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**
- Immunoglobulins (IgA, IgG, IgM), Quantitative
- Lymphocyte Subset Panel 6 - Total Lymphocyte Enumeration with CD45RA and CD45RO or Lymphocyte Subset Panel 7 – Congenital Immunodeficiencies
- Lymphocyte Antigen and Mitogen Proliferation Panel

**Low IgG, IgM, IgA OR poor antibody response to vaccination**

**ORDER**
- B-Cell Memory and Naive Panel

**B cells present**

- Low T-cell
- Low LAM
- Abnormal RTEs

- Immunology consult
- May need immunoglobulin replacement

- Agammaglobulinemia

- HIV positive

- See HIV algorithm

**No B cells present**

- Hyper IgM syndrome likely

- Consider genetic testing

**High IgM low IgG, IgA**

- Hyper IgM syndrome likely

- Consider genetic testing

**Low IgA**

- Immunoglobulin G Subclasses (1, 2, 3, 4)

- IgA absent
- Low IgG 2,4
- Poor pneumococcal vaccine response

- Warn of possible reaction to IgA-containing blood products

- Consider celiac testing

**IgA low Normal IgG subclasses**

- Innate immune deficiency

- ORDERS IRAK-4 and toll receptor

- Abnormal

- All normal

**Severe viral infections; Candida spp or other fungal infections; recurrent sinopulmonary infections**

**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

**Low T-cell**

- Low LAM
- Abnormal RTEs

- Possible severe combined immunodeficiency

**No B cells present**

- Agammaglobulinemia

- HIV positive

- See HIV algorithm

**Possible severe combined immunodeficiency**

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

**Abnormal complement activity**

- Possible complement deficiency

**Positive for deletion**

- DiGeorge syndrome

**Abnormal DHR**

- Chronic granulomatous disease

**Decreased CD11b/CD18**

- Leukocyte adhesion deficiency, type 1

**Decreased CD15**

- Leukocyte adhesion deficiency, type 2

**Increased IgE**

- Possible hyper IgE syndrome (Job syndrome)

**Candida spp specific IgE**

- Neutrophil oxidase deficiency, type 2

**Hematology consult**

- Absence of myeloperoxidase

**Autoimmune neutropenia**

- Myeloperoxidase deficiency

**Kostmann agranulocytosis**

- Low neutrophil count

**Neutrophil antibody positive**

- May need granulocyte colony stimulating factor

- Myeloperoxidase deficiency

**Hypogammaglobulinemia**

- Abnormal DHR

**Negative for deletion**

- All normal

**If all normal, contact immunology director for further evaluation**