Review of IgG Subclass and IgA Deficiency in a Tertiary Center

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Abstract
We retrospectively reviewed the clinical characteristics of twenty patients with IgA and/or IgG subclass deficiency attending a tertiary centre. The median age at diagnosis was 1.21 years and the median duration of follow up was 3.83 years. The most common presenting symptom was recurrent sinopulmonary infections (45%). This was followed by refractory asthma (30%), autoimmunity (15%), and other atopic manifestations (10%). Only one out of twenty patients presented with chronic gastrointestinal disease, which contrasted with the majority of previous literature. Three patients had significant pulmonary complications including bronchiolitis obliterans, bronchiectasis and pulmonary fibrosis. The prevalence of IgA deficiency in Hong Kong Chinese population is yet to be established, though it appears not as prevalent as that in western population.

Key words
Chinese; IgA deficiency; IgG subclass deficiency

Introduction
Selective IgA deficiency is the most common primary immunodeficiency and the estimated prevalence ranges from 1 in 400 to 3000 from various studies. Most of the IgA-deficient patients are asymptomatic but some of them are susceptible of recurrent sinopulmonary infections, atopic manifestations and asthma, autoimmunity and gastrointestinal tract disease. It is also known that IgA deficiency is associated with ataxia telangiectasia and chromosomal abnormalities (18q syndrome or ring chromosome 18). IgA deficient individuals with anti-IgA IgG antibodies on receipt of blood products may have anaphylactoid reaction. About 15-20% of IgA-deficient patients also have IgG subclass deficiency.

Synthesis of IgG2 and IgG4 lags relative to IgG1 and IgG3 during normal development. IgG2 is the most prevalent IgG subclass deficiency in paediatric patients and it is believed that IgG2 is responsible for immune response to polysaccharides antigens, especially in the context of infection with Streptococcus pneumoniae and Haemophilus influenzae. IgG1 and IgG3 are responsible for the antibody response to protein antigens. IgG subclass deficiency may occur as an isolated single IgG subclass deficiency or in combination with other IgG subclasses. Similar to IgA deficiency, IgG subclass-deficient patients can present with recurrent sinopulmonary infections, asthma, atopy and autoimmunity.

There is a lack of epidemiological and clinical study on IgA and IgG subclass deficiency in Hong Kong. We summarised hereto our experience in managing patients with IgA and IgG subclass deficiency in a tertiary centre.

Method
Case records of all patients with IgA and/or IgG subclass deficiency attending Paediatric Immunology Clinic at Queen Mary Hospital from July 1988 to December 2005 were retrospectively reviewed. These patients were referred