Aetiology of chronic suppurative lung disease

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

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 varied and was not out of the normal. The association between measles and secondary adenosine infection is well described.4

GARETH MORGAN


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 queries.

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 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 nature of the defect 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 of suppurative 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

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Sedation for investigations: prolonged effect of chloral and trimeprazine.

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