Blindness due to a rare form of Encephalitis

Two articles in a recent number of Brain call attention to a very rare condition of the central nervous system in which occur symptoms of great interest to ophthalmic surgeons. The first article is by Dr. Bouman of Amsterdam, and the second by Drs. Collier and Greenfield, National Hospital, Queen Square. The disease with which they deal has evidently been known in a somewhat obscure way for some time, but was first defined and identified as a clinical entity by Schilder in the year 1912, who gave it the name of encephalitis periaxialis diffusa. It is allied, on the one hand, to disseminated sclerosis, and seemingly, on the other hand, to a diffuse gliomatosis. Unlike disseminated sclerosis, however, it seems to commence in one focus and not in a series of isolated foci. It spreads from its initial focus diffusely in the white matter of the centrum ovale. In the preponderance of cases, it seems to commence in the white matter of the occipital lobes, and spreads from there into the white matter of the temporal lobes, parietal lobes and frontal lobes, so that, ultimately, the whole white matter of the cerebral hemispheres may be affected. The myelin sheaths are the portions of the white matter primarily affected, and in this respect it resembles disseminated sclerosis.

The predominant feature of interest to ophthalmic surgeons is the early onset of blindness, evidently usually commencing as a hemianopia, sometimes quadrantic, sometimes altitudinal, but gradually or rapidly becoming complete, and without any ophthalmoscopic changes in the majority of cases. In only a few cases of rapid onset slight degrees of papilloedema have been observed. In one or two cases, slight ocular-motor palsies have been noted, but these have not been a common symptom. The most characteristic symptoms, apart from the blindness, are the fairly rapid development of spastic paralyses (diplegias and quadriplegias) and progressive amentia. The disease usually occurs in childhood or early adult life and is progressive to a fatal termination within two or three years, the final stages being marked by complete spasticity and amentia.

Cause of Myopia

The discussion on the causes of myopia still proceeds and seems likely to continue for long enough. In a letter to the editor of the Lancet of October 25, 1924, Eccridge-Green repeats the statements made in his Arris and Gale lecture of 1921 that myopia
is produced by dilatation of the eye through increased intraocular tension caused by back pressure through the veins of the eye. He states that if the patient is instructed to avoid such muscular effort as will cause this the myopia may remain stationary or even diminish and the eyes may in some cases become emmetropic. We do not consider that Edridge-Green has brought forward facts to justify the following statement: "The eye evidently possesses a power of contracting under diminished intraocular tension." The use of the word "evidently" implies that it is obvious to all, which it certainly is not. This is quite clear from, e.g., the opening sentence of J. A. Wilson's reply in the Lancet of November 1. "I venture to state that few, if any, ophthalmic surgeons accept Dr. Edridge-Green's theory of the production of myopia." In the following paragraphs Wilson points out that sixty per cent. of myopic scholars are girls and that myopia is and has for long been known to be commoner among girls than boys, even before gymnastics were introduced into schools. He seems to hint at the finding of a mare's nest by Edridge-Green when he remarks that "artificial myopia due to spasm of accommodation does diminish." Wilson considers that the size of the eyeball is determined by the factor of heredity, though there are exceptions such as myopia due to keratitis. In this last case Wilson suggests that toxins pass through the cornea and filter back along the coats of the eyeball causing softening, and later, stretching at the corneo-scleral junction or at the posterior segment of the eyeball. J. L. Dick follows up the correspondence in the Lancet for November 29. He considers that myopia is essentially associated with debilitated states of the individual and is a disease of modern civilization. It is associated with debility rather than with continuous and excessive muscular action. Dick states that the thin sclera of the young child tends to be softened as the result of debilitating disease such as measles, rickets and whooping cough. "Diminished resistance of supporting structures rather than increased strain is, I believe, the essential factor in the production of myopia."

Dick, however, entirely omits to mention the heredity factor, and we do not think that any theory of myopia can possibly hold water that does not take account of this.

The correspondence is wound up in the Lancet of December 6 by Edridge-Green who admits the conditions mentioned by Dick as "contributory factors," and re-asserts his view that increased intraocular tension is the direct exciting cause. He repeats the "bull" case mentioned in the Arris and Gale Lecture which we transcribe from the text of the lecture itself: "A man staying at a farm had to help to hold a bull which had become wild, and he had to exert his strength to the utmost; he ruptured himself, became myopic to a low degree and blind in one eye through
detachment of the retina in the one afternoon; he stated that he had previously very good sight. Though such a disastrous result is fortunately very rare, this case admirably illustrates the causation of myopia, and had the increase been very slight might easily have passed unnoticed.” Now the strength of a chain is that of its weakest link, and the weak link here is that the previous ocular condition of the patient depends solely on the patient’s own statement.

Lastly, there is an annotation in the Lancet for January 3, 1925, which we take the liberty of summarizing in which the editor, referring to the recent correspondence in the columns of his journal, draws attention to the lecture by Sir Arthur Keith recently delivered to the British Optical Association. In this Sir Arthur compared the scleral structure of a baby’s eye to that of the outer cover of a pneumatic tyre; the strands of fibrous tissue are so arranged as to stand a much greater strain than they are expected to receive from the intraocular pressure. But the eye must grow. It grows by the action of fibroblasts which can bring about the required processes of growth and can control the whole process by which the eyeball is shaped so as to withstand the internal pressure to which the eyeball is subjected. We do not know how the fibroblasts do their work, but after youth has passed their activity ceases. Sir Arthur suggested that just as in acromegaly the activity of the fibroblasts is entirely thrown out of gear by some defect in the secretion of the pituitary body, so some comparable fault in the mechanism which normally regulates the growth of the sclerotic coat of the eye must be at the root of myopia. Excessive convergence, vascular engorgement, unusual muscular effort are none of them adequate to originate myopia, though when once started all these may be adverse factors favouring its increase, a fact which justifies the advice usually given to myopes by the ophthalmic surgeon. What exactly causes the defective action of the fibroblasts is a problem as yet unsolved. The editor concludes: “Sir Arthur Keith has pointed out the direction in which we must look in order to solve it.”