Primary Immunodeficiency

Primary immunodeficiency (PID) diseases are a group of potentially serious disorders in which inherited defects in the immune system lead to increased infections.

- Many people with primary immune deficiency diseases have experienced similar patterns of symptoms; often referred to as “Warning signs and symptoms” (See below)

Information Required

- Referral criteria are 1 or more of the following:
  - 4 or more new ear infections within 1 year
  - 2 or more serious sinus infections within 1 year
  - 2 or more months on antibiotics with little effect
  - 2 or more pneumonias within 1 year
  - Failure of an infant to gain weight or grow normally
  - Recurrent, deep skin or organ abscesses
  - Persistent thrush in mouth or fungal infection on skin
  - Need for intravenous antibiotics to clear infections
  - 2 or more deep seated infections including septicaemia
  - A family history of primary immune deficiency
- History of infections – date, type of infectious organism, severity, treatment trialled and response
- Family history
- Diagnostic test results – organisms identified
- Current management

Investigations Required

- Full blood count (with differentials)
- Liver function tests including Albumin/Protein
- Immunoglobulins: IgG, IgA and IgM
- Lymphocyte surface markers
- C4
- Chest X-ray (if history of chest infections)

Fax referrals to Allergy/Clinical Immunology Service

Flinders Medical Centre
Fax: 08 8204 7483

Red Flag

- Very low or absent IgG or IgM
- Low C4

- Suppurative lung disease / bronchiectasis
- Severe failure to thrive

Suggested GP Management

- Suspected cases of PID should be promptly referred to FMC Allergy/Clinical Immunology (ACI) Service for diagnosis and treatment, clearly highlighted as “suspected immunodeficiency”
- Treat infections in the usual manner; antibiotics often first line treatment.
- Hold live attenuated viral vaccinations (including rotavirus, MMR and varicella) pending specialist assessment

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 FMC is available to download from the SALHN Outpatient Services website www.sahealth.sa.gov.au/SALHNoutpatients.

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