or impression upon the pregnant woman, that, until the possibility of maternal impression has been annihilated by else than theory, we must continue to give to the doctrine due consideration. Of the many cases reported, most are subjects of physical defect, and most would seem to result from a sudden violent shock to the mother's nervous system, cognizance being through the sense of sight. Some, however, would seem to follow a prolonged impression of a milder nature, one accompanied by anxiety or fear; and still others have had the character of a reproduced injury to the mother, such as a burn, and this, too, within a few hours of labor (Whiting).

Dr. Wm. C. Dabney has collected and placed in tabular form, ninety cases of physical defect associated with a maternal impression. These cases have been carefully selected, and are reported by competent observers, and it would be difficult to dispose of them as merely accidental, or coincidental, as would for instance, Dr. E. T. Shelly (*Superstitions in Teratology, with Special Reference to the Theory of Impressionism*) who opposes, without convincing argument, the theory of maternal impressions as absurd and untenable.

Priestly Smith reports retinitis pigmentosa connected with a history of maternal shock. There were no constitutional nerve disorders in earlier generations, and no consanguineous marriages. A woman who had previously borne two healthy children suffered a nervous

shock during the early months of her third pregnancy. The child, and all other subsequent children, except one which died in infancy, developed retinitis pigmentosa with partial deaf-mutism. Smith believes maternal impression or shock to be a not improbable cause in this case.

Given a pregnant woman who has previously borne a defective child, for example, deaf, it is not unreasonable, in the opinion of the writer, to think it possible for her fear and anxiety lest the coming child be likewise affected, to so influence her pregnancy as to result disastrously. And we do find among deaf-mutes of large families a preponderance of consecutive defectives. On the other hand, the greatest irregularity may also be found.

In our series, we have in one instance of seven children, the 6th and 7th deaf-mutes; in another family of five children, the 2nd, 3rd, 4th, and 5th deaf; in still another family of eight children, the 4th and 5th are deaf. As examples of irregularity, one family of eleven children has the 1st and 7th deaf, and in a family of five, the 2nd only is deaf.

Mygind, in considering the influences during pregnancy bearing upon congenital deaf-mutism, says: "Although there can be no doubt that the connection between such maternal impression and deaf-mutism is often purely accidental, it cannot, on the other hand, be denied, that there is a possibility of powerful influence

during pregnancy, causing such considerable abnormalities in the auditory organs of the fetus as to result in deaf-mutism."

He refers to the statistics of Lent, who, among 303 deaf-mutes, found 13 cases ascribed to maternal impression. Wilhelm found 15 out of 519 deaf-mutes, and the Danish statistics show 11 among 553 deaf-mutes, declared to be the result of fright, grief, or agitation, etc., during pregnancy.

The evidence in favor of the possible baneful influences of maternal impression, the author thinks, is as yet too strong to be eliminated in considering the etiology of not only retinitis pigmentosa, but of many other congenital conditions, and he believes that it may be, and probably is, an important factor in causation.

Other causes ascribed by some for retinitis pigmentosa, are typhoid fever, meningitis, scarlet fever, erysipelas, epilepsy, sunstroke and eye-strain.

That retinitis pigmentosa should be first noticed after any one of these diseases, or that a victim of retinitis pigmentosa should also become a victim of any one of these diseases, cannot of course, in itself, show causal relationship. Furthermore, the probable fact that true retinitis pigmentosa is congenital, and the apparent preponderance of evidence pointing to etiological factors, such as heredity, consanguinity, maternal impression, etc., would militate against the acceptance of such causes. That eye-strain could have

any causal relationship to retinitis pigmentosa is to the author inconceivable.

RETINITIS PIGMENTOSA AND DEAF-MUTISM.

All observers recognize the frequent association of these two conditions, but the statistics at hand show considerable variance in the percentage findings. It will be noted that the percentage of retinitis pigmentosa found among deaf-mutes, is much lower than the percentage of deaf-mutism among cases of retinitis pigmentosa in general, and the figures must not be confounded. For example, Hocquard, among 200 deaf-mutes, found 5 with retinitis pigmentosa, or 2.5 per cent., and among 15 cases of retinitis pigmentosa, 5 were complicated with deaf-mutism, or 33.3 per cent. Leber concludes that 20 per cent. of retinitis pigmentosa is associated with deaf-mutism, while Derigs, among 60 cases of retinitis pigmentosa, found but four with deaf-mutism, or 6.6 per cent., a striking difference.

Schäfer, among 95 cases of retinitis pigmentosa, found 5 with deafness, or, 5.2 per cent.

In the Tübingen Klinik, 41 cases of retinitis pigmentosa showed 8, or 19.5 per cent., with defective hearing. From the standpoint of deaf-mutism, Liebreich found 14 cases of retinitis pigmentosa among 241 deaf mutes, or a percentage of 5.8.

Badal observed 7 cases of retinitis pigmentosa among 200 female deaf-mutes, or 3.5 per cent.

The author's 17 cases were found in the examination of about 700 deaf-mutes, which would give a percentage of 2.4. The percentages of retinitis pigmentosa among deaf-mutes as found by Hocquard (2.5 per cent.), Liebreich (5.8 per cent.), Badal (3.5 per cent.), and the writer (2.4 per cent.) are remarkably close. Combined, these four results give an average of *3.5 per cent. of retinitis pigmentosa among 1341 deaf-mutes.*

RETINITIS PIGMENTOSA AND IDIOCY.

It is not surprising, considering the nature of retinitis pigmentosa so far as we know it, that asylums and institutions for the feeble-minded should show among the inmates a liberal proportion of this affection. And still less surprising is it, when we consider the nature of idiocy,—a lack of development, or an arrest of development, of the central nervous system,—that the nervous tissues of the eye, direct offshoots of the brain, should likewise show marked structural defects.

DeWecker, in 1868, noticed the frequent occurrence of weak-mindedness in cases showing an early marked reduction of vision, and Leber found the mental faculties to be frequently underdeveloped in cases of typical retinitis pigmentosa.

In estimating the value of the mental faculties, we must not lose sight of the fact that one, particularly a child affected with retinitis pigmentosa, might well, from the contracted fields, night blindness, etc., appear

stupid; and most certainly would not, under the handicap, develop mentally as a normal child.

Liebreich, among 50 idiots, found 3 with retinitis pigmentosa (6 per cent.), while Höring found 4 cases among 30 idiots (12.18 per cent.).

In marked contrast to the view that retinitis pigmentosa is frequently found among idiots and the feeble-minded, are the results of Dr. James Thorington's examination of two thousand inmates of the Training Schools for Feeble Minded Children at Elwyn, Pa., and at Vineland, N. J. In not a single one of these cases was retinitis pigmentosa found.

Wider, among his *42 cases of retinitis pigmentosa,* found 12 with defective intellect (28.5 per cent.). Our series contains two mental defectives (11.7 per cent.).

RETINITIS PIGMENTOSA AND CIRRHOSIS OF THE LIVER.

Landolt reported two cases in which cirrhotic disease of the liver and retinitis pigmentosa coexisted. He considered the structural changes in the liver and retina to be analogous. Fuchs describes under the name of *ophthalmia hepatica,* a chorioretinitis, resembling in ophthalmoscopic appearances retinitis pigmentosa, occurring sometimes in cases of chronic disease of the liver associated with jaundice (inflammations or new growths). Night blindness and other symptoms are noted. Such cases are as he states chorioretinitis and should not be mistaken for true retinitis pigmentosa.

PLATE III.

FUNDUS SKETCH, CASE XIV.

This case shows extensive sclerosis of the retina and choroid. The disk is atrophied and has the dull, waxy appearance so often noted.

The vessels have not been uniformly attacked. Some are of fair size, while others are reduced to lines. The corresponding arteries and veins are about equal in size. In two places a vessel is completely ensheathed for a short distance with pigment. The deposition of pigment is also quite irregular, and not extensive.

The case is fifteen years old. Central vision and fields are relatively good.

CHAPTER IV.

ANALYSIS OF CASES.

CASE I.—C. B., male, age nine years, deafness congenital.

Family History.—Both parents born in Germany. Father and mother hear. No consanguinity. Seven children, two deaf (last two).

Personal History.—Walked at one year. No mental weakness; free from fits, scrofula, etc. No infirmity other than deafness. None of the ordinary diseases of childhood.

External Examination.—O_2 lacrimation; irritability; conjunctiva a little thickened; nothing expressed from canaliculi; pupils equal; irides functionally active; ocular movements full in all directions.

Fundus Examination. O.D.—Media clear; disk atrophic, margins blurred, color dull and waxy; faint atrophic excavation; vessels reduced in size, especially on disk where they taper; arteries and veins about equal in size. *Pigment;*—small amount, brown in color, wandering in character situated toward periphery, less below than elsewhere; some is feathery, and some is in lumps; it is not especially near the vessels.

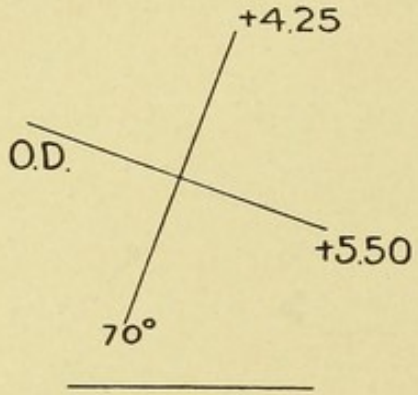
O.S.—Media clear; general condition same as in

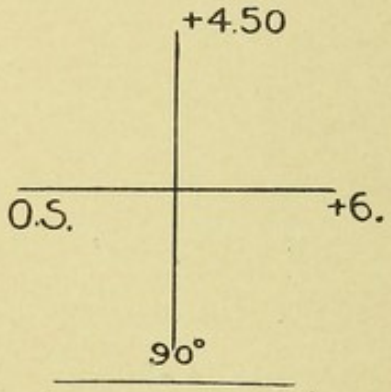
O.D. There is more wandering pigment than in O.D., and the vessels are smaller.

V. O. D.— $\frac{5}{10}$ V. O. S. — $\frac{5}{10}$

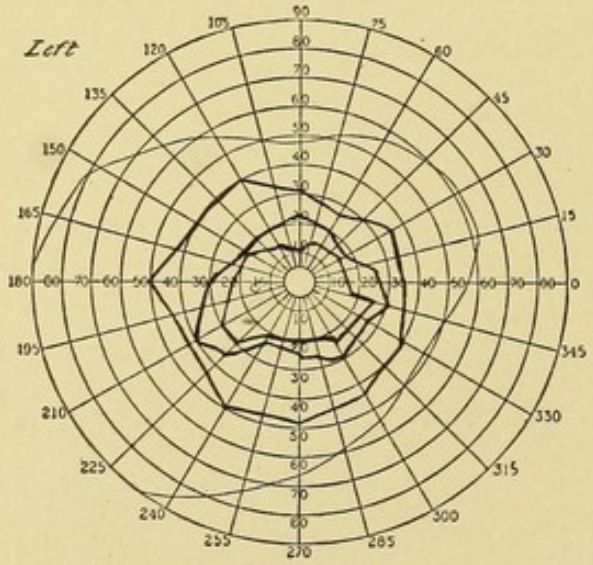
Uncorrected.

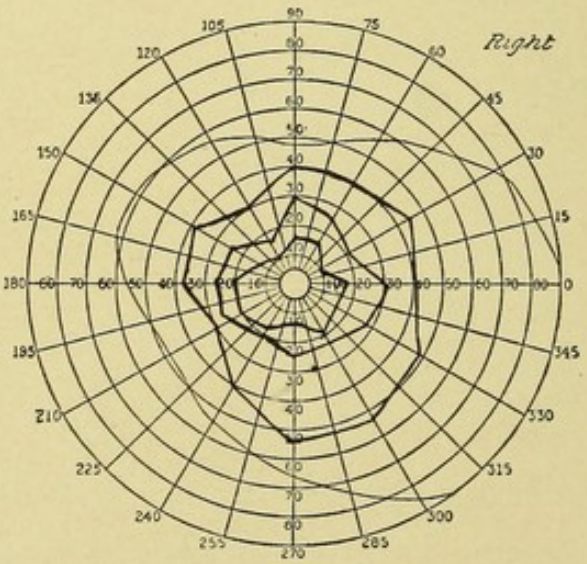
*Refraction.**





Fields.—O_2 Concentrically cut for 10 mm. white, red, and green. Central color perception good; recognizes 2 mm. green.





* The refraction in all cases was determined by retinoscopy under full scopolamin mydriasis.

Case II.—E. M. E., female (sister of III., IV., V.), age thirteen years. Deafness congenital; total.

Family History.—Father and mother born in Pennsylvania, have good hearing, and neither one has deaf relatives; were not related before marriage. They have seven children; numbers 2, 3, 4 and 5 born deaf.

Personal History.—Moral conduct and disposition good. General health good. Free from fits, scrofula, or cutaneous disease. Diseases of childhood:—measles.

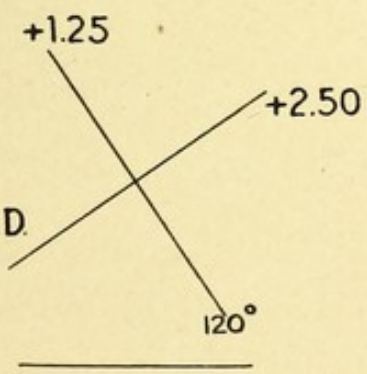
External Examination.—O_2 Conjunctiva clear, nothing expressed from canaliculi; pupils equal, irides functionally active; ocular movements full.

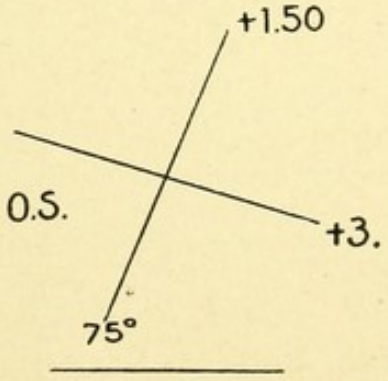
Fundus Examination.—O.D., a few vitreous opacities; rest of media clear. Disk is well defined by connective-tissue ring and markedly atrophic. The vessels are reduced in size, the arteries more so than the veins. Extensive pigment accumulations toward periphery; polar area free. O.S.—changes as in O.D.

V. O. D.—$\frac{5}{7\frac{1}{2}}$ V. O. S.—$\frac{5}{7\frac{1}{2}}$

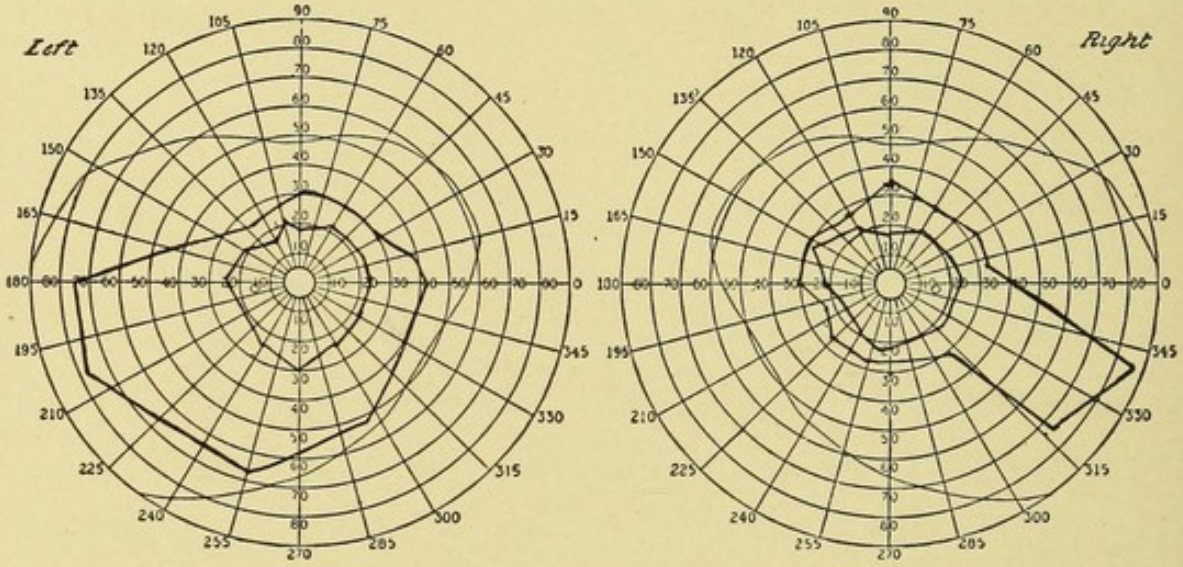
Uncorrected.

Refraction.





Fields.—O.D. concentrically cut for 10 mm. white, except in lower temporal quadrant where for an arc of about 20° it is nearly full. The red shows a full concentric contraction. O.S. concentrically cut for 10 mm. white and red. O_2—Central color perception for 2 mm. object, good.



Case III.—H. E., male (brother of II., IV., V.), age fifteen. Deafness congenital and total.

Family History.—Vide II.

Personal History.—Moral conduct and disposition good. General health good. No history of fits, scrofulous manifestations, or cutaneous disease. Diseases of childhood:—measles.

External Examination.—O_2 Conjunctiva clear, nothing expressed from canaliculi; pupils equal, irides functionally active. Ocular movements full.

Fundus Examination. O.D.—Media hazy (corneal), disk and retina are atrophic; disk margins are

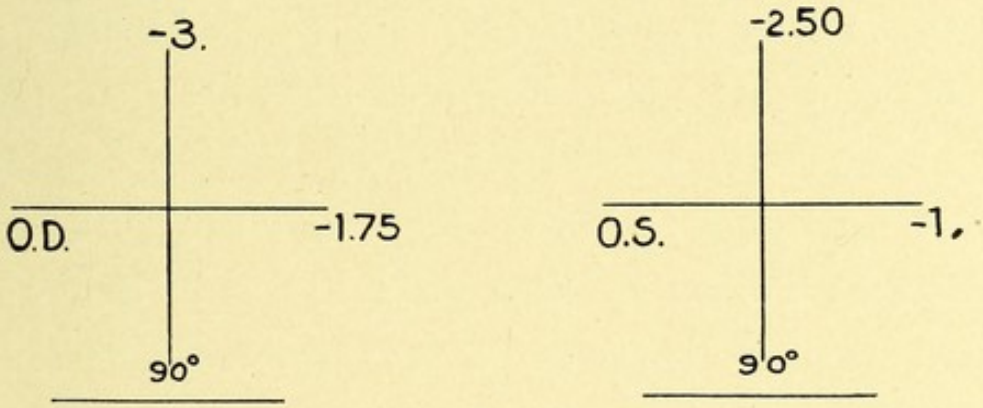
blurred. Wandering pigment in nerve fibre layer, around the periphery; polar portion free. Retinal vessels are much reduced in size.

O.S.—Small corneal opacity down and out; rest of media clear. Disk margins blurred; disk and retina atrophic; vessels much reduced. Wandering pigment as in O.D. around periphery, polar portion of fundus free.

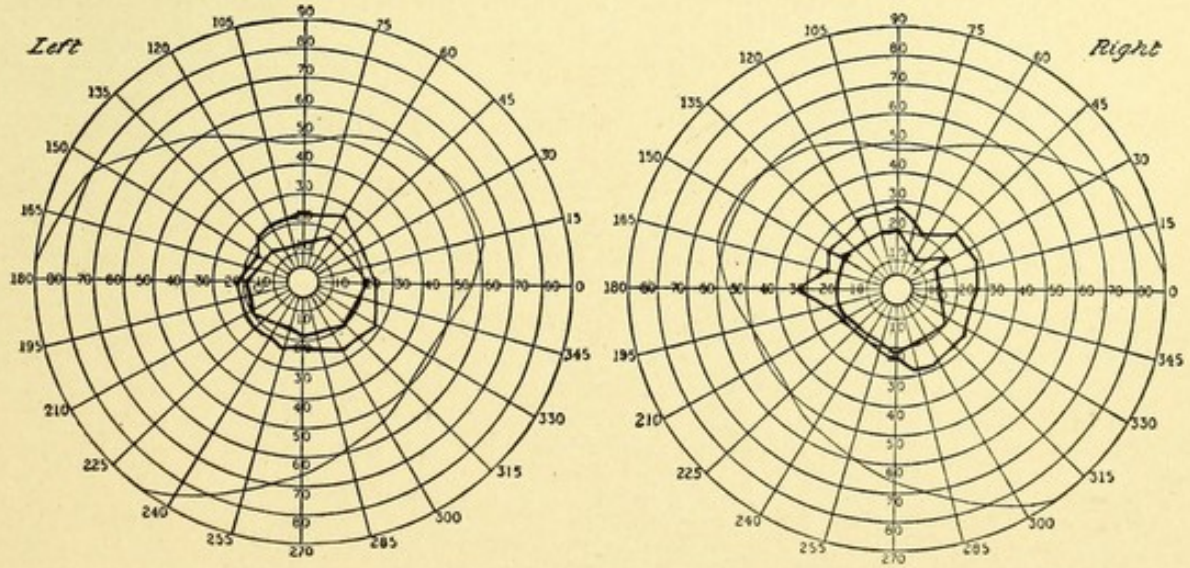
V. O. D.— $\frac{5}{9}$ V. O. S.— $\frac{5}{7\frac{1}{2}}$

Corrected.

Refraction.



Fields.—O_2 Very marked concentric contraction for 10 mm. white and red, the two fields almost coinciding. Central color perception for 2 mm. object is good.



Case IV.—N. E. E., female (sister of II., III., V.), age fourteen. Deafness congenital and total.

Family History.—Vide II.

Personal History.—Moral conduct and disposition good. General health good; no history of fits, scrofula, or cutaneous disease. Has had measles.

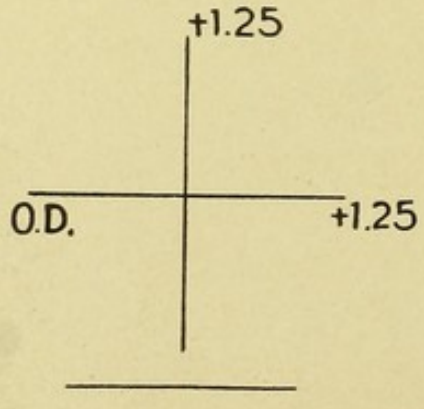
External Examination.—O_2 Conjunctiva clear, no lacrimal disturbance. Pupils equal, irides functionally active. Ocular movements full.

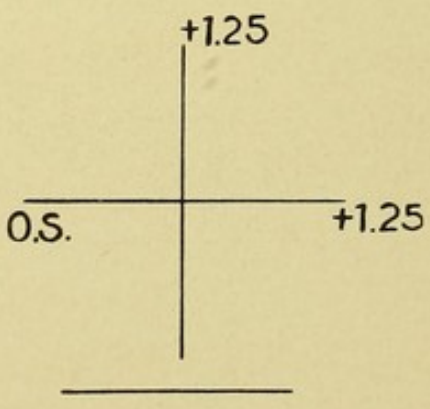
Fundus Examination.—O_2 Media slightly hazy; disk and retina markedly atrophic; disk blurred, dirty in color and opaque, surrounded by a hazy connective-tissue ring. The retinal vessels are greatly reduced in size. Pigment is rather sparce, fine and feathery; it is situated at periphery and does not extend very far toward centre. There are a few white spots near pigment accumulations. In the fundus of O.D., mostly on the disk, is considerable cholesterin.

V. O. D.—$\frac{5}{12}$ V. O. S.—$\frac{5}{7\frac{1}{2}}$

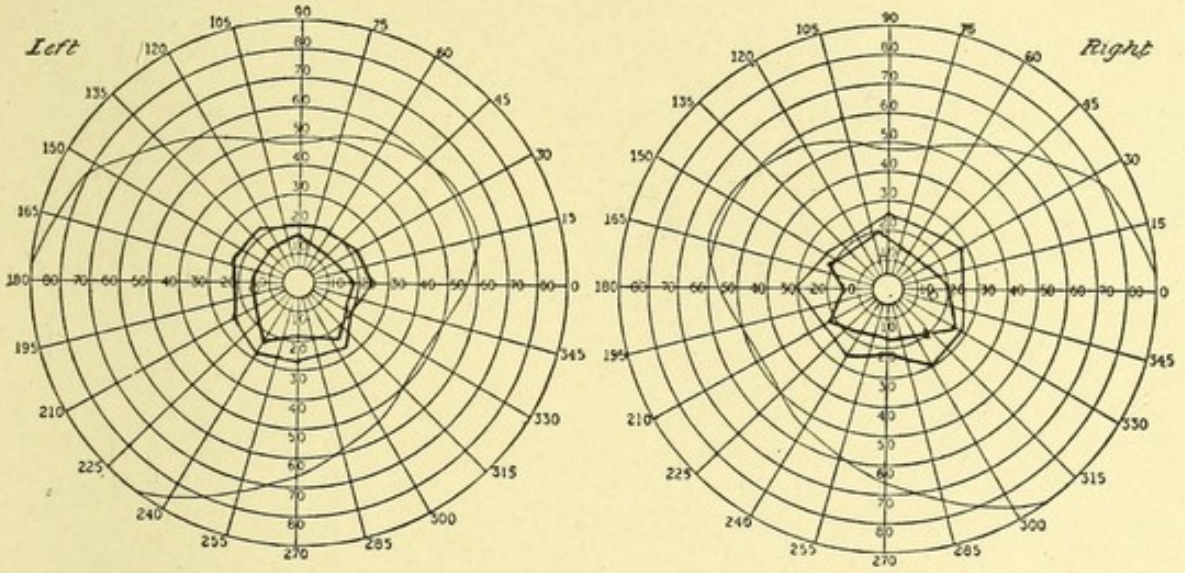
With and without correction.

Refraction.





Fields.—O_2 Concentric contraction to about 25° for 10 mm. white and red. Central color perception for 2 mm. object, good.



Case V.—M. J. E., female (sister of II., III., IV.), age eighteen years. Deafness congenital and total.

Family History.—Vide II.

Personal History.—Moral conduct and disposition good. General health good; no history of fits, scrofula or cutaneous disease; none of the diseases of childhood.

External Examination.—O_2 Conjunctiva clear, no lacrimal disturbance. Pupils equal, irides functionally active. Ocular movements full in all directions.

Fundus Examination.—O.D. Media clear, disk poorly defined by a connective-tissue ring; margins blurred; disk and retina are atrophic. Vessels are greatly reduced in size. Pigment dense and thickly

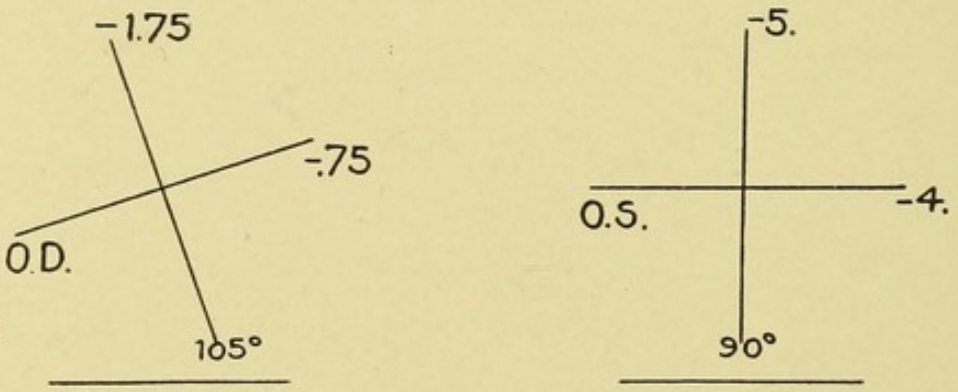
accumulated, occupying periphery and extending rather far toward the centre.

O.S.—Picture same as in O.D. except as regards the entrance of the vessels upon the disk; they emerge from the nasal side into a large, deep physiological excavation. In O_2 the temporal fundus is more free from pigment.

V. O. D.— $\frac{5}{15}$ V. O. S.— $\frac{5}{12}$

Without correction.

Refraction.



Fields.—Not obtained.

Case VI.—K. E., age eighteen and a half years. Deafness congenital and total.

Family History.—Father and mother born in Pennsylvania; not related; no deafness in either branch of family. Three children, patient being first and the only one deaf.

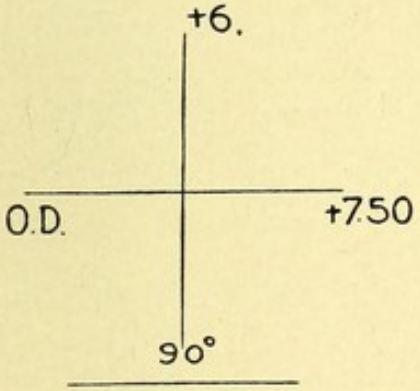
Personal History.—Moral conduct and disposition good; general health good; is free from disease. Whooping-cough in childhood. Night blindness pronounced; strikes objects in walking, very often. Walked at two and one-half years.

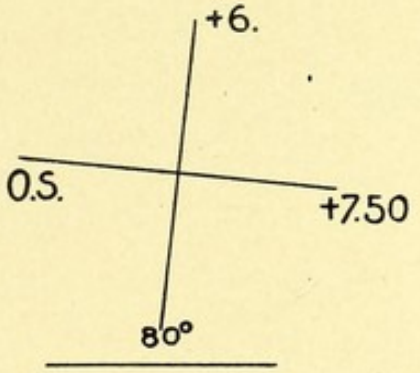
External Examination.—O_2 Conjunctiva clear, no lacrimal disturbance; pupils rather small; irides functionally active. Ocular movements full.

Fundus Examination.—O_2 Media clear, disks and retina atrophic; vessels small; extensive pigmentation around periphery; pigment is of the bone corpuscle variety.

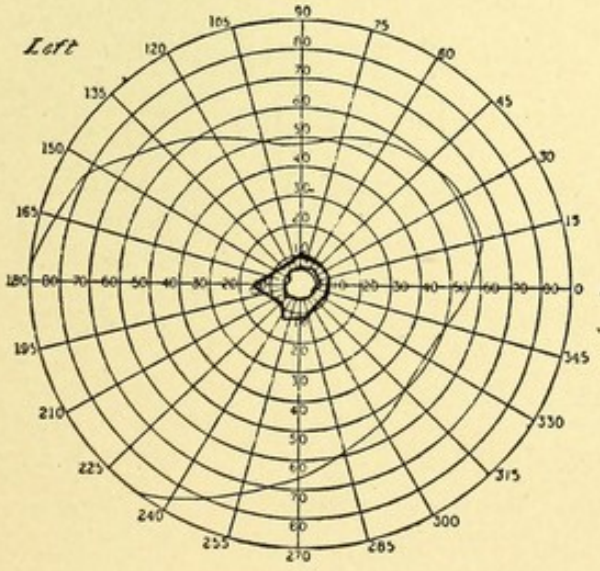
Vision.—Not recorded.

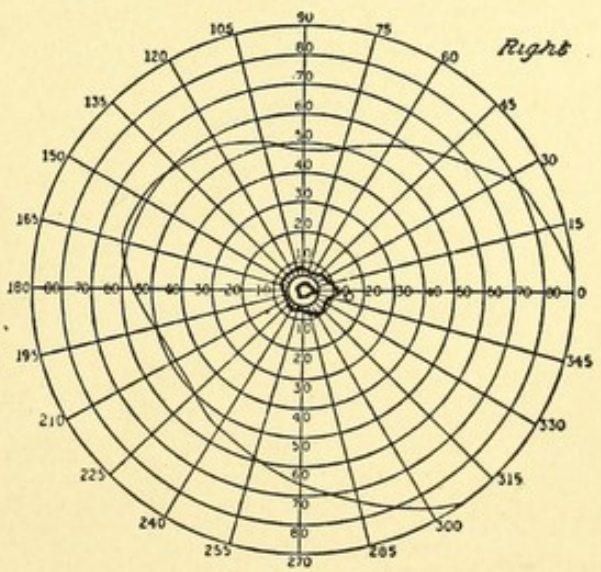
Refraction.





Fields.—O_2 Concentrically reduced for 10 mm. white and red; almost entire field is within 10°. Central color perception for 2 mm. object good.





Case VII.—E. G. C. F., male, age eighteen years. Deafness congenital and total.

Family History.—Parents second cousins; father born in Ireland. Mother born in England. Eleven children born, respectively, in 1874, '75, '77, '79, '80, '82, '84, '86, '88, '91, '94. Those born in '82, '88, and '91, died at 14 days, 18 months, and 11 months, respectively. Those born in '74 and '84 are deaf, '84 being the present case.

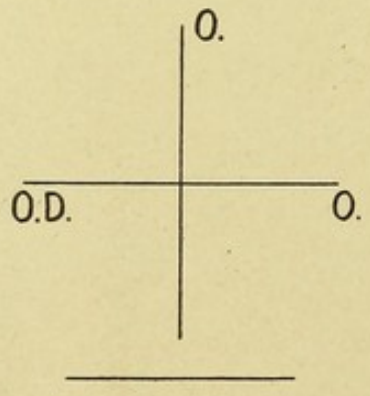
Personal History.—Seems to be mentally deficient; impossible to get satisfactory results from tests; has had typhoid fever and measles.

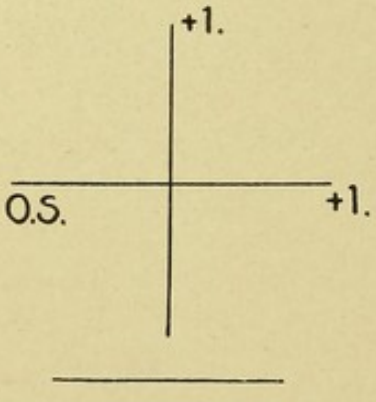
External Examination.—Conjunctiva clear; nothing expressed from canaliculi. Pupils equal, irides functionally active. Ocular movements full in all directions.

Fundus Examination.—O.D. Media clear; disk margins blurred; and retina atrophied. Vessels are small; general pigment absorption with wandering into retina mostly at equator. O. S.—Fine opacities in anterior vitreous. Fundus otherwise as in O.D.

Vision.—Not recorded.

Refraction.





Fields.—Impossible to record.

Case VIII.—E. V. F., female, age eight years. Deafness congenital and total.

Family History.—Father and mother born in Pennsylvania; not related; no deaf relatives. Five children, first dead at two and a half years. Patient is fourth and is the only one deaf.

Personal History.—Moral conduct and disposition good; no mental deficiency; no infirmity or disease other than deafness and retinitis pigmentosa; has had mumps and whooping-cough. Walked at three years.

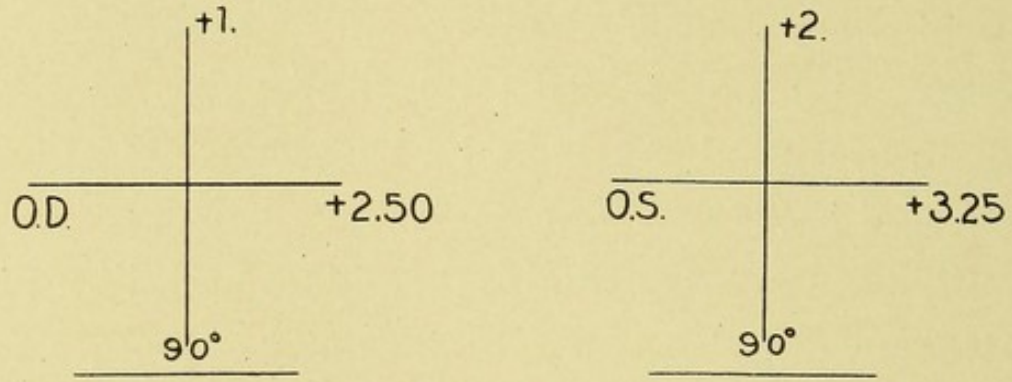
External Examination.—O_2 Conjunctiva clear, no lacrimal disturbance; pupils are equal; irides are functionally active. Ocular movements are full in all directions.

Fundus Examination. O.D.—Media clear; disk is flat and atrophied; parchment color; margins everywhere blurred and obscured. Retinal vessels are a little reduced in size. Retina is atrophied, there is pigment absorption throughout; the chorioidal vessels are everywhere visible. Toward the periphery are numerous white spots. At the nasal periphery below are small pigment spots in the nerve-fibre layer of the retina. O.S.—Changes essentially the same as in O.D., except that the retinal vessels are greatly reduced in size. There are white spots toward the periphery and small pigment spots in the nerve-fibre layer peripherally situated.

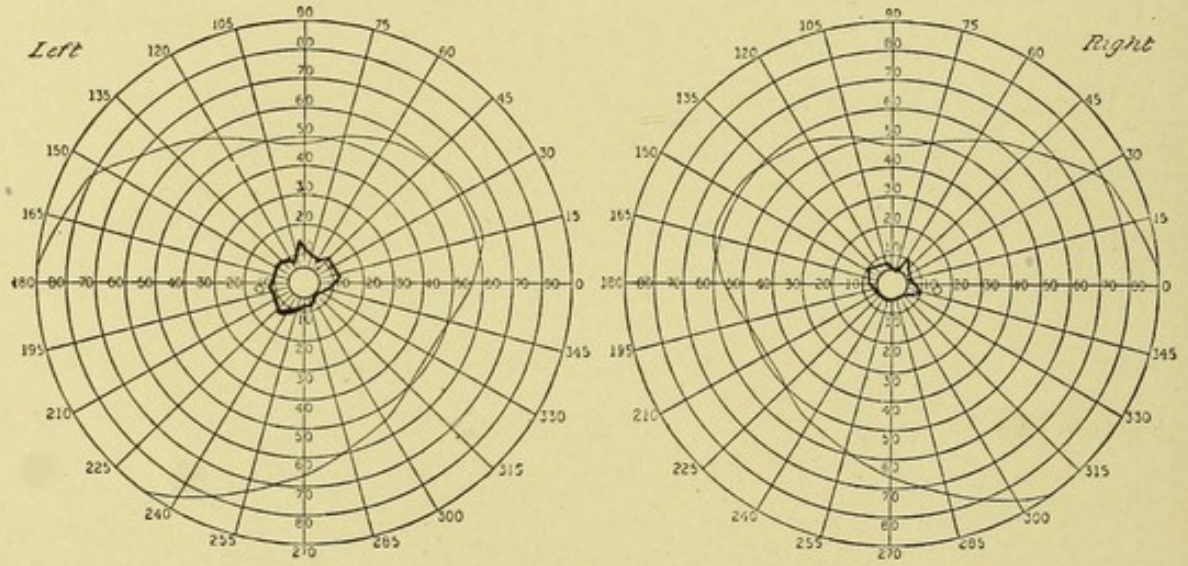
V. O. D.— $\frac{5}{10}$ V. O. S.— $\frac{5}{20}$

Not corrected.

Refraction.



Fields.—O_2. Reduced for 10 mm. white, concentrically to 8°–10°. No color fields obtainable.



Case IX.—N. G., female, age sixteen years. Partially deaf from birth.

Family History.—Parents American. Mother not deaf; no note of father. Consanguinity—not known.

Personal History.—Probably illegitimate. Moral conduct and disposition questionable. Some signs of

mental deficiency; general health is good; free from fits, scrofula, or cutaneous disease.

External Examination.—O_2 Conjunctiva clear, no lacrimal disorder; pupils equal; irides functionally active. Ocular movements full in all directions. Anterior chambers are deep.

Fundus Examination.—O. D. Media clear, disk is atrophied; surrounded by well marked connective-tissue ring broadest out; pigment mass at temporal margin; eccentric excavation out. The retinal vessels are reduced in size. The retina is atrophied; the chorioidal vessels are everywhere visible.

Pigment.—Round pigment spots are scattered throughout the fundus; some are situated in the nerve-fibre layer and some are deeper. There are no feathery or bone corpuscle-like formations.

O.S.—The changes are essentially the same as in O.D. Pigmentation extends very far toward periphery, both eyes.

V. O. D.— $\frac{5}{7\frac{1}{2}}$ V. O. S.— $\frac{5}{7\frac{1}{2}}$

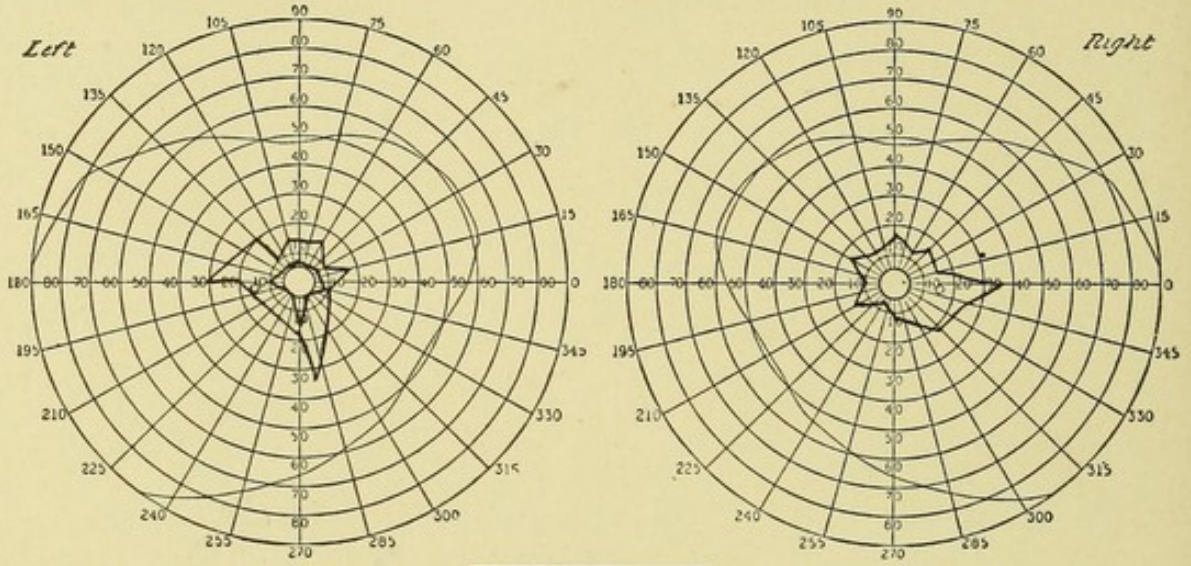
With and without correction.

Refraction.—Not recorded.

Fields. O.D.—Much cut for 10 mm. white and red. Outline angular and irregular. Central color perception good for 2 mm. object.

O.S.—Field for 10 mm. white, very much cut. Outline angular and irregular. Field for 10 mm. red,

reduced to fixation. Central color perception good for 5 mm. object, not for 2½ mm.



Case X.—M. K., female, age nineteen years. Deafness total and congenital, or resulted from a fall at four weeks.

Family History.—Parents born in Pennsylvania; not related; no deaf relatives. Three children born in 1878, '84, and '85. The second only is deaf.

Personal History.—Moral conduct and disposition good. Has had whooping-cough.

External Examination.—O_2 Conjunctiva injected. O.S.—Lymphatic swelling in lower fornix; no lacrimal disturbance; pupils are equal; irides are functionally active; ocular movements are full in all directions.

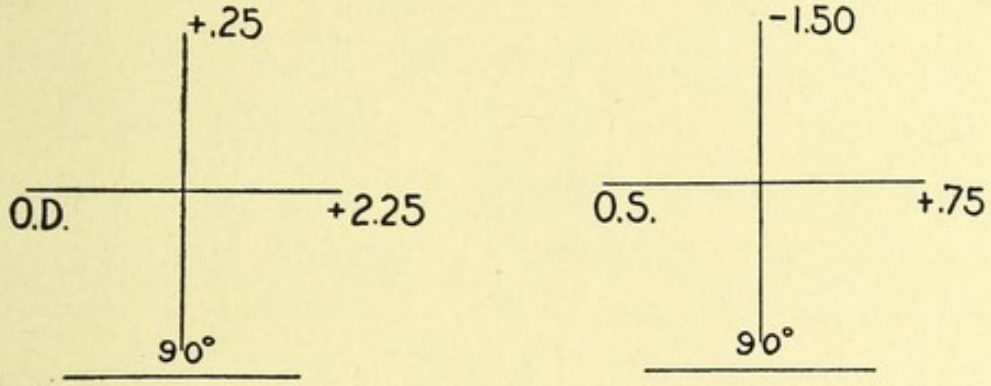
Fundus Examination.—O_2 Media clear, disks and retinæ are atrophied; the retinal vessels are small. A great deal of pigment is found in the retina toward the periphery, and extending rather far down. The pig-

ment is in *rather large dense masses* well interwoven.

V. O. D.— $\frac{5}{10}$ V. O. S.— $\frac{5}{10}$

Corrected.

Refraction.



Fields.—Not recorded.

CASE XI.—K. L. M., female, age eleven years. Deafness congenital and total (appreciates loud thunder and heavy vibrations).

Family History.—Father partially deaf left ear from scarlet fever at six weeks; mother not deaf. Parents are second cousins; there are no deaf relatives in either branch of family. They have had eight children, five of whom are living; two were born deaf.

Personal History.—(Is fifth child, fourth and fifth deaf; first three dead.) Moral conduct and disposition are good. No mental weakness shown. General health is good. Walked at two years. Cataract removed from O.S. some time ago; details of operation not known. No history of fits, scrofula or cutaneous disease. Has had scarlet fever, measles and whooping-cough. Complains much of night blindness.

External Examination.—O_2 Conjunctiva clear, no lacrimal disturbance.

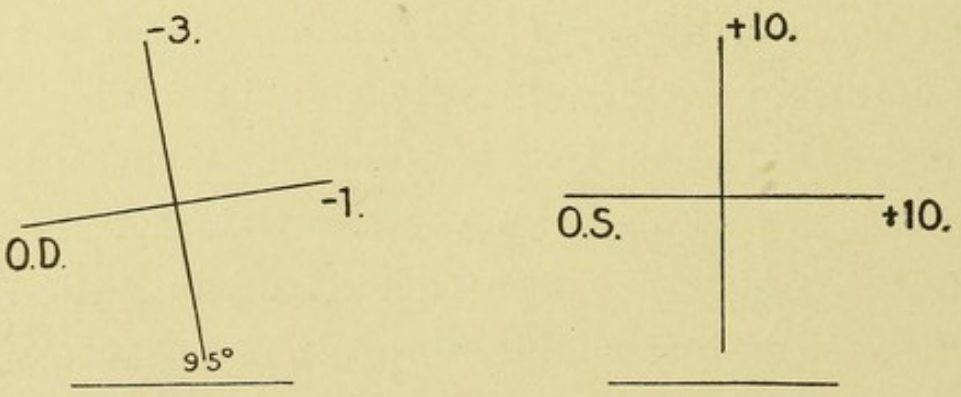
Fundus Examination. O.D.—Lens cataractous in posterior cortex; patch of opacity to temporal side; rest of media clear. Disk is well defined, waxy in color and appearance; very atrophic. The retinal vessels are much reduced in size; arteries and veins are of equal calibre. The retina is markedly sclerosed; there are a good many distributed spots or lesions, but no pigment is found.*

O.S.—Aphakic; iris is tremulous; there are some dense capsular remains peripherally, but the centre is clear. The disk is well defined, waxy in appearance and atrophied. The vessels, the retinal sclerosis and the retinal lesions are as in O.D.; there are no pigment foci noted.

V.O.D.—(Patient would not respond to tests.)

V.O.S.—Apparently blind; cannot fix; says V. is impossible.

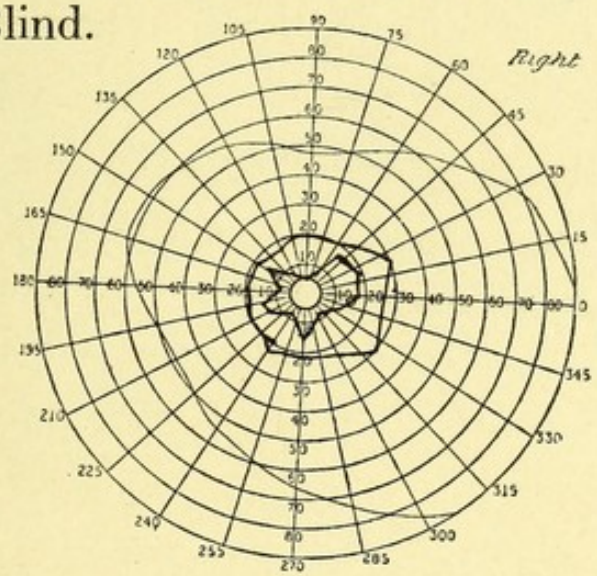
Refraction.



* See footnote on page 9.

Fields. O.D.—Concentric contraction for 10 mm. white, to about 20°, red field for 10 mm. 5°–10° within form. Green is not recognized except in 20 mm. object; smaller green objects are called blue. Other colors are centrally perceived in 2 mm. objects.

O.S.—Blind.



Case XII.—M. J. M., female (sister of XI), age fourteen years. Born deaf; deafness is total for ordinary sounds, but is conscious of sharp vibrations and very loud noises.

Family History.—Vide XI.

Personal History.—Moral conduct and disposition are good. No evidences of mental weakness or deficiency. Walked alone at three years. General health is good; no history of fits, scrofula or cutaneous disease. Has had measles, mumps and whooping-cough.

External Examination.—O_2 Conjunctiva clear; no lacrimal disorder. Pupils are equal, irides functionally active. Ocular movements are full in all directions.

Fundus Examination.—O.D. Few cobweb vitreous opacities; disk is well defined by a connective-tissue ring; it is atrophied and white in color, differing from the usual dirty, waxy looking disk. The retinal vessels are small, the arteries and veins being about equal in size. The retina is atrophied; there are white spots and placques throughout, with some little wandering pigment above, in the nerve-fibre layer.

O.S.—There are a few cobweb opacities in the vitreous, and a white flocculent opacity anchored by a thread. The disk, retina and vessels are as in O.D., excepting that the small amount of wandering pigment is in this eye, below.

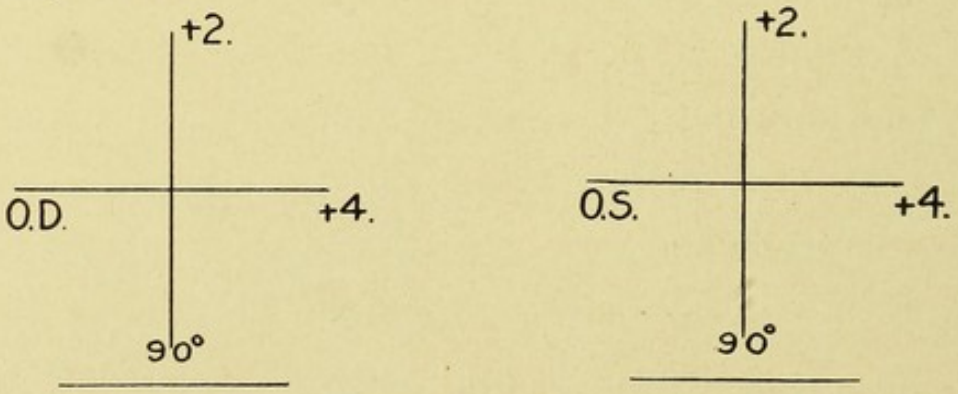
V. O. D.— $\frac{5}{9}$ V. O. S. — $\frac{5}{12}$

Not corrected.

V. O. D.— $\frac{5}{6}$ V. O. S.— $\frac{5}{6}$??

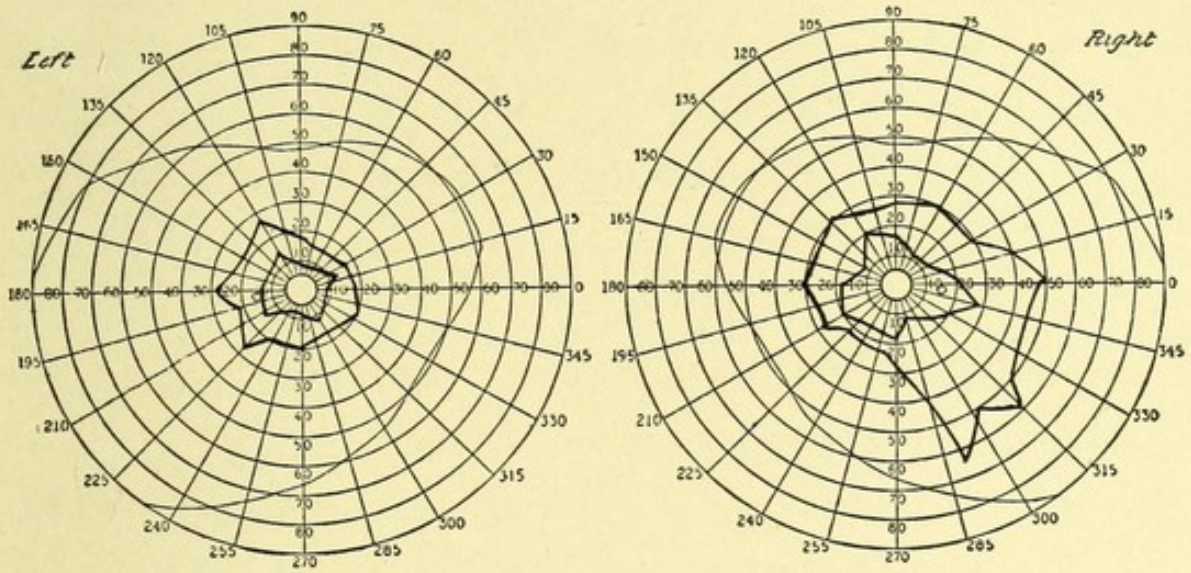
Corrected.

Refraction.



Fields. O.D.—Moderate contraction for 10 mm. white and red, boundary line for white rather irregular. Central color perception for 2 mm. good.

O.S.—Concentric contraction for 10 mm. white and red, more marked than in O.D. Central color perception for 2 mm. object good.



CASE XIII.—P. M., male, age eleven years. Hearing lost at thirteen and a half months following chicken-pox. Is partially deaf.

Family History.—Parents American; no deafness; no consanguinity. Number of children—not stated.

Personal History.—Walked at one year. Moral conduct and disposition good. No infirmity other than deafness and retinitis pigmentosa. Has had scarlet fever and chicken-pox.

External Examination.—O_2 Conjunctiva clear; no lacrimal disturbance; pupils equal; irides functionally active. Ocular movements full.

Fundus Examination. O.D.—Media clear; disk is atrophied, parchment-colored, and not sharply de-

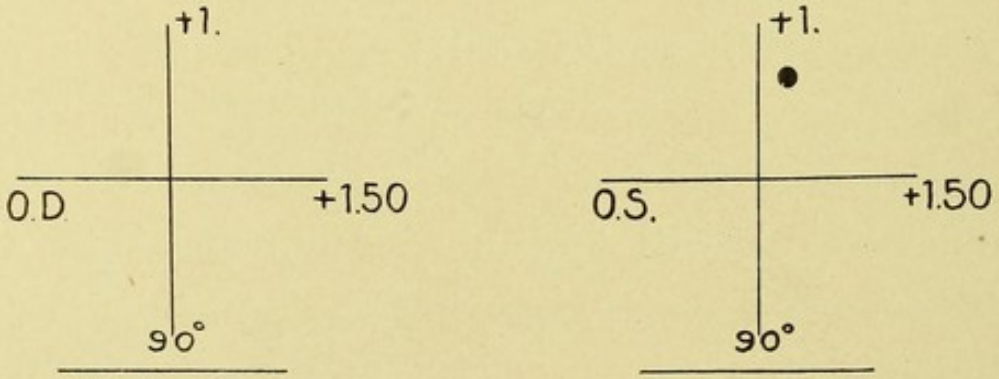
fined. The retinal vessels are much reduced in size and are irregular; the arteries show more reduction than the veins. A remnant of the hyaloid artery, white in color, is attached to centre of disk, and has its free end far forward in the vitreous. The retina is sclerosed throughout. Pigment extensive in its distribution; much of it is arranged along the vessels in chains. There are also many rounded isolated splotches as well as a few bone corpuscle-like formations. The retinal degeneration and the pigmentation are most marked around the periphery; the macular region is free.

O.S.—Conditions essentially the same as in O.D. The remains of the hyaloid are more rudimentary.

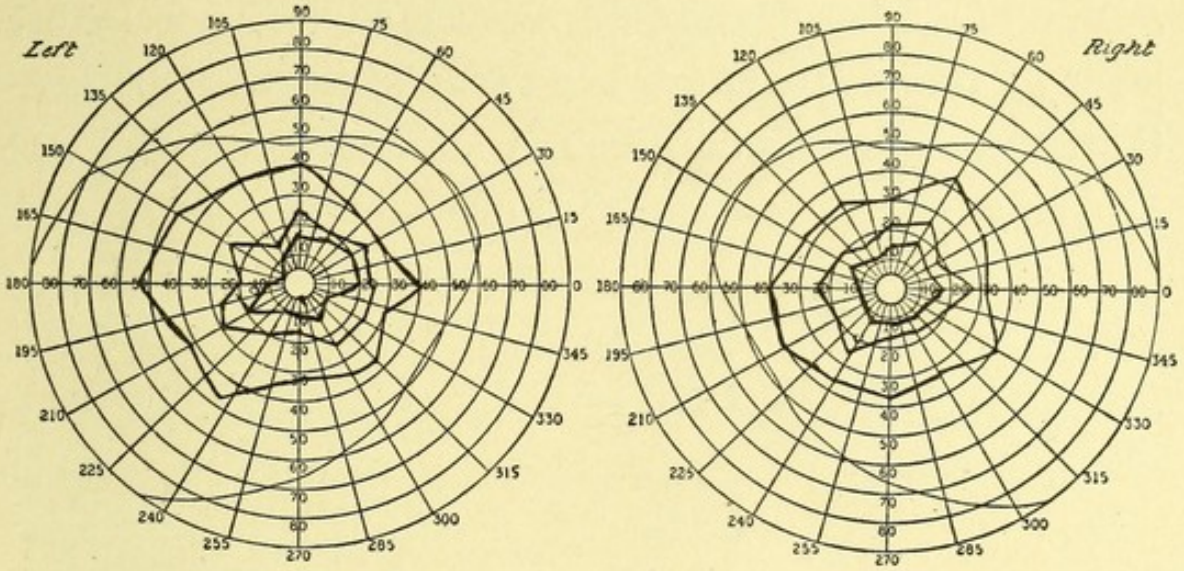
V. O. D. $-\frac{5}{6}$ V. O. S. $-\frac{5}{10}$

Not corrected.

Refraction.



Fields.—O_2 Concentric contraction for 10 mm. white, red and green. Central color perception for 2 mm. object is good. The fields show less contraction than would be expected from the extensive sclerosis of the retina.



CASE XIV.—H. S., female, age fifteen years. Deafness is total and congenital.

Family History.—Parents born in Ireland. Father's name is James Sullivan, mother's maiden name was Mary Sullivan, but consanguinity is denied. Have had three children; H. is first child and only one deaf. (?) Youngest child died at three weeks; no deaf relatives.

Personal History.—Moral conduct and disposition good; general health good; no history of fits, scrofula or chronic cutaneous disease; has had scarlet fever and measles.

External Examination.—O_2 Blepharitis marginalis; palpebral and bulbar conjunctiva injected; no lacrimal disturbance; pupils equal; irides functionally active; ocular movements full in all directions.

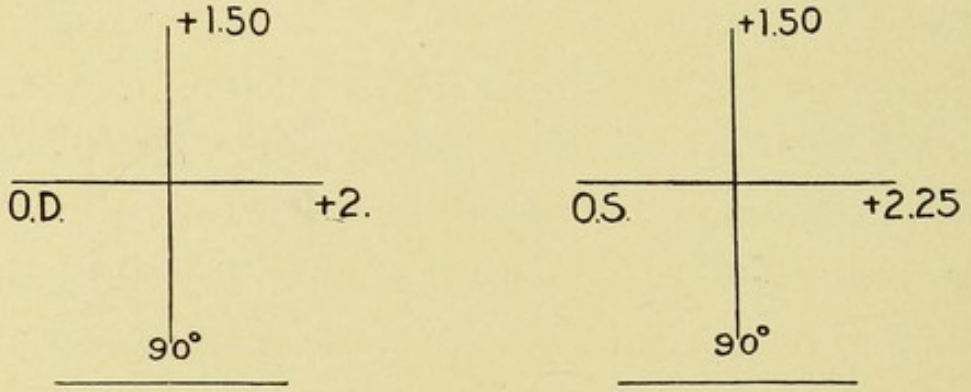
Fundus Examination.—O_2 Media clear. Disk is atrophied, waxy in color, and has blurred margins; the arteries and veins are quite small, and are of equal size.

Around periphery are a few pigment spots in nerve-fibre layer of retina; some are bone corpuscle-like in formation; there are also numerous whitish areas, which will probably later receive pigment deposits.

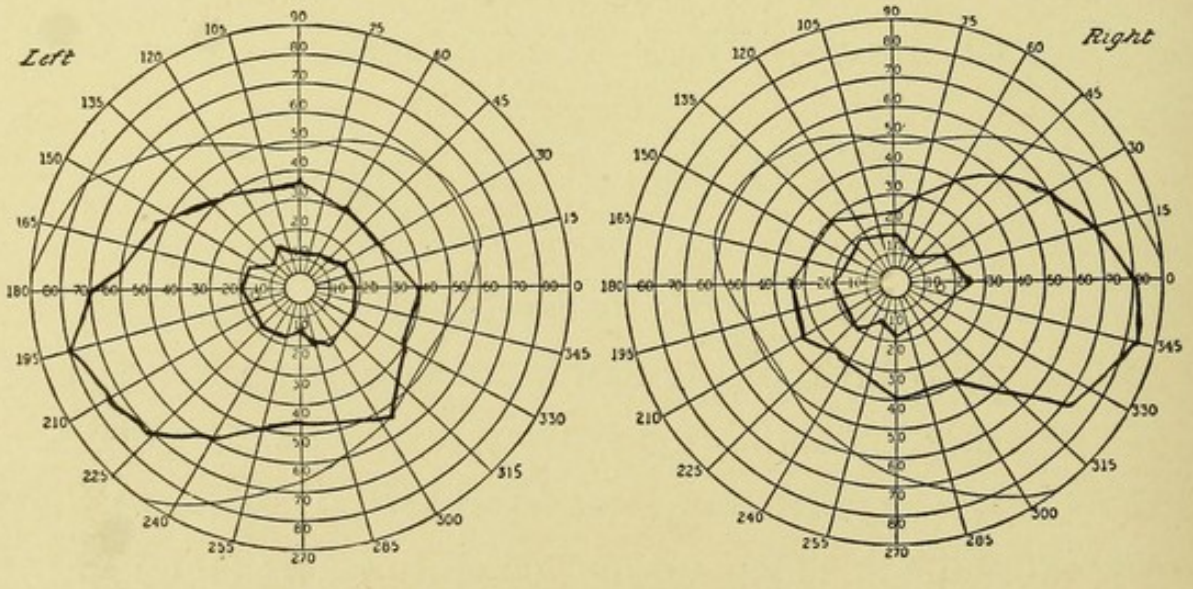
V. O. D.— $\frac{5}{9}$ V. O. S.— $\frac{5}{9}$

Not corrected.

Refraction.



Fields.—O_2 Fields are cut concentrically for 10 mm. white and red, about 20°. Central color perception for 2 mm. object is good.



CASE XV.—C. M. W., female, age ten years. Became totally deaf at four months, following convulsions.

Family History.—Parents born in Pennsylvania. Father and mother not deaf; no deaf relatives; no consanguinity; is only child.

Personal History.—Moral conduct and disposition good; general health good; convulsions in infancy; no subsequent history of fits, scrofula, or cutaneous disease; usual diseases of childhood negative.

External Examination.—O_2 Conjunctiva pale; no lacrimal disturbance. O.D.—*Vertical* nystagmus; the complete vertical movements are two or three per second and cease in downward rotation.

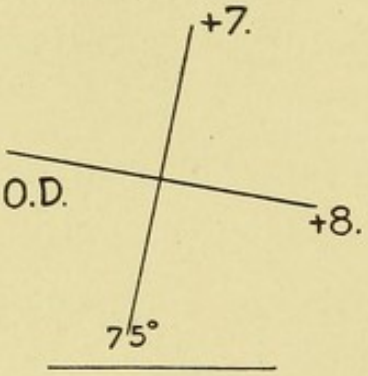
O.S.—Nystagmus is *rotary,* not so rapid as in O.D., and movements are of small amplitude.

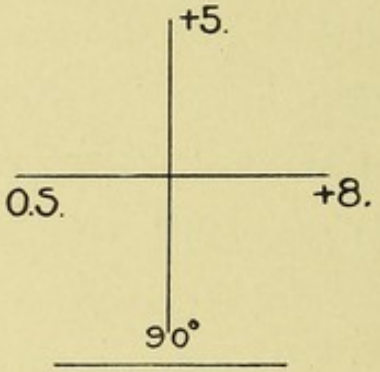
Fundus Examination. O.D.—Vitreous opacities; disk is atrophied, margins blurred; the retina is atrophied. The retinal vessels are very much reduced in size. There is a great deal of pigment wandering in retina; it bears no particular relation to the vessels, and is not lace-like nor bone corpuscle-like in arrangement, but takes the form of long lines and round deposits; no covering of the vessels is noted.

O.S.—Large vitreous opacities; disk is atrophied and well defined. The retinal vessels are much reduced in size; there is less pigment deposition, and less chorioido-retinal disturbance than in O.D. There are some few pigment lines and round deposits.

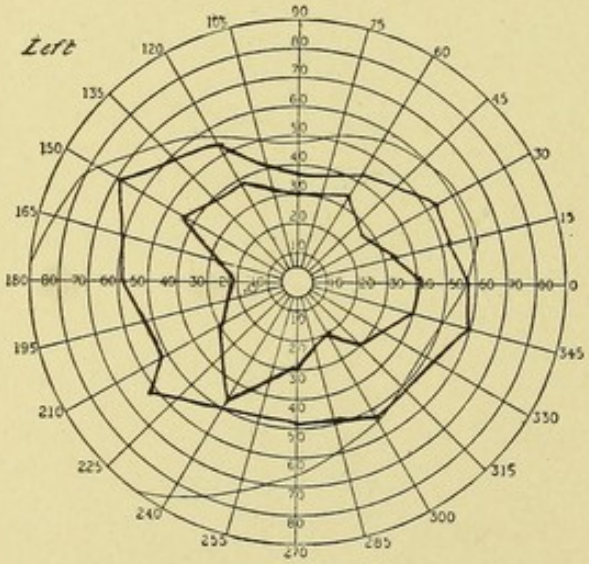
V. O. D.— $\frac{5}{15}$ V. O. S.— $\frac{5}{10}$??

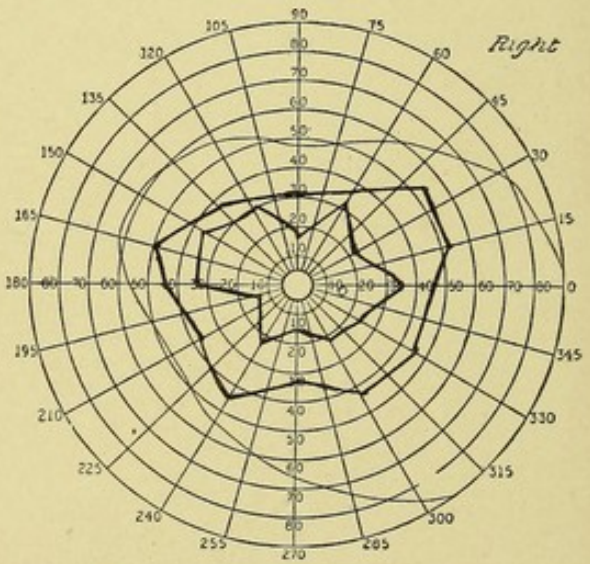
Refraction.





Fields.—O_2 Fields for 10 mm. white and red are moderately cut, mostly to temporal side, and relatively more for form than color. The field for O.D. is a little smaller than that for O.S. Central color perception is good.





Case XVI.—C. W., female, age nine years. Deafness congenital and total.

Family History.—Parents American; no con-

sanguinity; no deafness. Have five children, C. being number 2; all are living, and none are deaf but C.

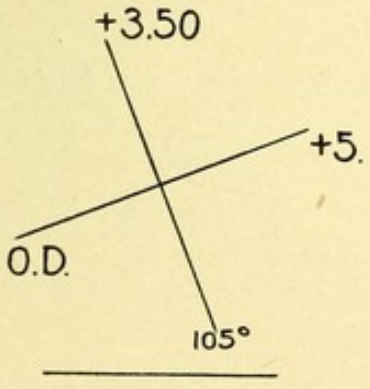
Personal History.—Understanding and intelligence are good; is free from signs of constitutional disease.

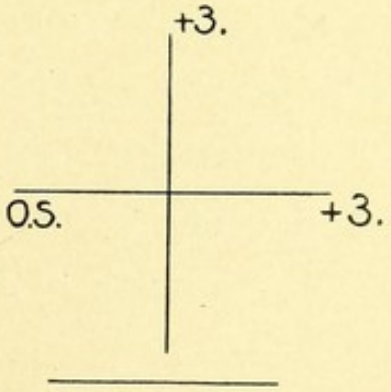
External Examination.—O_2 Conjunctiva pale and anemic looking; no lacrimal disturbance; pupils are equal; irides functionally active. Ocular movements are full.

Fundus Examination.—O_2. Media clear; disks and retinæ are atrophic; disks are not sharply defined. The retinal vessels are a little reduced in size. There are a number of white spots, more pronounced in O.D., and some pigment spots, not feathery, and more numerous in O.S.

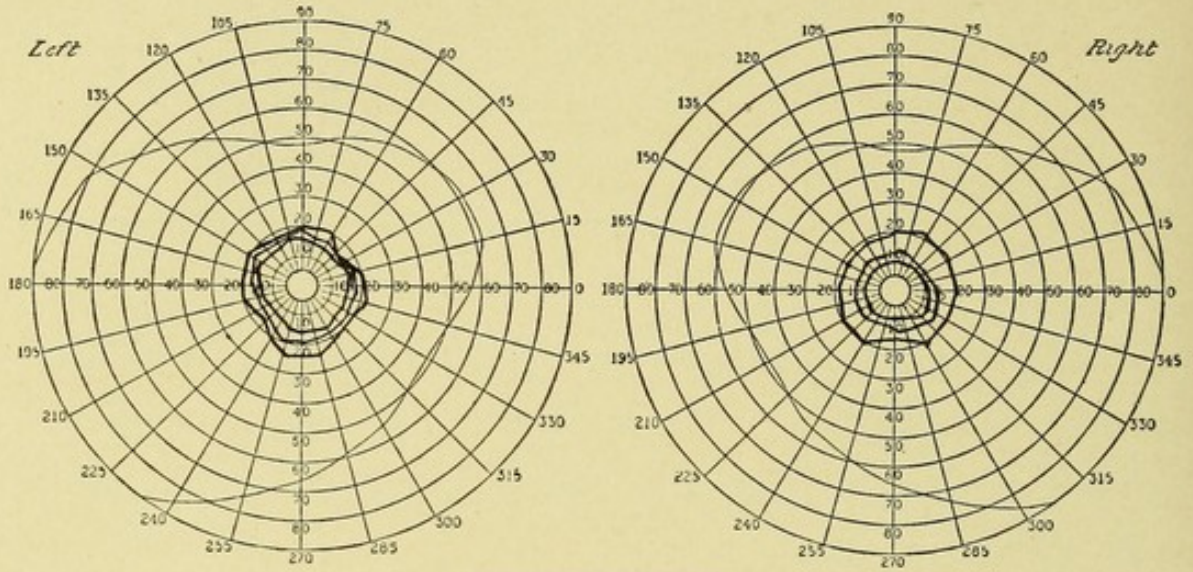
V. O. D.—$\frac{5}{15}$ V. O. S.—$\frac{5}{15}$

Refraction.





Fields.—O_2 Fields show great concentric contraction for 10 mm. White, red and green. Central color perception good.



Case XVII.—E. Y., female, age eighteen years. Deafness total; noticed at one year, following "cold."

Family History.—Parents born in Pennsylvania; not related; no deafness in either branch of family; two children; first child normal.

Personal History.—Moral conduct and disposition good. Pneumonia at three months; no signs of scrofula or chronic cutaneous disease; has had pneumonia, scarlet fever and measles.

External Examination.—Facial asymmetry; O.D. nearer median line; palpebral fissure O.D. is narrower than that of O.S.; O_2 conjunctiva clear; pupils equal; irides functionally active; ocular movements are full, convergence good.

Fundus Examination. O.D.—Media clear; the disk is very opaque and atrophic—"parchment-like;" margins are blurred. The arteries are a little reduced in size. There is typical degeneration of the retina,

extending and increasing as far toward the periphery as can be seen. The pigment is not uniform; there are a number of black disks and balls of pigment, and a few irregular splotches; the macular region is clear.

O.S.—The arteries are more reduced than in O.D., and irregular, dense pigment masses are more numerous. Other conditions as in O.D.

V. O. D.— $\frac{5}{7\frac{1}{2}}$ V. O. S.— $\frac{5}{5}$

Refraction.—O_2 Emmetropia.

Fields.—Not recorded.

CHAPTER V.

SUMMARY OF CASES.

The more important data noted in the preceding case histories may be briefly summarized as follows:

Sex.—It will be seen that *13 of the cases were females and but 4 were males.* This is a marked contrast to all other statistics relative to *retinitis pigmentosa and sex.* According to Leber, three-quarters of all cases of retinitis pigmentosa are males; Derigs—70 per cent. males, 30 per cent. females; Wider—66.6 per cent. males, 33.3 per cent. females. Sixteen of our cases are from the Pennsylvania Institution for the Deaf and Dumb, and one from the School for Deaf Children, at Bala.

The annual reports from the former institution, for the last six years, show that there have always been during this time, more boys in attendance than girls, the average attendance being 503 children, of whom 267 were males and 236 females.

These figures accord accurately with the combined statistics as given by Mygind, from all parts of the civilized world, viz., 210,190 deaf-mutes,—110,879 males, 91,682 females, or 83 females to 100 males.

From this, the conclusion would seem warranted that *among deaf-mutes, retinitis pigmentosa is much*

more frequent in the female, almost reversing the sex ratio for retinitis pigmentosa in general.

Age.—The ages of our cases at the time of examination range from 8 to 19 years. The essential changes throughout are as pronounced in the younger subjects as in the older. No "commencing cases" were recognized in the ophthalmoscopic examination of between 600 and 700 deaf-mutes from the one institution, where the age of entrance is six years, and of about 100 from the other institution, where the entrance age is two years.

We are, from our investigations, unable to say when first the subject would have been recognized as having retinitis pigmentosa, but believe the disease, if sought for, would have been detected at, or very shortly after birth. The well known slow progress of the disease, the comparatively little change for the worse noted between the younger and the older subjects (an interval of 10 years) and the advanced stage of degeneration found in the younger subjects, would seem to make this probable.

Heredity.—The parents of our cases number 26, with a total offspring, at the time of record, of 55. There is no evidence of direct transmission of either retinitis pigmentosa or of deafness, nor do our records show constitutional disease of any kind in any of the parents. Personal observation was, however, impossible, so that the elimination of such cannot be vouched

for. Of the 55 children, 19 were deaf, from which, of course, our 17 cases of retinitis pigmentosa come. But, of the remaining 36, two died within one month of birth, one at 11 months, one at 18 months and three more probably in infancy, so that it is impossible to say what the condition might have been in these cases.

The order of the defective children in their respective families, is interesting, and while perhaps not weakening the theory of heredity, would suggest, in certain cases, the operation of other determining causes, among them, maternal impression. In speaking of defective children in this connection, we refer mostly to deafness, and assume that where coupled with retinitis pigmentosa, the two manifestations are of common origin.

TABLE.

Cases.	Number in Family.	Defective.
I,	7	6th and 7th.
II, III, IV, V,	5	2nd, 3rd, 4th, and 5th.
VI,	3	1st.
VII,	11	1st and 7th (3 dead, 6–9–10).
VIII,	5	4th.
IX,	1	1 (illegitimate).
X,	3	2nd.
XI, XII,	8	4th and 5th (1st and 3rd dead).
XIII,	1	1.
XIV,	3	1st (1 dead).
XV,	1	1.
XVI,	5	2nd.
XVII,	2	2nd.

Consanguinity.—Consanguineous marriage of parents is noted in two instances, the relationship in each

case being that of second cousin. The total offspring of these two marriages was nineteen, four at least of whom were deaf. Of these four, case VII and cases XI and XII (sisters) are of our series.

Thus we have represented in this series 13 marriages, 2 of which, or *15.3 per cent., consanguineous.*

THE GENERAL CONSTITUTION AND STATUS OF THE PATIENTS THEMSELVES.

At the time of entrance upon institution life, and at the time of examination, all of these patients enjoyed good general health and seemed to be well nourished. Mental deficiency is noted in two cases (VII and IX). Moral conduct and disposition are good in all but these two, and with the exception of case XV, the records show no history of fits, scrofula, or cutaneous disease. Case XV had convulsions in infancy, but not subsequently. Furthermore, no infirmity other than the combined infirmity of deaf-mutism and retinitis pigmentosa, and in cases VII and IX, mental deficiency, was found in the series.

EXTERNAL OCULAR EXAMINATION.

A. The conjunctiva and lacrimal apparatus were negative in all but three cases.

I. Conjunctiva thickened, lacrimation.

X. Conjunctiva injected, lymphatic swelling in lower fornix O.S.

XIV. Blepharitis marginalis; palpebral and bulbar conjunctiva injected.

B. The pupils are equal, of average diameter and functionally active in all but two cases.

VI. Pupils abnormally small.

XI. Iridodonesis (Aphakia).

C. Ocular movements normal in all but one case.

XV. O.D.—*Vertical nystagmus;* movements 2–3 per second; cease in downward rotation.

O.S.—*Rotary nystagmus;* movements not so rapid as in O.D.; smaller amplitude.

VISUAL ACUITY.

This has been recorded in all but three cases (VI, VII, XI) and, as so often noted in descriptions of retinitis pigmentosa, was remarkably good considering the degeneration of the retina and optic nerve. Eight of the cases had vision of $\frac{5}{7\frac{1}{2}}$ or better, in one eye at least. The lowest uniocular vision was $\frac{5}{20}$, and the lowest binocular vision was $\frac{5}{15}$. It is probable, too, that these figures represent a lower standard than the subjects actually possessed, for any one who has had experience with young deaf-mutes, will know that it is very difficult as a rule to get them to make their best efforts in tests of this kind, about which they know nothing.

REFRACTION.

This was determined by retinoscopy under full scopolamin mydriasis in 16 of the cases, and can best be shown as follows:

E — O_2 in 1 case.
H — O_2 in 1 case.
H + A H, O_2 in 8 cases.
M + A M, O_2 in 2 cases.

ANTIMETROPIA.

E with H in 1 case.
H + A H with H + A M in 1 case.
H with H + A H in 1 case.
M + A M with H (Aphakia) in 1 case.

The refractive error was not high except in cases I–VI and XV. This series is entirely too small to add anything to the statistics upon refraction, but from the writer's records of several hundred refractions from the same institutions (not yet published) it would seem that refractive errors are not more numerous nor higher among deaf-mutes than among others not thus afflicted, of similar age and environment.

It was quite impossible in these cases to gain any information regarding the accommodation, and no attempt was made. It is probable, however, that could this be done, marked deficiencies would be found, for we know that in retinitis pigmentosa degeneration attacks practically the whole inside of the eyeball, and changes were found by Wagenmann and Deutschmann in the ciliary processes and ciliary body, iris, etc. Equally

impossible was it to test the extra-ocular muscles, but there is little reason to think that changes of special note referable to the disease under consideration would have been found.

OPHTHALMOSCOPIC EXAMINATION.

Media.—The media were clear both eyes in 10 cases, and in the remaining 7 cases showed changes as follows:

Vitreous opacities in 4.

Corneal opacities in 1.

Lens cataractous in posterior cortex temporal side, O.D.; Aphakia O.S in 1.

Media hazy both eyes; cause not stated in 1.

It will be noted that in not a single case was there the posterior polar cataract so often referred to as occurring in retinitis pigmentosa. It will also be observed that vitreous opacities were relatively frequent—being found in *23.5 per cent.* of the cases. This we urge against the tendency in doubtful cases, to consider vitreous opacities as evidence, more or less positive, of chorioretinitis specifica as opposed to retinitis pigmentosa.

Optic Disk.—The disk in retinitis pigmentosa is always atrophied, and no exception is to be found in this series. The disk margins were blurred in 11 cases, well defined in 4 cases, and not noted in 2. The color of a disk is not easily described so as to convey to a reader

an accurate idea of its appearance, but where noting the color, we have used those familiar terms as selected by others to best include the disk of retinitis pigmentosa. Thus we found it dull and waxy; dirty red gray and opaque; yellowish and opaque; the color of old parchment; the color of bees' wax; and in one case *white.*

However the color may be described, it is always far from normal, and once seen leaves no doubt in this regard even in the mind of a beginner. Another characteristic of the disk is its *flatness and uniformity.* In only one case of our series was a physiological excavation in evidence, and it might be said that as any portion of the disk appears, so appears the entire disk.

The Retina.—This as a tissue shows unmistakable sclerosis of varying degrees. We have noted it as *markedly* sclerosed 12 times. The chorioidal circulation when exposed by the dissipation or destruction of the retinal pigment, is not clearly and sharply defined, owing to the grayness and loss of transparency in the retina. In other words, the retina, which on account of its transparency, is normally invisible, becomes in retinitis pigmentosa no longer perfectly transparent, and hence visible.

The Retinal Vessels.—Both arteries and veins were reduced in size in all cases; great reduction was noted in 7 cases, and slight or moderate reduction in 10. In one case the diminution in size was great in one eye, and slight in the fellow eye. In three cases the arteries were

reduced relatively more than the veins; in 5 cases, the veins suffered the greater reduction, and in 9 the reduction was uniform. The smaller branches and terminals fall, of course, below the limit of observation and are ultimately entirely obliterated, as we know that the reduction in the size of the retinal vessels is greatly at the expense of the lumen.

Pigment.—Considering the pigment in regard to quantity, character, and distribution, considerable variation is found.

It is described as of small amount in 7 cases, including one case in which there was no pigment, of moderate amount in 2 cases, and as extensive in 8 cases. The ages of those showing little pigment range from 8 to 15 years, and of those showing extensive pigment deposit from 10 to 19 years.

The classical bone corpuscle-like or lace-like deposits, either alone or in combination with other formations, were seen in 5 cases. In other cases, round or irregular spots, disks, balls or masses, chains and lines were observed. Most characteristic is it, that only in exceptional cases of retinitis pigmentosa do we find *large* masses or accumulations of pigment, and even then there is apt to be considerable interspacing. This is the natural result of the method by which the pigment reaches the inner layers of the retina. It *wanders,* little at a time and very slowly, and is therefore less likely to settle in considerable amount at any one place.

The distribution was limited in this series to the periphery and equator, some cases showing pigment as far toward the periphery as could be seen. Although mostly in the nerve-fibre layer of the retina, it was occasionally noted in the deeper layers as well. In a number of cases the pigment seemed to bear no relation to the blood-vessels, while in others it lay along or close to the vessels, and in one case (Plate III) portions of the blood-vessels were completely ensheathed by the pigment. The polar area of the fundus was pigment-free in every case.

It is interesting to note that Case XI is sine pigmento, and is, perhaps, the worst case of the series. This case is of consanguineous origin, has cataract, very extensive sclerosis of the retina, and atrophy of the nerve, greatly reduced blood-vessels, complains much of night blindness, and has the form field reduced to 20°. Pigment will undoubtedly make its appearance in the retina sooner or later.*

VISUAL FIELDS.

It was possible to plot the visual fields in thirteen cases. This was most carefully done by Dr. Mary Buchanan. Efforts were made to obtain fields for 10 mm. objects of white, red and green; and central color perception was tested for 2 mm. objects. The

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* See footnote on page 9.

fields thus obtained are all very much contracted, and it will be observed that the greater the contraction, the greater is the relative loss to the temporal side, thus ultimately producing an almost circular field with the fixation point as the centre.

This is particularly apparent in Cases III, IV, VI, VIII, XI, XII (one eye), XIII and XVI, or in 61.5 per cent. of the cases. In most of the cases the form field shows a relatively greater reduction than the color fields. In cases II, XIV, and XV, a more normal relationship is shown.

Comparing cases of the series, the fields do not seem to correspond always with the fundus condition. For example, the fields in Case XIII are better than would be expected from an examination of the fundus, and a comparison of Cases VIII and XV shows a great want of harmony in this respect.

Scotomata of any kind were not discovered in a single case, and while the writer realizes that the sources of error in the perimetric examination of these patients were necessarily great, in so far as the ring scotoma is concerned, he would regard this defect as most unexpected. The retinal sclerosis, the vascular changes and the optic atrophy showed no local variations that could account for a ring scotoma.

Central color perception was remarkably good even for 2 mm. squares, the exceptions being VIII with no color fields, and indefinite recognition of color centrally; IX good for 2 mm. squares O.D., but requiring 5 mm.

squares for O.S., and XI, green recognized only in 20 mm. squares; smaller areas called blue.

The conclusions drawn from these fields are:

A. In retinitis pigmentosa the form field suffers greater and more rapid contraction than the color fields, indicating that the peripheral retina is first to become affected. This is in accord with the fundus findings.

B. The fields tend to become *circular* around the fixation point.

C. Color perception is well maintained in the presence of the most extensive atrophy and degeneration.

D. Ring scotoma must be in this disease a rare field defect.

CHAPTER VI.

LABORATORY FINDINGS IN CASES OF RETINITIS PIGMENTOSA.

Blood-counts were made in eleven of the cases of retinitis pigmentosa, in order to determine whether or not the condition was accompanied by a uniform change in the blood picture. By reference to the appended results, it will be seen that there was no definite change in the blood of these patients.

The erythrocytes varied between 6,120,000, the highest, and 4,130,000, the lowest. The leucocytes varied from 25,120, the highest, to 10,960, the lowest. All but one of these counts were made in the middle of the afternoon, in the neighborhood of three o'clock, and the slight degree of leucocytosis, both absolute and relative, observed was probably due to the normal increase of white blood-corpuscles following a meal. The leucocytosis of 25,120 in Case XI was apparently purely accidental.

The hemoglobin varied from 125 per cent., the highest, to 80 per cent., the lowest. The color-index, it will be seen, was in all cases that of a slight degree of chloroanemia, except Case IX, in which the color-index was high.

The differential leucocyte count showed nothing abnormal. All of the patients were under twenty-one

years of age, with the exception of one (the last), and the comparatively high percentage of lymphocytes in the counts may be ascribed to the normal lymphocytosis of childhood. In the last case there was anisocytosis, and a few poikilocytes and some degenerated leucocytes were observed. This cannot be considered a characteristic of the disease under consideration.

The counts were made in the customary way, with the Thoma hemocytometer. The hemoglobin was determined by means of Fleischl's hemoglobinometer. The differential counts were made in specimens stained by Wright's method, and in every case five hundred leucocytes were counted.

Urine examinations in these cases were made at intervals of two or three months, in order to determine whether the disease, retinitis pigmentosa, was accompanied by any constant or characteristic change in metabolism, as expressed by alterations of the composition of the urine. All of the patients except one were inmates of an institution and were subject to practically the same conditions. They were on a mixed diet containing a fair proportion of the various ingredients of a normal diet.

Breakfast for the younger children consisted of oatmeal, bread and butter, and milk. On Sunday morning, in addition to these articles, each child had one egg. Dinner on Monday consisted of roast beef, potatoes, stewed tomatoes, gravy, and rice pudding; on

Tuesday of vegetable soup, mutton stew with potatoes and peas; on Wednesday of Hamburg steak, potatoes and gravy, corn, and corn-starch pudding; on Thursday of rice soup, beef stew with potatoes and beets; on Friday of fish, scrambled eggs, or oyster stew, potatoes, coleslaw and bread pudding; on Saturday of bean soup, roast beef, potatoes and gravy, and stewed onions; and on Sunday of Hamburg steak, mashed potatoes, baked beans, pickles, and fruit, custard or junket. None of these children received coffee or tea. For supper the diet consisted of bread and butter, milk and stewed fruit or a cereal.

The breakfast for the older children consisted of bread and butter, stewed fruit, oatmeal, milk, coffee, and on one day of the week eggs, and on another day hash. For dinner on Monday they received corn soup, roast mutton, boiled potatoes and peas; on Tuesday roast beef, mashed potatoes, corn, and rice pudding; on Wednesday mutton stew, potatoes, stewed tomatoes, and bread pudding; on Thursday vegetable soup, corned beef and cabbage and boiled potatoes; on Friday fish, potatoes, coleslaw, and coffee; on Saturday bean soup, sometimes roast veal, turnips, and potatoes; on Sunday roast beef, mashed potatoes, baked beans, pickles, and custard or fruit. For supper, on Monday, they received bread and butter, beef hash with potatoes, syrup, and milk; on Tuesday bread and butter, mutton hash, milk, stewed peaches, biscuit, and tea; on Wednesday

boiled rice or cold meat, bread and butter, milk, tea, and stewed prunes; on Thursday hash, milk toast, syrup, bread and butter, ginger-cake, tea, and milk; on Friday hominy grits or scalloped potatoes, stewed pears, bread and butter, tea and milk; on Saturday hash, bread and butter, syrup, and tea.

At the time that the urine was to be collected for examination, the child was removed from its dormitory and placed in the infirmary for twenty-four hours, where under the care of a trained nurse, the 24-hour urine was collected in a clean vessel, and subsequently sent to the laboratory. During the child's residence in the infirmary he received the same diet as that received by the children in the dormitories.

The urine was first measured to determine the total quantity, and then was examined for its total nitrogen, for ammonia nitrogen, for preformed sulphates, for conjugate sulphates, or phosphates, and for volatile fatty acids, indican, phenol, urobilin, acetone, albumin, and glucose.

The total nitrogen was estimated by the Kjehdal method, using mercury in the digestion. The ammonia nitrogen was determined according to the method of Krueger and Reich (*Zeit. f. physiol. Chem.,* Bd. xxxix, p. 165). Twenty c.c. of urine were diluted with about 400 c.c. of distilled water, and twenty drops of alcohol were added and 10 c.c. of a saturated solution of barium hydroxide. This mixture was then distilled in a vacuum

for thirty-five minutes. The distillate was collected in $\frac{N}{10}$ solution of sulphuric acid and titrated with $\frac{N}{10}$ solution of sodium hydroxide, using litmus or congo red as the indicator.

The preformed sulphates were determined according to the method of Baumann (*Zeit. f. physiol. Chem.*, Bd. i., p. 70). One hundred c.c. of urine were filtered and diluted with an equal volume of water. The diluted urine was then acidulated with acetic acid, warmed, and an excess of saturated solution of barium chloride added. The mixture was then kept warm on an iron plate covered with asbestos, until the precipitate had settled. The precipitate was collected on a filter having an ash of known weight, and the filtrate was saved for the determination of the conjugate sulphates. The precipitate was washed with hot water, hot dilute hydrochloric acid, hot alcohol, and ether, and the dried precipitate was placed in a platinum crucible together with the ash of the filter paper, and the whole was incinerated. After the first incineration the cool residue was moistened with dilute sulphuric acid in order to replace any barium sulphate which might have been reduced by the organic matter in the precipitate, and, after driving off the excess of sulphuric acid by carefully heating, the crucible was again strongly heated, cooled, and weighed.

In order to determine the conjugate sulphates, the first filtrate from the determination of the preformed sulphates was saved, and 10 per cent. of its volume of

hydrochloric acid was added, and the whole boiled for ten minutes. A small additional quantity of saturated solution of barium chloride was added, and the precipitate was allowed to settle, was collected, washed, incinerated, and weighed, in the same manner as indicated for preformed sulphates.

The phosphates were determined by titrating with a standard solution of uranium acetate, the end reaction being determined by using a 10 per cent. solution of potassium ferrocyanide as an indicator.

The indican was determined by the method of Obermayer (*Wien. klin. Woch.*, 1890, No. 9, p. 176). To 20 c.c. of urine 5 c.c. of basic acetate of lead solution were added, and the mixture filtered. A small quantity of the filtrate was agitated with an equal volume of a solution of ferric chloride in hydrochloric acid (ferric chloride, 1 gram, strong hydrochloric acid, 500 c.c.). Five c.c. of chloroform were then added, and after a thorough shaking the presence of indican was determined by the blue color of the chloroform.

The acetone was determined by the method of Penzoldt (*Archiv f. klin. Med.*, 1883, No. 34, p. 132). To 100 c.c. of urine 2 c.c. of 50 per cent. solution of acetic acid were added, and the mixture was distilled until about 30 c.c. had been obtained as distillate. A small crystal of orthonitrobenzaldehyde was dissolved in hot water, and when cool, a portion of the distillate was added to it, together with a small quantity of 10 per cent. sodium hydroxide solution. In the presence of acetone

the color of the liquid changes to yellow, green, and finally to blue.

In order to determine the presence of volatile fatty acids and phenol, 50 c.c. of dilute sulphuric acid were added to 250 c.c. of urine. This mixture was then distilled in a current of steam until the distillate was no longer acid in reaction. One-half of this distillate was then titrated with $\frac{N}{10}$ sodium hydroxide solution, and the amount of volatile fatty acid was expressed in terms of cubic centimetres of $\frac{N}{10}$ sodium hydroxide solution required for neutralization. Congo red or litmus was used as the indicator. The other half of the distillate was treated with a small quantity of 10 per cent. solution of ferric chloride or with bromine water. In the presence of phenol the iron solution produces a violet-blue color; bromine water produces a yellowish-white, crystalline precipitate of tribromphenolbrom.

Urobilin was determined by treating 50 c.c. of urine with a few drops of hydrochloric acid and adding 25 c.c. of amyl alcohol. The mixture was then thoroughly agitated in a separatory funnel, and the alcoholic extract examined for the absorption of bands of urobilin by the spectroscope.

The albumin was tested for by the heat and nitric acid method. Glucose was tested for with Fehling's solution. None of the specimens of urine contained either albumin or glucose.

The examination of the urine of these patients by the methods above enumerated failed to reveal any constant expression of abnormal metabolism. As a rule, the total quantity of urine passed in twenty-four hours was low. The total nitrogen excretion was also lower than normal, taking 15 grammes of nitrogen as a high normal average excretion per day, for an adult. The majority of these patients were children, however, and it is possible that the low nitrogen excretion may be accounted for by the age of the patients. The ammonia nitrogen was in no case excessive. Excretion of sulphates showed no uniform increase or decrease. Some patients presented about a normal sulphate excretion, others a decrease, and still others a slight increase in the amounts of these bodies excreted during the twenty-four hours. The phosphate excretion was uniformly low, never rising to as much as 2 grammes of P_2O_5 in twenty-four hours. The volatile fatty acid excretion presented wide fluctuations. Indican, phenol, urobilin, and acetone were usually absent. Occasionally a trace of one or the other of these substances would be met with, but there was never enough of these bodies present to lead to the conclusion that any abnormal metabolic process was going on in the patients.

As a result of the examination of the blood and the urine in these cases of retinitis pigmentosa, we should say that *no light can be thrown on the nature of the disease, and no assistance in its diagnosis given, by the examination of either of these body fluids.*

BLOOD EXAMINATIONS

	CASE II	CASE III	CASE IV	CASE VI	CASE VIII	CASE IX	CASE XI	CASE XII	CASE XIV	CASE XV	Case Not in Series
Erythrocytes	5,360,000	4,760,000	5,030,000	4,130,000	4,190,000	4,650,000	5,560,000	5,510,000	4,980,000	4,170,000	6,120,000
Leucocytes	12,080	25,120	13,360	10,960	15,840	14,240	12,400	13,040	12,640	11,040	10,480
Ratio	1 : 444 —	1 : 189 +	1 : 307 +	1 : 376 +	1 : 265 +	1 : 326 +	1 : 448 +	1 : 422 +	1 : 394 —	1 : 368 +	1 : 583 +
Hemoglobin	84%	82%	88%	91%	80%	87%	85%	93%	89%	79%	105%
Color Index	0.78 +	0.86 +	0.87 +	1.10 +	0.95 +	0.93 +	0.76 +	0.84 +	0.89 +	0.94 +	0.85 +
DIFFERENTIAL COUNTS:											
Polymorphonuclears	61.2%	75.2%	67.2%	65.0%	70.8%	70.4%	63.2%	73.0%	61.0%	66.0%	78.8%
Lymphocytes	31.4%	19.0%	24.4%	24.8%	23.2%	25.2%	29.2%	16.4%	33.6%	28.0%	11.6%
Transitionals	5.0%	3.0%	3.8%	6.2%	3.6%	2.6%	5.6%	7.0%	3.0%	5.4%	5.4%
Eosinophiles	2.2%	1.6%	3.0%	4.0%	1.8%	1.8%	1.8%	2.2%	1.6%	0.6%	3.8%
Basophiles	0.2%	1.2%	1.6%	.....	0.6%	.....	0.2%	1.4%	0.8%	.....	0.4%
	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%	Erythrocytesvary in size. Few poikilocytes. Some degenerated leukocytes.

URINE ANALYSES

	CASE II		CASE III		CASE IV		CASE VI		CASE VIII		CASE IX	
	Nov. 9, 1903	Feb. 15, 1904	Jan. 18, 1904	Mar. 21, 1904	Nov. 3, 1903	Feb. 8, 1904	Dec. 20, 1903	Mar. 14, 1904	Jan. 11, 1904	Mar. 14, 1904	Dec. 14, 1903	Mar. 7, 1904
Total quantity	600 cc.	550 cc.	795 cc.	790 cc.	680 cc.	1130 cc.	650 cc.	750 cc.	540 cc.	900 cc.	940 cc.	940 cc.
Total nitrogen	4.6200	8.2544	9.7053	8.6931	6.7553	9.9982	9.828	6.237	7.5448	7.2828	11.7387	8.6066
Nitrogen expressed as urea	9.8960	17.6809	20.7888	18.6207	14.4700	21.4162	21.0515	13.3596	16.1611	15.5997	25.2444	18.4354
Ammonia nitrogen	0.4284	0.4158	0.3116	0.2709	0.5521	0.2689	0.31950	0.1941	0.2608	0.3061	0.5461	0.3816
Preformed sulphates,												
Expressed as H_2SO_4	0.6913	1.9446	1.7479	2.7407	0.9266	1.7725	2.6782	2.4335	1.1700	2.2142	2.3134	3.5794
Expressed as SO_3	0.5642	1.5872	1.4267	2.2370	0.7563	1.4467	2.1860	1.9862	0.9549	1.8073	1.8882	2.9215
Conjugate sulphates,												
Expressed as H_2SO_4	0.0754	0.1863	0.1670	0.1201	0.0775	0.1518	0.1245	0.0748	0.0026	0.2216	0.1389	0.1120
Expressed as SO_3	0.0615	0.1520	0.1363	0.0980	0.0632	0.1239	0.1016	0.0611	0.0021	0.1808	0.1134	0.9148
Phosphates as P_2O_5	lost	1.0997	1.7584	1.6413	lost	1.4769	1.7265	0.9875	1.0253	1.1587	1.9367	1.7604
Volatile fatty acids	58.08	7.7	17.31	23.70	38.624	33.448	34.84	6.00	27.0	14.4	24.816	37.6
Indican	none	slight	sl. trace	present	marked	trace	marked	present	trace	none	marked	trace
Phenol	none	none	none	none	none	none	none	none	none	none	none	none
Urobilin	trace	marked	present	trace	trace	lost	present	none	none	present	none	present
Acetone	none	present	trace	trace	none	trace	none	none	trace	none	none	none
Albumin	none	none	none	none	none	none	none	none	none	none	none	none
Glucose	none	none	none	none	none	none	none	none	none	none	none	none

URINE ANALYSES—*Continued*

	CASE XI		CASE XII		CASE XIV		CASE XV		CASE NOT IN SERIES	
	Nov. 24, 1903	Feb. 29, 1904	Dec. 7, 1903	Feb. 29, 1904	Nov. 16, 1903	Feb. 22, 1904	Nov. 30, 1903	Mar. 7, 1904	Nov. 13, 1903	Dec. 23, 1903
Total quantity	950 cc.	1130 cc.	690 cc.	855 cc.	800 cc.	960 cc.	860 cc.	700 cc.	600 cc.	930 cc.
Total nitrogen	4.6018	5.5786	7.6738	8.7141	9.5244	9.408	6.4534	9.1924	9.8582	11.369
Nitrogen expressed as urea	9.8570	11.9494	16.4374	18.6657	20.4012	20.1519	13.8232	19.7901	21.1162	24.3523
Ammonia nitrogen	0.4522	0.3717	0.4250	0.3291	0.8736	0.3964	0.6862	0.3626	0.7812	0.3255
Preformed sulphates,										
Expressed as H_2SO_4	0.7609	1.6004	1.7742	3.6506	1.7041	2.3190	1.1186	2.7885	1.7805	3.3215
Expressed as SO_3	0.6211	1.3063	1.4381	2.9796	1.3909	1.8928	0.9130	2.2760	1.4532	2.7110
Conjugate sulphates,										
Expressed as H_2SO_4	0.09069	0.0862	0.0840	0.0969	0.1537	0.1879	0.9223	0.0189	0.1022	0.1056
Expressed as SO_3	0.0740	0.0704	0.0686	0.0791	0.1219	0.1534	0.7527	0.0154	0.0834	0.0862
Phosphates as P_2O_5	lost	0.8597	1.3519	1.5738	lost	1.2079	1.2334	1.6388	lost	1.8316
Volatile fatty acids	11.4	*424.88	33.672	54.72	31.36	75.58	94.944	18.2	34.56	67.704
Indican	none	trace	trace	trace	marked	marked	trace	present	trace	marked
Phenol	none	none	none	none	trace	none	none	none	none	none
Urobilin	trace	present	none	present	trace	present	none	present	trace	none
Acetone	none	none	none	none	none	none	none	none	none	none
Albumin	none	none	none	none	none	none	none	none	none	none
Glucose	none	none	none	none	none	none	none	none	none	none

*There was probably an error in this determination.

CHAPTER VII.

PROGNOSIS; TREATMENT AND MEDICAL DIRECTION; CONCLUSIONS.

PROGNOSIS.

The victim of retinitis pigmentosa if he live his allotted time of three score years and ten, will in most cases be blind. Long before this time, however, he will have lost possession of useful vision. The contracted field and the night blindness will rob him of the power of orientation, and if able to read at all, it will be by slowly passing from letter to letter, the place once lost being difficult or impossible to find. The degeneration doubtless has a limit, and it is possible for a certain amount of central vision to escape when that limit has been reached, the disease then remaining stationary; but so far as the writer knows, this stationary phase never occurs early, leaving useful vision for an indefinite time.

If a curve of degeneration could be constructed for this disease, it would probably show a rather rapid ascent to the point of very pronounced deterioration, then a gradual ascent toward extinction, and finally terminate in a parallel.

From the teaching born of experience in the observation of this disease by countless physicians, we are not at the present time justified in offering a prognosis better than above indicated.

TREATMENT AND MEDICAL DIRECTION.

Until medical science can regenerate atrophied nerve fibres and sclerosed tissue; specialize connective tissue; enlarge blood-vessels, reduced in lumen from thickening of the walls, etc., retinitis pigmentosa must be considered incurable in so far as correcting damage already done is concerned. Whether or not the progressing changes when recognized can be checked, is open to discussion. As a broad principle, we know that the greater the power of resistance possessed by an individual, the better can invasion of any kind be combated, and the ultimate chances of an individual attacked by retinitis pigmentosa would be theoretically at least, better, were hygienic and other measures instituted from its first recognition, and continued throughout.

But right here comes the greatest of difficulties. Medical attendance and advice extended over a term of years, ten, twenty, thirty, or more, when pain and discomfort are absent, however reasonable theoretically, are practically impossible of attainment.

When advice is first sought, much damage has in most cases been established, and so gradual is the subsequent deterioration, that adaptation to existing conditions is apt to claim more of our attention than remedial measures. The literature contains numerous reports of cases thought to have been benefited by treatment of one kind or another; vision has been reported improved and the fields extended, but most of these reports fail when

knowledge concerning the permanency of the improvement is sought.

Thus strychnia hypodermically, phosphorus, mercury, the iodides, the constant and intermittent electric current, etc., have all been advocated, and improvement claimed from their use. Marcus Gunn even suggested, some years ago, that the galvanic current might cause an enlargement of the blood-vessels. Surely such a result must be considered highly improbable when we think of the structural changes which have overtaken these vessels, and produced the diminution in size.

If drugs or electricity can directly influence the retina and optic nerve in retinitis pigmentosa, such influence must, in the opinion of the writer, be in the nature of a temporary stimulation. A jaded horse might be stimulated to greater activity by judicious application of the whip, but as a horse, he is not thereby improved.

On the other hand, as indicated above, the early and continued institution of all means directed toward constitutional normality, with perfect assimilation, nutrition, and functional activity, would in all probability deter the degeneration and benefit the victim to that extent.

Unfortunately such a course can seldom be pursued. In the patients of this series it would be impossible. Their social surroundings and possibilities are as unalterably opposed to progressive medicine as would be Gibraltar to an invading fleet. In a word, retinitis pigmentosa would seem to be *incurable;* real improvement

is doubtful, and arrest of the processes is possible but not proven, the evidence at hand being more constructive then analytical.

The advice and direction to be given these patients form no small part of the physician's duty toward them.

With practical, if not absolute blindness before them, and often with years of warning, time might well be devoted to a certain amount of preparation. So much progress has been made by the Societies interested in the publication of literature for the blind during the last few years, that the usefulness and happiness of these unfortunates have been increased a hundred fold. The mastery of one or more of the several systems of reading for the blind, while there is yet vision, then a comparatively easy task, should be advised. Many dark and gloomy hours would thus be replaced by those of pleasure and profit. Javal, the eminent French ophthalmologist, despite his affliction, turned night into day, and in a little work entitled, "*Entre Aveugles,*" which has been translated into many languages, has opened for the blind avenues to enjoyment and usefulness which were hitherto closed. When blindness is contemplated, a study and practice of Javal's experiences would amply repay the time expended.

Finally, advice may be sought on the question of marriage. The writer is not in sympathy with the most advanced ideas of a few, striving to correct the world's evils, who believe that diseases should be stamped out by

enforced celibacy. He believes that every man or woman, of *sound mentality,* has a *right* to marry, the already existing laws, of course, being complied with. The contracting parties should most certainly know of all infirmities, and their ultimate prospects. That being the case, marriage becomes a matter of choice, and the ones interested should do the choosing.

It is unquestionably the duty of the physician to *advise* against unions likely to result in defective offspring, but to concoct *prohibitive legislation* would, if carried to its ultimates, entail hardships not consistent with a free people.

With regard to retinitis pigmentosa and kindred diseases, statistics show that while direct transmission to the next generation is rare, the appearance of the disease in subsequent generations is common. It must be noted also that the lives of many who have had retinitis pigmentosa have been useful, and not spent in vain. Through childhood and adolescence, and even through manhood and womanhood, sunshine has entered in fair, or perhaps, full amount. Is it just, then, to conclude that because A is less fortunate than B, A never should have existed?

The writer would urge, therefore, that medical men, when called upon to advise in matters of this kind, take a broad and careful survey of the situation presenting, and render that advice per all things considered, rather than condemn without a hearing, because it is customary.

CONCLUSIONS.

From the foregoing review of the literature, and from personal observations in the study of a number of cases, including those among deaf-mutes here reported, the writer would derive the following *fundamental* conclusions concerning retinitis pigmentosa.

The disease is a distinct and definite process, always presenting a combination of phenomena absolutely characteristic and practically invariable. It is a degeneration of the entire neurovascular tract of the peripheral end organ of vision, extending certainly beyond the orbit and into the brain, but just how far, pathological examinations into the central nervous system of these cases have not as yet revealed. *It is not an inflammation;* a fact early recognized and now very generally conceded.

Within the eyeball, the primary tissue involved is the chorioid, and the striking retinal changes are, for the most part, secondary. The name *retinitis pigmentosa,* in that it does not correspond well to the true nature of the disease, is poorly selected. The retinal changes are inseparable from those in the chorioid, and are secondary to them; therefore, if the term retina is to enter the title, it should follow chorioid as *chorioretinal.*

The terminology indicative of inflammation is likewise out of place, as has been so frequently suggested, and should give way to the term *degeneration,* or *sclerosis.* This being done, descriptive adjectives can be

introduced ad libitum, and correctness be maintained as far as they go.

The name retinitis pigmentosa is, however, established and well understood, and is, furthermore, practical in that it carries with it an idea of the disease intended. For this reason it has been used throughout this essay.

The disease is congenital in origin, however late its manifestations come to light. It might well be looked upon as a stigma of degeneracy, and finds its purest expression in the presence of other degeneration stigmata. For example, the retinitis pigmentosa found in association with congenital deaf-mutism is the type par excellence.

Heredity is a potent etiological factor; parental consanguinity is of importance only in connection with heredity and environment; impressionism cannot be excluded; and syphilis as a cause of true retinitis pigmentosa has not been established, nor has it commended itself to the majority of observers.

The literature teems with cases of supposed retinitis pigmentosa and conclusions derived therefrom, but many of these cases, in the opinion of the author, do not, through carelessness and inattention on the part of the observers, represent this disease. Pigment in the retina is not retinitis pigmentosa, and chorioretinitis of various kinds, notably syphilitic, may closely simulate in ophthalmoscopic appearances true retinitis pigmentosa.

Some observers even go so far as to designate certain of their cases as the " syphilitic cases," which provokes the inference that such are cases of syphilitic chorio-retinitis.

The symptoms and manifestations of the disease are well understood, and aside from opposing the generally accepted frequency of ring scotoma, nothing of importance remains to be said. The presence of a ring scotoma would indicate a selective tendency on the part of the process. The evidence is against such selection in retinitis pigmentosa, almost all of the structures involved showing a uniform and similar invasion.

Retinitis pigmentosa sine pigmento is, as a disease, identical with retinitis pigmentosa.

Finally, unilateral retinitis pigmentosa is inconsistent with the author's ideas of the nature of the process.

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