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BY

WM. F. NORRIS, M.D.

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HEREDITARY ATROPHY

OF THE

OPTIC NERVES.

BY

WM. F. NORRIS, M.D.,

PHILADELPHIA, PA.

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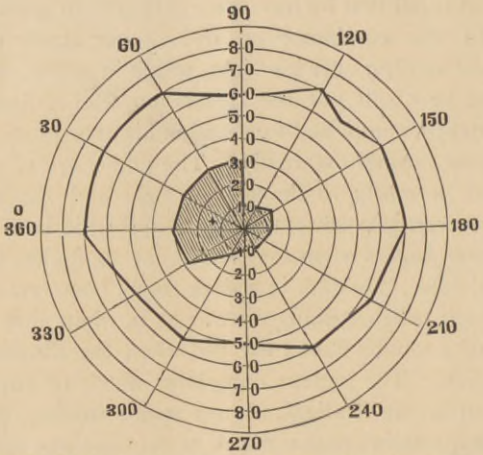
HEREDITARY ATROPHY OF THE OPTIC NERVES.

CASE I.—A. H. A., aged forty-nine, is a well-developed, vigorous-looking man, never addicted to the use of either alcohol or tobacco; complains that for about a year he has been losing sight, and is now so blind that he has some difficulty in getting about by himself. He was conductor on one of our steam railways, but found a cloud settling over his sight, which in a few weeks became so dense that he could not see signals nor read ordinary print, and was consequently compelled to abandon his avocation. At present, his eyes appear normal externally. The pupils are of medium size, and respond promptly to changes of light and shadow. Central vision is, however, almost completely abolished in both eyes, and he can only count fingers when they are about six inches to either side of the visual axis. He can, in a dark room, however, see a gaslight five feet distant, when looking directly at it, when it is turned down to the size of a candle-flame, but not when the illumination is farther diminished. The patient complains much of any bright light, and when out in the sunlight, always wears smoked glasses. The ophthalmoscope shows that the disk of the right eye is slightly ovoid vertically, with a choroid ring all around, passing into an irregular black-peppered *conus* to the outside, which is one and one-half times the width of the main central veins. There is a slight central excavation, but the rest of the disk is of a greenish-gray hue, a trifle prominent. It is entirely devoid of capillarity, but the central vessels have about their usual calibre. The choroid is perhaps more granular than usual; but beyond this, and a very slight retinal haze around the disk, there are no demonstrable changes in either it or in the retina. In the left eye the disk is more nearly circular; there are remains of a choroidal ring which has existed all around, but is now absorbed in places. The scleral ring is too marked all around, and outward there is a black speckled *conus* one and one-half times the width of the main retinal veins.

The patient was given $\frac{1}{32}$ of a grain of strychnia three times

daily; the quantity of this drug was gradually increased till he took $\frac{1}{16}$ grain at each dose, and in about a month, there was marked improvement, so that the accompanying field of the right eye could be taken with a Sherk's perimeter, candles being used for the fixation point and also for the peripheral object—the scotoma was outlined by the use of a black penholder—the candle-flame still being used for fixation. The vision was now $\frac{4}{CC}$, when the patient looked about six inches below the letter to which his attention was directed. The peripheral vision has so much improved that he can walk about with much greater confidence. The strychnia treatment was continued for some time longer, but apparently with no further improvement of the vision.



Field of right eye of A. H. A., showing extent and outline of central scotoma and limits of peripheral light perception.

Case II.—C. H. K., aged fifty years. He is a first cousin of the former patient on the mother's side, and complains of the same central cloud in his field of vision. The right eye distinguishes Sn. CC at one foot, the left, Sn. CC at five feet; in both cases with eccentric fixation. In a dark room, patient could distinguish through the central cloud, when looking directly at it (with the right eye), a gas-flame turned down to about the size of a usual candle-flame, and with the left eye, the same object when it was diminished to one-fourth of this size. Examination of the field with candles shows in right eye an irregular central scotoma, varying from about 15° to 20° around the point of fixation. The candle is seen most distinctly in the lower outer part of the field. In the left eye the

image is dim at fixation point, and there is a cloud extending from it in every direction about 5° below and about 20° above; vision much the best in the lower part of the field. The pupils here, too, were of medium size and prompt, but the patient is not so much inconvenienced by strong light as in the other case. The ophthalmoscope here, also, shows a greenish atrophy of the optic nerves, with entire want of capillarity of the disk, and but little change in the calibre of the central vessels. In each eye, the scleral ring is unduly marked all around the disk. A similar treatment was inaugurated, and after some weeks' use of the strychnia the central cloud was slightly diminished in density, and the peripheral vision markedly improved. The patient remarks that "now, when looking in the glass, he can see his own gray moustache." O. D. V. $= \frac{3}{cc}$; O. S. V. $= \frac{6}{CLXX}$; in both, eccentric fixation.

In both patients, the patellar tendon reflex was examined and found to be prompt.

These patients are first cousins, their mothers being sisters; and thus give a most interesting history of hereditary atrophy, beginning with the maternal great-grandfather and extending through four generations. The blindness has usually been transmitted by the female side of the house, and has affected both males and females, but the former much more frequently. It is noteworthy that an uncle, after being "blind," became so much better, that he could resume ordinary occupations, but this has not been the case in any other affected member of the family. The patients deny any consanguineous marriages. The following genealogical table exhibits the affected members of the family:

G.g.g.f. — G.g.f. — G. m. (affected). (affected). (affected).

- Uncle (affected, but improved). — Cousin (affected).
- Mother (not affected).
  1. A. H. A. (patient), male, 49 yrs. (affected).
  2. Female, 46 years (not affected).
  3. Died in infancy.
  4. Male, 43 years (affected).
  5. Died in infancy.
  6. Died in infancy.
  7. Female, 33 years (not affected).
  8. Died in infancy.
  9. Died in infancy.
- Aunt (affected). — Cousin, patient C. H. K., male (affected).

These cases form, to a great extent, a complement and contrast to those presented by typical pigment degeneration of the retina. They both have this in common, that they are frequently transmit-

ted from generation to generation, but in the one the affection starts always after puberty, often in middle life, with a neuritic atrophy of the optic nerve, and cuts off the central vision at the start, while peripheral vision remains comparatively unaffected. In the other, we have also an hereditary affection of the nervous system, localizing itself in the periphery of the retina often early in life, producing a secondary atrophy of the optic nerve and leaving central vision intact often for many years. Both are said frequently to arise from consanguineous marriages, but both often exist without any such cause.

Most of the individuals of this series became affected between the ages of twenty-five and forty years. The two who were my patients were much benefited by the persistent use of strychnia, which, although it failed to remove the central scotoma, so improved their peripheral vision that they could get about far more comfortably.

Treatment, however, in the great majority of recorded cases, has been unsatisfactory, and very different treatment seems to have succeeded in the hands of different individuals, showing either that the recorded improvements were the natural course of the disease uninfluenced by medication, or, what is more probable, that the various groups of cases are really different in their nature, and that we must endeavor by careful clinical study to further differentiate them. Thus Leber,[1] who has given us a most careful study of this subject and a *résumé* of its literature, remarks, that in his hands, even in the cases which subsequently improved, strychnia has been totally without effect. He recommends mercurial inunctions, and states that he once saw marked improvement after galvanization of the sympathetic in the neck—a proceeding which, however, in other cases proved unavailing. Graefe[2] saw marked improvement from systematic sweating, and Mooren[3] records the only complete cure with which I am familiar, obtained in a patient who came to him two weeks after the commencement of the affection, where, after five months' complete rest of the eyes, and the application of setons to the back of the neck, nitrate of silver was administered with such effect that the patient was able again without effort to read Jaeger 1 with either eye.

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[1] Arch. f. O., XVII., ii., pp. 249–291; and later in Graefe and Sämisch's Handbuch der Augenheilkunde, Vol. V., pp. 824–828.

[2] Graefe, A. f. O., IV., ii., pp. 266–268.

[3] Mooren, Ophthalmologische Mittheilungen, 1873, p. 88.

I cannot, however, but believe that in these, as in almost all other atrophies, that a certain degree of improvement in the *peripheral* vision is to be obtained by the persistent use of strychnia, and that *it is due not to any specific action on the nervous system, but to the fact that it so increases arterial pressure, as to drive a greater supply of blood into the shrunken capillary vessels of the optic nerve and retina, and thus restore proper nutriment to tissues which have been to a greater or less degree in a state of starvation.*[1]

REMARKS.

THE PRESIDENT remarked that the subject was one of considerable interest. A genealogical table was given in the last number of the new British *Journal of Ophthalmoscopic Science* of four generations of patients afflicted with retinitis pigmentosa, most of them minors.

DR. HARLAN, of Philadelphia, said that in the Institution for the Blind, in Philadelphia, there were patients in whose family a history of retinitis pigmentosa could be distinctly traced back seven generations.

DR. KIPP, of Newark, N. J., said Mauthner reported cases in which atrophy of the optic nerve was hereditary in a family several generations, but later color-blindness had developed instead of atrophy, while in the last members of the family the eyes were normal.

DR. LITTLE, of Philadelphia, mentioned the case of a family of Wilmington, Del., in which atrophy of the optic nerve could be traced back two or three generations.

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[1] It is well known that strychnia is an excito-motor and stimulant to the nervous system, especially to the spinal-motor centres. Richter, however, has shown that it causes a very decided rise in the arterial pressure. Sigmund Mayer confirms this, and shows that the rise in blood-pressure is independent of the convulsions and of the frequency of the heart's action. He concludes that the rise of the arterial pressure is due to vaso-motor spasm. The later experiments of Schlesinger (1874) and Klapp (1878) further substantiate these statements. *Vide* a "Treatise on Therapeutics," H. C. Wood, 4th ed., p. 306.