RETINITIS PIGMENTOSA: WITH AN ANALYSIS OF SEVENTEEN CASES OCCURRING IN DEAF-MUTES

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WILLIAM T. SHOEMAKER

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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.

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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,