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THE PROGNOSIS IN PAPILLOEDEMA*

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PAPILLOEDEMA is one of the common conditions which is a meeting ground of the ophthalmologist and the neurologist, and for academic as well as practical reasons concerns the practice of both, and no aspect of this condition demands more attention than its prognosis as regards the preservation of vision.

I am aware that in speaking to you to-day on the *Prognosis in Papilloedema* I am dealing with a subject which has been repeatedly discussed by this Society; in our *Transactions* you can find noteworthy contributions to it, particularly by Dr. James Taylor and Mr. Leslie Paton. Mr. Paton's articles on Papilloedema are in fact so comprehensive and based on such extensive experience that I fear I can add little to the conclusions he has already drawn. My excuse for introducing the subject again is that I frequently find that the presence of papilloedema is regarded as an immediate danger to vision. For instance, recently in an examination for a higher medical diploma I presented to several candidates a case showing early swelling of the discs, and was

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told by almost all of them that steps should be at once taken in order to save vision. There is obviously, therefore, considerable misapprehension on the matter of prognosis among our colleagues in different branches of our profession, and it is on us the duty of instruction rests.

It is my intention to discuss to-day only that form of papilloedema associated with increased intracranial pressure and to exclude the question of its diagnosis. Its pathology need not delay us, for it is now recognised by everyone that it is essentially an oedema of the nerve head in which, for a time at least, there is little structural damage or functional disturbance of the nervous elements, though these may be strangled and destroyed by overgrowth of interstitial tissue when the oedema subsides into atrophy. Unfortunately we must also recognise that in this stage no recovery of vision can be expected.

As the essential aetiological factor in papilloedema is increase in intracranial pressure, the relief of this pressure will lead to its disappearance and remove the risk of blindness or serious deterioration of vision if it can be effected before secondary changes have developed in the disc. It is therefore natural that those who are primarily concerned in the conservation of the patient's sight should demand early or immediate operation, for it is only by operation that permanent relief of pressure can be obtained. But there are often cogent reasons against immediate operation. Complete removal of the tumour, which is the condition with which papilloedema is most often associated, is frequently impossible, more especially as it is in cases with extensive infiltrating tumours of the glioma type that the severer degrees of papilloedema are found, and in these cases operation frequently reduces the length and utility of life. Even a cerebral decompression often leads to considerable physical disablement. Further, in even the most experienced hands operation, whether radical or palliative, entails considerable risk to life, and for this reason or on other grounds, as the lack of symptoms that can establish a definite localisation, delay is often advisable.

We are consequently often faced with the questions: What will happen if pressure is not relieved at once? and, How long can we delay without endangering the patient's vision? To these questions no general answer can be given; each case must be considered on its own merits. But the statement in one of the most recent textbooks on neurology that "every longstanding case of papilloedema inevitably leads to blindness" cannot be accepted without qualification.

It is of course known to every ophthalmologist that even intense papilloedema may not cause any disturbance of vision; Hughlings Jackson recognised this a few years after the introduction

of the ophthalmoscope and emphasised it again in his Presidential address to this Society 48 years ago. It is not, however, so generally recognised that vision may remain unimpaired for long periods, that in other words the papilloedema may be non-progressive. It is a frequent experience of neurologists that even in the presence of considerable congestion and swelling of the optic discs there may be no deterioration of sight for months, and occasionally we meet with cases in which vision remains unchanged for years. In the earlier volumes of our *Transactions* such cases have been described by Hartridge, Marcus Gunn, Frost and others, and Gowers, an unrivalled observer, stated in his *Medical Ophthalmoscopy*, "The time taken for the development of changes varies within wide limits. A neuritis may remain for months or even for years unchanged, or the most intense strangulation may be developed in a few weeks," and on another page he wrote "at any stage the morbid process may stop, remain stationary for a long time and then recede."

I have seen many such cases in which the existence of a papilloedema was beyond doubt and yet vision remain unimpaired for long periods. One man, for instance, whom I was able to follow for six or seven years, preserved normal visual acuity and unrestricted visual fields. It is true such cases are exceptional, but it should be recognised that they do occur. In these cases there are probably either quiescent or slowly growing tumours to the presence of which the intracranial contents can adapt itself so that there is no great or sudden rise of pressure.

In other, but rarer, cases the papilloedema subsides without surgical intervention or other specific treatment, and vision remains unaffected. Hughlings Jackson described several such cases and Gowers wrote many years ago, "now and then a neuritis rapidly subsides although the cerebral disease progresses." In a later volume of our *Transactions* Mr. Maitland Ramsay referred to "a considerable number of cases in which, in spite of intense oedema, the condition did subside, vision was unimpaired and no operation became necessary." I have personally followed several such cases. In most of them it is true the swelling of the discs was low, and hæmorrhages and exudates were absent or scanty, but this is not always so. One of the most striking instances was a doctor's wife in whom several expert ophthalmologists had no doubt that papilloedema with considerable swelling and hæmorrhages existed. Its association with headache suggested the presence of an intracranial tumour, though she showed no other signs of cerebral disease. Though no operation was undertaken the papilloedema subsided slowly, vision remained intact and there has been no recurrence or the development of other symptoms during the past four years.

In most of these cases it is impossible to ascertain the exact nature of the condition to which the papilloedema is due; in some there may be a tuberculous or gummatous tumour which becomes quiescent, or a gliomatous growth which undergoes degeneration, but internal hydrocephalus is probably responsible in many of them for the elevation of intracranial pressure, and in others there may be an external hydrocephalus or "serous meningitis," that is an excess of cerebrospinal fluid in the sulci and on the surface of the brain, such as is frequently found in association with otitis and other infective processes in the skull.

It is true that spontaneous subsidence of papilloedema is rare and that non-progressive papilloedema is not common, but their occurrence and the fact that swelling and congestion of the discs may persist for long periods without serious deterioration of vision, shows that papilloedema as such does not always demand immediate surgical intervention.

In most cases of papilloedema, however, the relief of intracranial pressure is, or soon becomes, necessary to save sight, and we are consequently faced with the important question: what indications are there?, or what warning may we have, that further delay is dangerous? In the first place there can be no doubt that the more rapidly the congestion and swelling of the optic discs develop, the more intense does the papilloedema become and the greater is the danger to sight if the intracranial pressure is not relieved. Swelling of five or more dioptries is usually a danger-signal against undue delay. Intense congestion, as shown by great engorgement of the retinal veins and early and extensive hæmorrhages on the surface of the swollen disc, are equally serious features, in fact they may be regarded as the most important measure of the severity of the process. The early appearance of exudates in the form of large soft greyish patches on the swollen discs, or of a macular fan, are also significant signs, though they may not themselves interfere with vision. An even more important danger-signal is, as Mr. Paton and others have pointed out, a narrowing or decrease in calibre of the arteries in the swollen disc and its neighbourhood, in fact when the narrowing is pronounced it is improbable that vision will remain unimpaired even though cerebral decompression is at once undertaken. Contraction of the arteries may be in fact regarded as the first evidence of constriction of the structures in the nerve-head by the organising fibrous and glial tissue, and irreparable damage of the nerve fibres soon follows. The appearance of white lines along the vessels usually belongs to a later stage of consecutive atrophy.

In the later stages increasing greyness and density of the swollen disc, a subsidence of the swelling accompanied by even the slightest deterioration of visual acuity, or commencing con-

striction of the visual fields, are indications that in the near future vision may be affected by the histological changes associated with consecutive atrophy. Of these signs of an oncoming atrophy special importance must be given to peripheral contraction of the visual fields; it is often the most prominent of the symptoms associated with early atrophy and frequently the first indication of secondary changes.

To you it is unnecessary to emphasise the importance of frequent and careful observations to follow the evolution of the fundal changes and to detect the earliest signs of secondary atrophy which constitute such a serious threat to vision.

Another symptom which is generally regarded as a warning that vision is in danger is the transient loss of sight of which many patients complain. It may be only momentary, but occasionally it *lasts* for a few minutes, or on rare occasions it may pass off even more slowly. It has been attributed by some to a temporary increase of congestion, and by others to a temporary failure of circulation in the retinae and optic nerves, and the fact that it is often brought on by a sudden change in posture, as rising quickly from the prone or sitting position, favours the latter view. I agree, however, with Mr. Paton that it more probably results from compression of the chiasma and optic tracts by the bulging or distended floor of the third ventricle. It certainly occurs more commonly with tumours in the posterior fossa of the skull, which almost invariably lead to internal hydrocephalus, than with growths in other regions. The fact that it occasionally precedes the appearance of pathological changes in the discs or apart from them, as Dr. Hughlings Jackson and Mr. Paton have observed, an observation that I have been able to confirm, indicates that it is not directly due to papilloedema though often associated with it. When, however, this temporary amaurosis occurs in a patient with highly swollen discs it must be regarded as a threat of permanent failure of vision, as the intracranial as well as the intra-ocular changes with which it occurs, are usually so intense that sight may be soon endangered.

The treatment of papilloedema is of course removal of its cause, which is increased intracranial pressure, either by extirpation of the tumour or other pathological condition which raises pressure, or when this is not practicable, by a decompression craniectomy. As a rule the swelling and congestion of the discs subside within two to four weeks after operation, and when secondary changes had not appeared by then there is no further risk of impairment of vision. When visual acuity is seriously reduced before operation it is unfortunately rare for appreciable recovery to take place. When, however, the impairment of acuity is slight vision may improve considerably; I may recall to you that Mr. Paton observed

such improvement in one-third of a series of cases which he followed after operation, but my own experience has not been so fortunate. Visual fields that have become seriously constricted rarely recover fully, though a moderate degree of peripheral narrowing often disappears as the papilloedema subsides.

Occasionally there is immediately after operation a rapid deterioration of sight, which is, however, often transient; it is probably due to a sudden fall of pressure in the central artery of the retina as a result of too rapid relief of intracranial tension, and is therefore comparable to the diminution of vision which sometimes follows a successful operation for glaucoma.

Unfortunately vision may also fail permanently after a decompression operation in patients in whom neither the ophthalmoscopic changes nor the state of the visual fields had suggested the risk of such a catastrophe. This, in my experience, has been most common after either partial or complete removal of a tumour from the posterior fossa of the skull. For instance, I recently saw a woman with good visual acuity and unrestricted visual fields and a degree of papilloedema which could not be regarded as severe before operation, who, a few days after the partial removal of an acoustic nerve tumour and an extensive decompression, complained that her sight was failing and within a week or two was permanently blind. An equally tragic case was under my care recently. Visual acuity was 6/9 in each eye and the visual fields were full, and though the swelling of each disc amounted to about 5 dioptres the ophthalmoscope did not reveal any changes suggestive of impending atrophy. A fortnight after complete removal of a large dural endothelioma which compressed his left frontal lobe he complained of impairment of vision which progressed to complete blindness of one eye and left 6/36 vision only in the other. The swelling of the discs subsided slowly into a characteristic secondary atrophy.

Such progressive failure of vision after operation in cases in which it had not been expected or foretold is naturally distressing to all concerned, particularly when the cerebral operation promises permanent relief, but fortunately it is not common. The factors which determine the rapid secondary atrophy to which it is due are uncertain, and I fear I cannot suggest any signs which may warn us of its possible occurrence.

But an accurate prognosis of the state of vision after relief of intracranial pressure can usually be given by careful and if necessary repeated observations, though, as much else in medicine, a reliable judgment depends largely on experience, and as a guide to it we are often unable to formulate definite rules.