Immunodeficiency Evaluation for Chronic Infections in Infants and Children Testing

Recurrent respiratory infections with or without chronic diarrhea

**CONSIDER**
- Immunoglobulins (IgA, IgG, IgM), Quantitative
- Lymphocyte Subset Panel 6 - Total Lymphocyte Enumeration with CD45RA and CD45RO or Lymphocyte Subset Panel 7 – Congenital Immunodeficiencies
- Response to polyvalent pneumococcal vaccine if >2 years
- Response to DT vaccine
- Sweat chloride testing (at accredited cystic fibrosis center)

**ORDER**
- HIV PCR – test of choice in children ≤18 months (antibodies do not function in infants)
- CD4+ T-Cell Recent Thymic Emigrants (RTEs)
- Lymphocyte Subset Panel 6 - Total Lymphocyte Enumeration with CD45RA and CD45RO or Lymphocyte Subset Panel 7 – Congenital Immunodeficiencies
- Lymphocyte Antigen and Mitogen Proliferation Panel

**Immunoglobulin disorder evaluation**

- Low IgG, IgM, IgA OR poor antibody response to vaccination
- B cells present
- Immunology consult
- May need immunoglobulin replacement
- Common variable immune deficiency (CVID) or other immune deficiency

**T-cell disorder evaluation**

- High IgM
  - Low IgG, IgA
  - Hyper IgM syndrome likely
  - Consider genetic testing

- Positive sweat chloride
  - ORDER Genetic testing
  - Cystic fibrosis
    - IgA absent
    - Low IgG 2,4
    - Poor pneumococcal vaccine response
    - Warn of possible reaction to IgA-containing blood products
    - Immunology consult
    - May need immunoglobulin replacement with low IgA-containing preparation

- Low IgA
  - ORDER Immunoglobulin G Subclasses (1, 2, 3, 4)
  - IgA absent
    - Normal IgG subclasses
  - IgA low
    - Normal IgG subclasses
  - Consider celiac testing

- Abnormal complement activity
  - Possible complement deficiency

- Low IgG or IgM
  - ORDER Individual complement testing based on results of CH50, AH50

- Chronic granulomatous disease
  - Leukocyte adhesion deficiency, type 2
  - Decreased CD16
  - Decreased CD15
  - Increased IgE
  - Possible hyper IgE syndrome (Job syndrome)

- decreased CD11b/CD18
  - Leukocyte adhesion deficiency, type 1
  - Decreased CD15
  - Increased IgE
  - Possible hyper IgE syndrome (Job syndrome)

- Low neutrophil count
  - Neutrophil oxidative burst assay (DHR)
  - Neutrophil adaptation deficiency, type 2
  - Neutrophil antibody positive
  - Myeloperoxidase deficiency

- Kostmann agranulocytosis
  - Low neutrophil count
  - Neutrophil adaptation deficiency, type 1
  - Neutrophil antibody positive
  - Myeloperoxidase deficiency

- DiGeorge syndrome
  - Chromosome FISH, Metaphase (specify 22q11.2 deletion)

- Possible severe combined immunodeficiency
  - SCID genetic panel

- Abnormal complement activity
  - Possible complement deficiency

- All normal
  - Contact immunology director for further evaluation

- Low IgM, IgM, IgA
  - ORDER B-Cell Memory and Naive Panel
  - No B cells present
  - Bruton X-linked agammaglobulinemia
  - Consider genetic testing

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HIV algorithm

Hematology consult
- May need granulocyte colony stimulating factor

Autoimmune neutropenia

Myeloperoxidase deficiency

Job syndrome