



## THE RETINA IN HAEMOCHROMATOSIS

BY

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HAEMOCHROMATOSIS consists of an extraordinary derangement of liver and possibly pancreatic metabolism, whereby excessive deposits of iron and copper are laid down in almost all organs and tissues, including the ectodermal structures. Enlargement of the liver, associated with a fine type of biliary cirrhosis occurs, and also an interstitial fibrosis of the pancreas. In consequence of these changes, a clinical picture is produced of diabetes mellitus, accompanied by a slaty-blue discolouration of the skin, prompting the appellation, "diabète bronzé."

The pigmentation prefers certain skin areas, such as those exposed to light. The mucous membranes are usually said to be uncoloured. In a search of the available literature I have been unable to find any reference to retinal pigmentation. In the course of a study of four examples of this disease from another point of view, the fundi were carefully examined through the dilated pupil. In all four patients, retinal pigmentary changes of varying degree were discovered, of a special character now to be described.

The pigment deposition was of a diffuse and uniform character, but was especially concentrated around some or all of the disc margin. The edge of the disc, therefore, appeared exceptionally sharply cut, owing to the colour contrast. This intensity of darkness was maintained through a radius of 3 or 4 mm. towards the periphery, and then commenced to fade and assume the normal hue of a "tigroid" fundus. The pigment itself was not the common jet black colour seen in dark skinned races or brunettes, but a deep slaty-blue colour, through which the retina appeared as a kind of ochre background. The artist (see Fig.) has not quite caught the exact shade, and has made the disc itself too purplish, whereas it really displayed its normal light pink. The illustration is from the patient with these changes least advanced, as he made the best "sitter."

These appearances varied directly in depth as the skin changes, as far as this small series of patients is concerned, of whom none had any special degree of natural pigmentation. In fact, the one (a woman) with the darkest fundus had the lightest shade of hair. None had any great degree of refractive error.

Of the nature of the pigment, one can only surmise that it is haemofuscin, identical with that of the cutis externa, since, so far, I have not been able to obtain autopsy material. The mucous mem-

branes usually remain uncoloured, while the ectodermal tissues, skin and retina are pigmented. It is highly probable that the rare unpigmented examples of haemochromatosis have retinæ of normal aspect.

As regards the remaining features of the fundus, a little scattered irregularity of the arterial lumen was apparent in one patient. None of the other components of a diabetic retinitis were seen in these or other reported sufferers. The carbohydrate disturbance is here of a different order from that of idiopathic diabetes mellitus. The disorder is usually diagnosed at a late stage, and milder cases can readily pass unnoticed under the clinician's eyes, especially in a sunny country, so it may happen that the oculist may be able to add one more to the list of diseases first recognised through the ophthalmoscope.

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## THE PRIMARY POSITION OF THE EYES

BY

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THE purpose of this paper is to show that the conception of the Primary Position of the Eyes, as described in text-books, is wrong.

Duke-Elder<sup>1</sup> states:

"When the eyes are looking straight in front, with the head vertical, with the visual axes parallel, and the vertical meridians of the two corneae both vertical and parallel, they are said to be in the PRIMARY POSITION."

Yet every thinking person must realise that when the eyes are looking—straight ahead or elsewhere—the visual axes are *not* parallel, but convergent. Therefore, in physiological ophthalmology it cannot be admitted that the visual axes of the *looking* eyes ever are parallel.

Perhaps no objection should be raised against this definition of the visual axes as parallel, if it is done arbitrarily in order to explain something. But then it is necessary that the description of the *true* physiological state should follow, in order to correct the wrong impression caused by so arbitrary a statement. The above text-book<sup>2</sup>, however, goes on:

"This forms the basis of the physiological definition of the primary position of the eyes."