Primary Immunodeficiency

Primary immunodeficiency (PID) diseases are a group of potentially serious disorders in which inherited or acquired defects in the immune system lead to an increased risk of infections. While many of these disease present in childhood, some may emerge later in life.

Examples of infections that occur in PID include:

- Infections that are unusually severe, persistent, recurrent or resistant to treatment
- Infections involving unexpected spread or unusual organisms

Information Required

- Referral criteria are 1 or more of the following:
  - 4-6 sinopulmonary infections/year requiring antibiotics
  - Sinopulmonary infections with a prolonged course requiring multiple course of oral antibiotics or IV antibiotics
  - Chronic suppurative lung disease/bronchiectasis, unknown cause
  - Recurrent meningitis
  - Recurrent and severe infections with environmental mycobacteria and usually harmless viral or opportunistic pathogens
  - Recurrent deep skin or organ abscesses
  - Persistent extensive oral thrush or cutaneous fungal infection
  - Recurrent deep-seated infections including septicemia in the absence of an alternative cause
  - A family history of primary immune deficiency
- History of infections – date, type of infectious organism, severity, treatment trialed and response
- Family history of immunodeficiency and autoimmunity
- Current management

Investigations Required

Please include results of the following:

- Full blood count (with differential)
- Immunoglobulins: IgG, IgA, IgM and IgE
- Chest X-ray or CT (if history of recurrent chest infections or bronchiectasis)

Not seen in Immunology OPD clinic

- HIV infection (refer to Infectious Diseases)
- Secondary immunodeficiency due to immunosuppressive medication (refer back to specialist- can be seen in Immunology clinic on specialist referral)

Fax referrals to Allergy/Clinical Immunology Service

Royal Adelaide Hospital Fax: 08 7074 6241

Red Flag

- Very low or absent IgG plus either low IgA or IgM
- Suppurative lung disease / bronchiectasis of unknown cause

Suggested GP Management

- Suspected cases of PID should be promptly referred to the RAH Allergy/Clinical Immunology Service for diagnosis and treatment, clearly highlighted as **"suspected immunodeficiency"**
- Treat infections in the usual manner; however every effort should be made to isolate the causative organism and promptly clear the infection
- Hold live attenuated viral vaccinations (including rotavirus, MMR and varicella) pending specialist assessment
- Exclude concomitant bronchiectasis

Clinical Resources

- Australasian Society of Clinical Immunology and Allergy (ASCIA) www.allergy.org.au
- Immune Deficiencies Foundation Australia (IDFA) www.idfa.org.au

General Information to assist with referrals and the referral templates for the RAH are available to download from the CALHN Outpatient Services website: https://www.rah.sa.gov.au/health-professionals/outpatient-referrals

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1.0	June 2018	July 2020	Original

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