Light at the end of the tunnel for the blind leading the blind?

Editorial

The September bulletin of the British Paediatric Surveillance Unit (BPSU) stated that the Sir Peter Tizard research bursary has been awarded to a proposed epidemiological survey of paediatric idiopathic intracranial hypertension (IIH), also known as benign intracranial hypertension or pseudotumour celebri. IIH can lead to visual failure but is treatable. Its diagnosis, natural history, and medical and surgical management in childhood remain controversial.[1,2]

Paediatric neurologists, ophthalmologists, and neurosurgeons agree that it is a syndrome defined by a triad of features: raised cerebrospinal fluid (CSF) pressure in the absence of an intracranial mass lesion or dilatation of the cerebral ventricles; normal spinal fluid composition; and normal neurological findings, with the exception of papilloedema and sixth cranial nerve palsies.

Headache or visual impairment, alone or together, are cardinal symptoms (IIH is sometimes regarded as a 'headache syndrome') but some patients are asymptomatic. Transient visual obscurations are common, but only if specific enquiry is made.[1] Symptoms, signs, and CSF pressure do not necessarily vary in phase with each other and the condition is associated with a number of headache disorders not caused by IIH.[3] Causal associations with IIH include recurrent otitis media, being overweight, iron deficiency, marasmus, over- or undersupply of vitamins, endocrine factors, and certain drugs. By convention, the term IIH is used to denote cases that are both truly idiopathic and for which a probable causal association has been identified, while frank cerebral venous thrombosis is excluded. Little is known about its pathophysiology.

Underdiagnosis may result from false reassurance by a normal 'resting' CSF pressure or from using reference ranges for pressure that fail to take account of age, but is less common than overdiagnosis. The diagnosis usually rests on interpretation of fundoscopy, CSF opening pressure at lumbar puncture, and on a clinical judgement on whether symptoms, fundoscopic appearance, and CSF pressure are causally related. Caution is needed, particularly in asymptomatic children with abnormal optic discs or in any child with normal fundoscopy.

Drusen, (literally 'hidden') glial tissue within the retina unrelated to intracranial pressure but presenting the fundoscopic appearance of 'swollen' discs, can be diagnosed with the help of, in increasing order of sensitivity, calcification on CT, auto-fluorescence, or increased reflectivity on ultrasound 'B' scanning of the disc. Sometimes fundoscopy of the parents can provide helpful evidence of familial drusen. Papillitis is often not considered as an alternative diagnosis nor optic neuritis as a cause of visual failure in children with modestly elevated CSF pressure and normal discs. Although young children presenting with visual failure and without headache feature in some series of children with IIH, this, as with brain tumours, often reflects the difficulty in obtaining a history from the very young. For the same reason, young children with optic neuritis rarely give a history of eye pain and, if they do, it may sound like headache. The signs that enable optic neuritis to be differentiated from IIH may also be lacking, as affected young children are seldom able to cooperate for Goldman perimetry. Thus, although ophthalmologists may be correct in asserting that 'visual evoked response testing does not have a role in the detection or monitoring of visual dysfunction in IIH',[1] it is a test that may lend valuable support to a clinical suspicion of optic neuritis.

On this care pathway, abnormal fundoscopy often prompts cranial imaging. In an ideal world this will include magnetic resonance (MR) venography and also sequences that are sensitive to white matter disorders, but IIH may only be considered for the first time after exclusion of a mass lesion on imaging that is inadequate to exclude venous thrombosis or optic neuritis. Young macrocephalic children with normal optic discs, especially those with macrocephalic parents, whose scans show plump ventricles and large extra-cerebral spaces, do not have IIH and should not be exposed to the risks of its treatment. Most have benign external hydrocephalus (BEH) and are asymptomatic (with elevated CSF pressures relative to population norms).

CSF composition, including cell count and levels of protein and glucose, must be normal. CSF opening pressure is liable to elevation by anxiety, distress, hyperflexion for lumbar puncture, breath-holding, over-sedation, or general anaesthesia. Reference pressure ranges that take account of being overweight are available for adults but not for children. Conditions other than IIH may lead to swollen discs and elevated CSF pressure, notably optic neuritis. Thus, for every child that truly has IIH, I typically see four or five other children without IIH but with a CSF pressure measurement between 17 and 27cm of CSF. These include healthy children, those with optic neuritis, and those with BEH.

If the evidence base for management of IIH were ice, I would not walk on it, even if I were a duck, and I will not air my views here. We need to reach a sufficient consensus to start the long journey towards a randomized controlled trial of alternative treatments. Incidence figures for paediatric IIH have been hamstrung by diagnostic uncertainty, but the relative rarity of IIH clearly warrants a multi-centre approach. Clear case definition will, therefore, be significant for epidemiological survey and management.

Light in this murky area of clinical practice is badly needed. I wish the BPSU-backed investigators success in achieving consensus. The starting point must be a precise algorithm for diagnosis.

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References

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