EDITORIAL

Ophthalmic complications of spina bifida and hydrocephalus

Ophthalmic involvement is well known to occur in neurodevelopmental problems, but it is not so widely appreciated how frequent this is or its relevance to management. This is highlighted in this issue by Hannah Gaston who over a six-year period examined 322 children with spina bifida and hydrocephalus. She found a wide range of neuro-ophthalmic involvement including: strabismus, nystagmus, refractive errors, sunsetting, optic atrophy, papilloedema, Parinaud's syndrome, cortical visual impairment, visual field defects and visual failure due to shunt blockage.

Strabismus was seen in 42% and of these 33% were only identified by the ophthalmologist. Most were convergent, with over half exhibiting signs of a 6th nerve pasly. Optic atrophy was observed in 17%, and although it was often long standing, being associated with the underlying cause of the hydrocephalus, it could also signify recent shunt failure. Primary optic atrophy is not a safe diagnosis and shunt function should always be investigated. The low incidence of cortical visual impairment in this study (3%) is perhaps surprising, recognising how difficult this is to diagnose, particularly in the presence of pre-existing anterior visual pathway dysfunction. Less than one-third had completely normal visual functions and even 13% of these had exhibited transient eye problems. Severe visual impairment was invariably accompanied by severe general handicap.

Papilloedema was generally considered to be infrequent in congenital hydrocephalus, but two recent studies have reported incidences of around 14%. This sign was invaluable in diagnosing shunt malfunction and raised intracranial pressure, which otherwise would have been made on the basis of non-specific signs. Of course the absence of papilloedema is not an assurance of normal intracranial pressure. Most important, in those children examined during a period of uncontrolled hydrocephalus, there was positive ophthalmic evidence in 70%. Also in a small group in whom clinically the shunt appeared to be working, ophthalmic signs were the only clue to malfunction which was eventually proven at surgery. This study emphasises that ophthalmic assessment in these children is worthwhile not only for long-term care, but also as part of the management of acute problems.

Alistair Fielder