**LETTERS TO THE EDITOR**

Aetiology of chronic suppurative lung disease

**EDITOR,—**Nikolazik and Warner report that eight of 41 children with suppurative lung disease had a demonstrable primary immunodeficiency. 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 immunodeficiency and may be immunosuppressive, a severe and complex course of measles is often associated with a primary immunodeficiency.

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**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. 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 interpretation of results 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 causes 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 two 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 might argue against this. The association between measles and secondary adenovirus infection is well described.4


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Sedation for investigations: prolonged effect of chloral and trimipramine

**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 trimipramine (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 noted for trivialisation of 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.

Children were therefore asked 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 return to normal 90 minutes (range 90 minutes to 3 hours). Thirteen parents commented that their child was drowsy on the next day. One child fell asleep at school and the other was too drowsy to attend school. Parents commented that their children acted as if drunk, with dizziness and unsteadiness.

Chloral has been widely used for the sedation of children for painless procedures. The main emphasis in published studies has been on the ability to perform a successful investigation.2 The use of monitoring and sedation for children's imaging 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 trimipramine for sedation of children is associated with prolonged drowsiness. Standardisation of monitoring and postsedation care in children requiring heavy sedation should be similar for those having general anaesthesia.

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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 likely to be in the developmental
stages that affected children have reached and the
developmental and emotional environments
with 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,\(^2\) 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 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.

<table>
<thead>
<tr>
<th>Sex</th>
<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</td>
<td>1974</td>
<td>Anophthalmia</td>
<td>3-5</td>
<td>Hospitalisation</td>
</tr>
<tr>
<td>M</td>
<td>1975</td>
<td>Hypoplastic retinal membrane</td>
<td>2-3-25</td>
<td>Separation from parents</td>
</tr>
<tr>
<td>F</td>
<td>1979</td>
<td>Retinal aplasia</td>
<td>2-2-5</td>
<td>None recognised</td>
</tr>
<tr>
<td>M</td>
<td>1979</td>
<td>Optic nerve hypoplasia*</td>
<td>2-5</td>
<td>Nursery placement, birth of brother</td>
</tr>
<tr>
<td>M</td>
<td>1982</td>
<td>Optic nerve hypoplasia*</td>
<td>2-5</td>
<td>Nursery placement</td>
</tr>
<tr>
<td>M</td>
<td>1986</td>
<td>Ocular haemangioma</td>
<td>2-2-5</td>
<td>Note recognised</td>
</tr>
</tbody>
</table>

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

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 emergen-
dy department and to reduce short stay admissions. The initiative was initially sup-
ported 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.

We are concerned, however, that the 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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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 prospective studies 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
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.\(^3\) 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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EDMUND HEY
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National Perinatal Epidemiology Unit,
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2 Gardiner HM, Duns PM. Controlled trial of immediate splinting versus ultrasound surveil-


*Membership: Dr Rosemary Arthur, Mr Nicholas
Clarke, Dr Carol Dezaubeux, Dr Diana Elbourne, Dr Adrian Grant, Dr Alastair Gray, Dr Edmund Hey,
Ms Lesley Morgan. Professor Charles Normand.
Developmental setback in severe visual impairment.

L Rosenbloom

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Updated information and services can be found at:
http://adc.bmj.com/content/71/2/179.3.citation

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