

Naval Medical Center Portsmouth
Allergy/Immunology Referral Guidelines: Immunodeficiency

Definition(s)

- **Primary Immunodeficiency** - disorders of the immune system that predispose affected individuals to increased rate and severity of infection, immune dysregulation, autoimmune disease and malignancy, distinct from secondary immunodeficiencies.
- **Secondary Immunodeficiency** - disorders of the immune system secondary to viral infections, immunosuppression to prevent graft versus host after transplant, treatment of systemic autoimmune disease or cancer chemotherapy.

When to Suspect Primary Immunodeficiency

- Recurrent bacterial infections (more frequent than expected at the patient's age)
- More than one severe infection (ie. meningitis, osteomyelitis, pneumonia, sepsis)
- Infections that present atypically, are unusually severe or chronic or fail regular treatment (ie. require IV antibiotics)
- Two or more months on antibiotics with little effect
- Recurrent subcutaneous abscesses or any abscess of an internal organ
- Any infection caused by an unexpected or opportunistic pathogen (ie. pneumocystis)
- Severe or long lasting warts, generalized mollusca contagiosa
- Extensive candidiasis, recurrent oral thrush in children > 1 year
- Failure of an infant to gain weight or grow normally
- Family history of primary immunodeficiency

Initial Diagnosis and Management

- Initial evaluation of suspected immune deficiency may begin with the primary care provider. A basic evaluation to exclude many forms of primary immune deficiency includes:
 - CBC with differential (check platelet volume, absolute lymphocyte, neutrophil and eosinophil counts)
 - IgG, IgA, IgM and IgE (IgGAME)
 - Consider lymphocyte subpopulations (T, B, and NK cells) in cases of severe infections, recurrent viral or fungal infections, or failure to thrive with eczema
 - Consider HIV ELISA

Importance of Early Recognition

- Severe Combined Immune Deficiency (SCID) is a medical emergency, and successful treatment is dependent on rapid recognition.
- Timely recognition of primary immunodeficiency in children and adults is important for prevention of organ damage, decreased quality of life, and decreased life span.

Indication for Specialist Referral

- Specialist referral is recommended for cases of suspected primary immunodeficiency for comprehensive immunological evaluation.
- Patients with any kind of severe or recurrent infection under 6 months of age, especially if accompanied with failure to thrive, (even if this failure starts after 3 or 4 months of age) warrants an urgent referral. Please call the Allergy/Immunology duty pager to facilitate.

Reference:

1. de Vries E, et al. Patient-centered screening for primary immunodeficiency, a multi-stage diagnostic protocol designed for non-immunologists: 2011 update. Clin Exp Immunol 2011;167:108-119.
2. Practice parameter for the diagnosis and management of primary immunodeficiency. Ann Allergy Asthma Immunol, 2005. 94(5 Suppl 1): p. S1-63.