

## Clinical details of six children studied

Sex	Year of birth	Ophthalmological diagnosis	Period of normal development (years)	Perceived environmental stress factors
M	1974	Anophthalmia	3-3.5	Hospitalisation
M	1975	Hypoplastic retinal membrane	2-3.25	Separation from parents
F	1979	Retinal aplasia	2-2.5	None recognised
M	1979	Optic nerve hypoplasia*	2.5-3	Nursery placement, birth of brother
M	1982	Optic nerve hypoplasia*	2.5	Nursery placement
M	1986	Ocular haemangioma	2-2.5	None recognised

\*Light perception present. All six are now severely retarded with overt autistic behaviour.

professionals who are concerned with assessing and attempting to predict the developmental outlook for children with severe visual handicap.<sup>1</sup>

The authors make clear that it is those children with the most severe degrees of visual impairment who are at greatest risk of sustaining developmental setbacks and suggest that component causative factors include primary maldevelopment of the central nervous system, the degree of visual impairment, the developmental stages that affected children have reached and the developmental and emotional environments to which they are subjected.

The developmental profiles of some of the children they describe are reminiscent of children who show the features of disintegrative psychosis in childhood,<sup>2,3</sup> in which it may be that inherently vulnerable children develop a severe acquired autistic syndrome often in association with evidence of severe environmental stress. A similar picture can be seen in children with severe visual impairment and over recent years I have followed up the progress of six children all of whom had severe visual impairment and who have shown evidence of severe and permanent developmental setbacks. Their clinical details are summarised in the table.

Clearly the experiences of Cass and her colleagues are likely to represent not uncommon phenomena. I would endorse their recommendations that careful attention be given to the developmental and environmental vulnerability of this group of handicapped children, particularly as the developmental setbacks that they describe are likely to cause major and long term disability.

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- 1 Cass H, Sonksen P, McConachie H. Developmental setback in severe visual impairment. *Arch Dis Child* 1994; 70: 192-6.
- 2 Evans-Jones LG, Rosenbloom L. Disintegrative psychosis in childhood. *Dev Med Child Neurol* 1978; 20: 462-70.
- 3 Hill AE, Rosenbloom L. Disintegrative psychosis of childhood: teenage follow-up. *Dev Med Child Neurol* 1986; 28: 34-40.

## Epilepsy with myoclonic absences

EDITOR.—The paper by Drs Manonmani and Wallace has, very succinctly, described an under-recognised epilepsy syndrome.<sup>1</sup> It is possible that some of the intellectual difficulties noted in their children may have been

related to subclinical epileptiform discharges as has been reported previously<sup>2</sup> (and continues to be seen in my experience) in this type of epilepsy. The improvement in seizure control is frequently mirrored by an improvement in attentional skills (and therefore 'learning'), and by disappearance of the subclinical epileptiform discharges.

The identification of specific epilepsy syndromes is important for both pragmatic and academic reasons; it provides information on prognosis and guidelines for antiepileptic treatment (that is, which drug should be used). Lamotrigine does appear to be effective in epilepsy with myoclonic absences.<sup>1,2</sup> However, the drug may be associated with an idiosyncratic or hypersensitive (allergic) rash, particularly if 'added' to sodium valproate, which may militate against its continued use. Lamotrigine must be introduced slowly; a recommended starting dose is  $\leq 0.5$  mg/kg/day, initially on alternate days, increasing to 2-5 mg/kg/day over 5-6 weeks - failure to do so may limit the usefulness of this new antiepileptic drug.

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- 1 Manonmani V, Wallace SJ. Epilepsy with myoclonic absences. *Arch Dis Child* 1994; 70: 288-90.
- 2 Gibbs J, Appleton RE, Rosenbloom L, Yuen WC. Lamotrigine for intractable childhood epilepsy: a preliminary communication. *Dev Med Child Neurol* 1992; 34: 369-71.

## Ambulatory paediatrics

EDITOR.—The paper by Doug Heller gave an excellent overview of the philosophy of ambulatory paediatrics.<sup>1</sup> I agree that it is a change in attitude, making the service more child and family oriented, rather than a new speciality.

The definition of ambulatory paediatrics is broad, and many people find the concept a little abstract. I would like to give concrete examples of two successful developments within our ambulatory service.

A daily urgent consultant led clinic has been introduced. Many similar clinics suffer from being overcrowded by inappropriate referrals. We have avoided this by introducing a 'hotline', which is a direct line manned by a consultant and runs for one hour before the urgent clinic. General practitioners can phone for advice and to discuss patients. Those thought to need an urgent appointment are seen that day. Preliminary data suggest that

less than half the calls to the hotline result in an urgent appointment, and many problems can be dealt with by advice only.

The second development concerns our children's home care nurses who rotate through the accident and emergency department. The aim was to improve the quality of care for children in the accident and emergency department and to reduce short stay admissions. The initiative was initially supported by and is currently being evaluated by the King's Fund. Preliminary data suggest this has been very successful, and we are committed to continuing the service.

Ambulatory paediatrics is an exciting and challenging area to be involved in, with much scope for development. Even small changes in attitude and in the way the service is delivered can make a huge difference to the quality of care provided.

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- 1 Heller DR. Ambulatory paediatrics: stepping out in a new direction? *Arch Dis Child* 1994; 70: 339-42.

## Role of ultrasound in congenital hip dysplasia

EDITOR.—We were pleased that the publication of the annotation on this subject in the April issue gave prominence to the potential of this approach to the management of babies with clinically suspected hip instability.<sup>1</sup> We are concerned, however, that some readers might wrongly conclude that ultrasound screening for these babies is an established strategy for selecting which of them actually need early prophylactic splinting. This is a technique that, though promising,<sup>2</sup> still needs further controlled trial evaluation. We need to confirm that the potential advantages really do outweigh the disadvantages when this approach is adopted in everyday practice. As the annotation says, it remains 'important that prospective trials provide data to ensure that errors in diagnosis and subsequent treatment are not compounded'. The MRC have just funded such a trial. Anyone in a position to help recruit patients is urged to contact Lesley Morgan at the Perinatal Trials Service in Oxford (where the trial is being coordinated) for further information.

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- 2 Gardiner HM, Dunn PM. Controlled trial of immediate splinting versus ultrasound surveillance in congenitally dislocated hips. *Lancet* 1990; 336: 1553-6.

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