Authors' response to 'TAP deficiency is also a cause of bronchiectasis'

We thank Zimmer *et al* for their comments¹ on our recent article 'Immunity and bacterial colonisation in bronchiectasis'.²

We accept that indirect activation of natural killer (NK) cells by bacterial pathogens has been well described.^{3–4} We also recognise that a role for NK cells may be suggested by the link between patients with familial bronchiectasis with impaired human leucocyte antigen (HLA) class I expression and as a result NK cell dysfunction.⁵ We, therefore, agree that there is merit in discussing the potential role of NK cells in the pathogenesis of non-cystic bronchiectasis. However, the aim of our article was to describe in some detail the functions of aspects of the immune system in bronchiectasis with particular focus on those aspects that lead to the propensity for bacterial colonisation seen in these patients. In particular, we wished to highlight the bacterial factors that may facilitate bacterial colonisation and, crucially, may impair response to antibiotic therapy with particular emphasis on *Pseudomonas aeruginosa*. As such it was not practical to include the potential role of every factor of the innate immune system that may potentially have a role in the pathogenesis of bronchiectasis.

Similarly, we acknowledge the relationship between transporter associated with antigen processing (TAP) deficiency and bronchiectasis. However, we feel that although early diagnosis of this condition (in childhood), for example, in the case of a family member of a known patient, may allow for the introduction of appropriate management of lower respiratory tract infections and thereby perhaps delay the appearance of bronchiectasis,⁶ the inclusion of tests to detect this in diagnostic algorithms as suggested would be unlikely to change the

management of these patients. In the absence of an effective definitive treatment for this rare condition, patients should be managed as per the guidelines for the treatment of non-cystic fibrosis bronchiectasis with appropriate antibiotic therapy during exacerbations, physiotherapy and antibiotic prophylaxis where indicated. Therefore, although Zimmer *et al* feel it is important to diagnose TAP deficiency as early as possible, we feel the inclusion of tests to diagnose this in diagnostic algorithms cannot be justified at this current time.

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