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