LETTERS TO THE EDITOR

Aetiology of chronic suppurative lung disease

EDITOR,—Nikolaizik and Warner report that eight of 41 children with suppurative lung disease had a demonstrable primary immunodeficiency.¹ Four of these patients were shown to have abnormalities of neutrophil function on the basis of some form of in vitro functional assay. Three further patients with other causes for their lung disease were said to have similar defects.

None of these deficiencies are recognised within the comprehensive World Health Organisation classification of immunodeficiency. This is probably because such functional abnormalities have proved difficult to characterise in a consistent manner. The possibility exists that the observations in these assays are due to immunological activation secondary to chronic infection.

I am surprised at the relative absence of immunoglobulin and antibody deficiencies in this series. These are the most common primary immunodeficiencies and are well characterised as leading to suppurative lung disease. A proportion of the 37% of children without identifiable predisposing factors might have such deficiencies if they had detailed investigation of their immunoglobulin and antibody production. The methods section does not outline the extent of the immunological evaluation or whether all patients received such an evaluation. The identification of such patients is important as replacement immunoglobulin treatment may be the only way of preventing progression of their pulmonary disease.

Finally the two patients with measles and adenovirus infection and measles and disseminated *Mycobacterium intracellulare* infection are placed in the secondary immunodeficiency group. These patients may also have primary cell mediated immunodeficiencies. Though measles is immunosuppressive, a severe and complex course of measles is often associated with a primary immunodeficiency.

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1 Nikolaizik WH, Warner JO. Aetiology of chronic suppurative lung disease. *Arch Dis Child* 1994; 70: 141-2.

Professor Warner comments:

Dr Morgan makes a number of important points about our publication that certainly require answering. The presentation of a short report does rather restrict the number of references that can be given which might perhaps have answered some of the questions.

Dr Morgan is concerned that the deficiencies described are not recognised within the World Health Organisation classification of immunodeficiency. This classification is very exclusive. The criteria are very restrictive and this is rather like placing blinkers on immunologists in suggesting that significant and reproducible abnormalities of immune responsiveness are not evidence of

immunodeficiency. This is patently not the case. It is important to emphasise that none of the immune investigations were done when children had active infection. The C reactive protein was always normal at the time of investigation, otherwise the results were rejected. Furthermore, the vast majority had more than one investigation to confirm the abnormality. I would refer Dr Morgan to our publication which was reference 6 in the original paper.¹ This describes the neutrophil defects in more detail. It also provides extra evidence which might eventually allow them to be recognised within the classification system.

Sadly most of the investigation in these children was done at a time before there was a common awareness of IgG subclass deficiencies. Therefore, this was a notable shortcoming of the study and I do accept that we should perhaps have identified some immunoglobulin and antibody deficiencies. However, the presence of IgG subclass deficiency remains a very difficult area where the relevance to any disease is sometimes in doubt.² It is, however, important to note that some of the patients with serum dependent neutrophil defects responded well to immunoglobulin infusions where we suspect that the antibodies provided exert an opsonising and organism killing promoting (procidin) role.

Concern is also expressed about the presence of immunodeficiency among children with other explanations for bronchiectasis. In this respect, two of the patients with primary ciliary dyskinesia syndrome had neutrophil defects. Dr Morgan might be interested to know of some studies which have been done, suggesting that there might be primary defects of neutrophil function in patients with this syndrome which may relate to abnormalities in cytoskeletal proteins.³

The patients with measles all had their immune functions studied long after recovery from measles and the responses were normal. We certainly had no evidence of any abnormality of cellular immune response at the time that they were investigated for their suppurative lung disease. This, of course, does not totally exclude the possibility that there was a rather specific susceptibility to measles, though the severity of the measles' illnesses in each case was not outstandingly severe. The association between measles and secondary adenovirus infection is well described.⁴

- 1 Parkin JM, Bridge J, Sloper KJ, Warner JO, Pinching AJ. Defective candida opsonin and procidin in patients with recurrent pyrogenic infection - a common immunodeficiency. *Scand J Immunol* 1991; 33: 89-95.
- 2 Zielen S, Baktai G, Marialigeti T, Ahrens P, Szekely E, Hofmann D. Humoral immunity in children with proven bronchiectasis, bronchial deformations and chronic bronchiectasis. *Pediatric Allergy and Immunology* 1991; 2: 117-23.
- 3 Kantar A, Oggiano N, Giorgi RL, Fiorini R. Membrane fluidity of polymorphonuclear leucocytes from children with primary ciliary dyskinesia. *Pediatr Res* 1993; 34: 725-8.
- 4 Warner JO, Marshall WC. Crippling lung disease following measles and adenovirus infection. *British Journal of Diseases of the Chest* 1976; 70: 89-94.

Sedation for investigations: prolonged effect of chloral and trimeprazine

EDITOR,—Sedation is increasingly required for procedures such as computed tomography which are not painful but require the child to remain still during the investigation. Young children in particular may need to be

heavily sedated to achieve satisfactory results. We have performed a survey of the use of sedation for investigations in our paediatric department. This included telephone follow up of children after discharge.

Data were obtained for 34 children, median age 2.8 years. Procedures included 17 computed tomograms and 12 auditory evoked response studies. The drugs most commonly prescribed were chloral (31 patients, median dose 50 mg/kg) and trimeprazine (32 patients, median dose 2.9 mg/kg). Seventeen procedures were recorded as easy to perform, 12 were performed with difficulty, and five failed. Ward staff commented on problems relating to sedative medication in 18 children. Two were too drowsy for discharge from the day case unit and required admission overnight. We also audited the duration of starvation before sedation and the use of monitoring. In general these fell below standards that would be considered safe in children having general anaesthesia, despite the use of heavy sedation.

Parents of 24 children were contacted to ask about their child's behaviour after discharge. We were particularly concerned to determine how long it took to return to a normal pattern of behaviour. The median length of time from sedation to normality was 21 hours 30 minutes (range 90 minutes to 93 hours 50 minutes). Thirteen parents commented that their child was drowsy on the next day. One child fell asleep in school and was sent home and another was too drowsy to attend school. Parents commented that their children acted as if drunk, with dizziness and unsteady gait.

Chloral has been widely used for the sedation of children for painless procedures.¹ The main emphasis in published studies has been on the ability to perform a successful investigation.² The use of monitoring and need for starvation before heavy sedation has been addressed by the American Academy of Pediatrics.³ Similar standards are recommended as for general anaesthesia.

Anaesthetists should have a central role with paediatricians and radiologists in the development of sedation protocols for children. We have found that the use of chloral and trimeprazine for sedation of children is associated with prolonged drowsiness. Standards of monitoring, starvation, and postsedation care in children requiring heavy sedation should be similar for those having general anaesthesia.

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- 1 Keeter S, Benator RM, Weinberg SM, Hartenberg MA. Sedation in pediatric CT: national survey of current practice. *Radiology* 1990; 175: 745-52.
- 2 Hubbard AM, Markowitz RI, Kimmel B, Kroger M, Bartko MB. Sedation of pediatric patients undergoing CT and MRI. *J Comput Assist Tomogr* 1992; 16: 3-6.
- 3 American Academy of Pediatrics Committee on Drugs. Guidelines for monitoring and management of pediatric patients during and after sedation for diagnostic and therapeutic procedures. *Pediatrics* 1992; 89: 1110-5.

Developmental setback in severe visual impairment

EDITOR,—The data presented by Dr Cass and her colleagues are of importance and significance for paediatricians and other

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.¹

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,^{2,3} 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.¹ 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 previously² (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.^{1,2} 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 ≤ 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.¹ 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.

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.¹ 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,² 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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- 1 Clarke NMP. Role of ultrasound in congenital hip dysplasia. *Arch Dis Child* 1994; 70: 362-3.
- 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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