Immunodeficiency Evaluation for Chronic Infections in Infants and Children Testing

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1. 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 diphtheria and tetanus (DT) vaccine
   - Sweat chloride testing (at an accredited cystic fibrosis center)

2. Hypogammaglobulinemia
   - Low IgG
   - IgM, IgA OR poor antibody response to vaccination
   - B cells present
   - Common variable immune deficiency (CVID) or other immune deficiency
   - Immunology consult
   - May need immunoglobulin replacement
   - Agammaglobulinemia
   - No B cells present
   - Consider genetic testing

3. Hyper IgM syndrome likely
   - High IgM, low IgG, IgA
   - Cystic fibrosis
   - ORDER Genetic testing

4. Positive sweat chloride
   - ORDER Genetic testing
   - Immunology consult
   - May need immunoglobulin replacement with low IgA-containing preparation

5. Low IgA
   - Immunoglobulin G Subclasses (1, 2, 3, 4)
   - IgA absent
   - Low IgG 2,4 Poor pneumococcal vaccine response
   - IgA absent Normal IgG subclasses
   - Warn of possible reaction to IgA-containing blood products
   - Consider celiac testing

6. Normal IgG subclasses
   - Abnormal T cell recent thymic emigrants (RTEs)
   - Decreased response to Candida spp
   - Immunity consult
   - Possible severe combined immunodeficiency

7. Abnormal RTEs
   - CD4+ T cell recent thymic emigrants (RTEs)
   - May need immunoglobulin replacement
   - Agammaglobulinemia
   - Low T cells
   - Abnormal RTEs
   - Decreased response to Candida spp
   - Immunology consult
   - Possible severe combined immunodeficiency

8. HIV positive
   - HIV NAAT – test of choice in children ≤18 months (antibodies do not function in infants)
   - Abnormal RTEs
   - Decreased T cells
   - Possible severe combined immunodeficiency
   - ORDER Toll-Like Receptor Function

9. Possible severe combined immunodeficiency
   - Positive for deletion
   - Decreased IgE
   - Increased IgE
   - Possible hyper IgE syndrome (Job syndrome)
   - Leukocyte adhesion deficiency, type 1
   - Decreased neutrophil count
   - Neutrophil antibody positive
   - Deficiency of myeloperoxidase
   - Myeloperoxidase deficiency

10. ORDER Individual complement testing based on results of CH50, AH50
    - Abnormal complement activity
    - Possible complement deficiency
    - If all normal, contact immunology director for further evaluation

11. Low IgG or IgM
    - Hypogammaglobulinemia
    - Abnormal DHR
    - Decreased CD11b/CD18
    - Decreased CD15
    - Candida spp
    - Specific IgE neutrophil chemotaxis

12. Genetic testing
    - Chronic granulomatous disease
    - Leukocyte adhesion deficiency, type 1
    - Leukocyte adhesion deficiency, type 2
    - Candida spp
    - Specific IgE neutrophil chemotaxis

13. Kostmann agranulocytosis
    - May need granulocyte colony stimulating factor

14. Neutrophil antibody positive
    - Autoimmune neutropenia

15. Absence of myeloperoxidase
    - Leukocyte adhesion deficiency

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