Australian Paediatric Surveillance Unit

PROTOCOL SHEET - PRIMARY IMMUNODEFICIENCY DISEASE (PID)
(Including Severe Combined Immunodeficiency Syndrome, from January 1997)

Background to study

The individual forms of primary immunodeficiency diseases are rare and their true incidence is not known. The Australian Society of Clinical Immunology and Allergy (ASCIA) formed a Primary Immunodeficiency Disorder (PID) study group in 1990 and developed a register of patients with PID with the aim of collecting clinical and epidemiological data and quantifying the potential requirements for replacement and other therapy. However, the findings of the Register are incomplete. To March 1993, 500 paediatric and adult patients have been ascertained and classified according to the WHO classification of PID. Other, as yet unclassified, forms of PID have also been sought. It is hoped that information collected through APSU surveillance will provide more complete information about the incidence, type, management and outcome of PID in Australian children.

Objectives:

To determine

1. the incidence of and demographic features of PID in Australia
2. the relative occurrence of its different causes
3. its management, mortality and morbidity
4. potential national requirement for immunoglobulin therapy and bone marrow transplantation

CASE DEFINITION and REPORTING INSTRUCTIONS

Please report any child or adolescent under 15 years who has been diagnosed with a primary immunodeficiency disorder in the last month. Diagnosis should be based on clinical and laboratory findings which suggest one of the following conditions :-

1. ***Predominantly antibody defects, eg; X-linked agammaglobulinaemia, IgA deficiency, IgG subclass deficiency.***
2. ***Combined immunodeficiency, eg; severe combined immunodeficiency, common variable immunodeficiency.***
3. ***Immunodeficiency with other major defects, eg; Wiskott-Aldrich syndrome, Di George syndrome, Ataxia Telangiectasia.***
4. ***Complement deficiencies, including C1 inhibitor deficiency (heredity angioneurotic oedema).***
5. ***Defects of phagocytic function, eg chronic granulomatous disease, leucocyte adhesion deficiency, Schwachman's syndrome.***
6. ***Chronic mucocutaneous candidiasis.***
7. ***Other as yet unclassified.***

The immunodeficiency must not be secondary to any infection such as HIV/AIDS or to any other disease process such as malignancy or trauma.

Follow-up of notifications

Clinicians notifying a case of PID will be sent two reply-paid questionnaires but will only be required to complete one of these. If you notify a case of SCID you should complete the SCID questionnaire only (which is unchanged from previously). Clinicians reporting any other case of PID will be asked to complete the first page of the PID questionnaire. If the case has been referred to an immunologist you will be asked to supply their name and they will be contacted for further information, including laboratory results. This will ensure that all cases are being reported but should minimise the workload for non-immunologists. All paediatric immunologists in Australia are involved as investigators in this study and have agreed to this proposal.

Investigators

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References

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- Baumgart K, Britton WJ, Kemp A, French M, Roberton D. Spectrum of Primary Immunodeficiency Disorders in Australia : ASCIA Register. Proceedings of 5th Annual Scientific Meeting of the Australasian Society of Clinical Immunology and Allergy, Canberra, December 1994.

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