Clinical details of six children studied

<table>
<thead>
<tr>
<th>Year of birth</th>
<th>Ophthalmological diagnosis</th>
<th>Period of normal development (years)</th>
<th>Perceived environmental stress factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>M 1974</td>
<td>Anophthalma</td>
<td>3–5</td>
<td>Hospitalisation</td>
</tr>
<tr>
<td>M 1975</td>
<td>Hypoplastic retinal membrane</td>
<td>2–3–25</td>
<td>Separation from parents</td>
</tr>
<tr>
<td>F 1979</td>
<td>Retinal aplasia</td>
<td>2–5</td>
<td>None recognised</td>
</tr>
<tr>
<td>M 1979</td>
<td>Optic nerve hypoplasia*</td>
<td>2–5</td>
<td>Nursery placement, birth of brother</td>
</tr>
<tr>
<td>M 1982</td>
<td>Optic nerve hypoplasia*</td>
<td>2–5</td>
<td>Nursery placement</td>
</tr>
<tr>
<td>M 1986</td>
<td>Ocular haemangiomatosis</td>
<td>2–25</td>
<td>Note recognised</td>
</tr>
</tbody>
</table>

*A 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.

The authors make clear that it is those children with the most severe degrees 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, in which it may be that inherently vulnerable children develop a severe acquired autistic syndrome in association with evidence of severe environmental stress. A similar picture can be seen in children with severe visual impairment and in 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 lasting disability.

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**Epilepsy with myoclonic absences**

**EDITOR.—**The paper by Drs Manonmani and Wallace has, very succinctly, described an under-recognised epilepsy syndrome. 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 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. However, the drug may be associated with an idiosyncratic or hypersensitive (allergic) rash, particularly if ‘added’ to sodium valproate, which may mitigate against its continued use. Lamotrigine must be introduced slowly; a recommended starting dose is ≤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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**Ambulatory paediatrics**

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

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.

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. We were concerned, however, that some 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, 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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Role of ultrasound in congenital hip dysplasia.

C Dezateux and E Hey

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