

Educational material

APPROACH TO LYMPHOPENIA IN CHILDREN

➤ Lymphopenia: definition-

Absolute numbers of lymphocytes vary during childhood due to the maturation of the immune system. Lymphocytes constitute around 60-75% of circulating leukocytes in a newborn and around 20-40% in an adult.

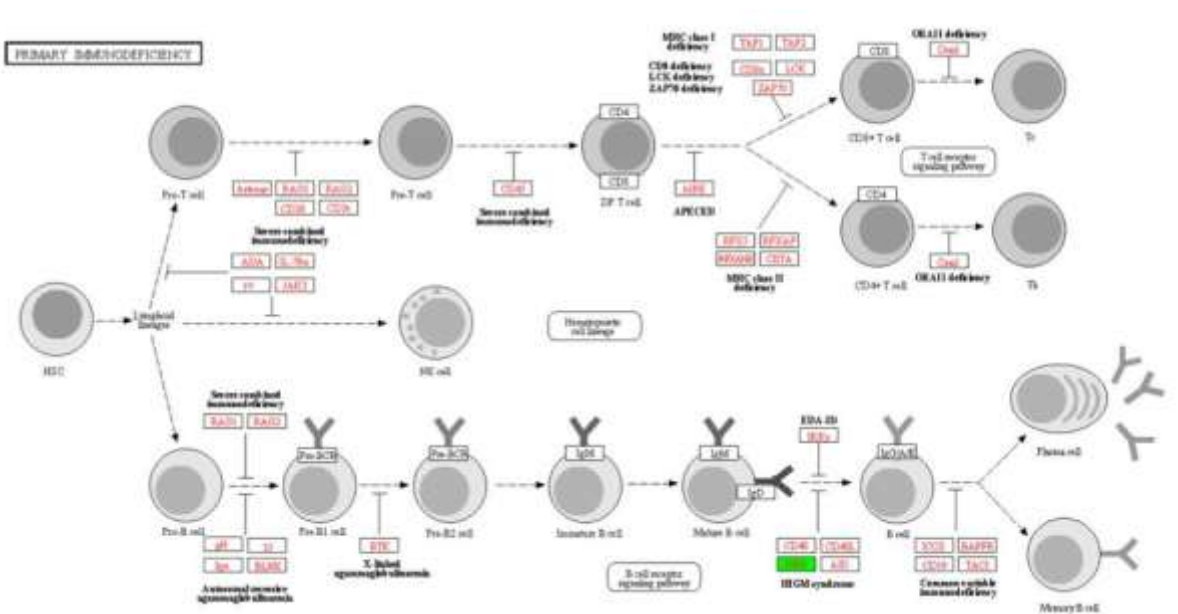
Absolute lymphocyte count (meriting further evaluation)-

<2000 cells/ μ L at birth

<4500 cells/ μ L in an infant

<1000 cells/ μ L in an older child or adult.

**Always observe the trend of ALC*



➤ **Lymphopenia: etiology-**

- Infections: **Transient lymphopenia** is observed frequently in viral infections-

Influenza, Dengue, HIV, Rubella

Enteric fever

- Autoimmune diseases: SLE
- Drugs: immunosuppressive therapy, corticosteroids, cytotoxic agents
- Primary immunodeficiency-

Severe combined immunodeficiency (prototype)

WAS

Di George syndrome

Ataxia telangiectasia

X linked agamma globulinemia

Idiopathic CD4 lymphopenia

➤ **How do lymphopenic disorders present?**

Failure to thrive from early infancy (including intractable diarrhea, severe eczema). Unusual infections or unusual severity of infections.

Encountered pathogens

- **Viruses** (CMV, EBV, VZV, HSV, adenovirus, HHV8, HPV, Molluscum contagiosum, RSV)
- **Fungi** (superficial *Candida*, *Aspergillus*, *Cryptococcus*, *Histoplasma*, *Pneumocystis jirovecii/carinii*)
- **Protozoa** (*Toxoplasma*, *Microsporidium*, *Cryptosporidium*)
- **Intracellular bacteria** eg. *Mycobacterium* sp, *Salmonella*

Non immunological diagnosis

- Gastrointestinal, Renal, Cardiopulmonary, Endocrine, Neurological, Metabolic and Congenital causes.
- Malignancy
- Chronic lead poisoning.
- Perinatal infection.
- Severe malnutrition
- HIV

➤ **Approach to lymphopenia-**

< 6 months

-Symptomatic (infected): observe ALC, repeat within a week

-Asymptomatic: repeat after 2 weeks and document normal ALC.

-If low ALC persists, refer to a specialist

6 months- 2 years:

-SCID is still possible at this age

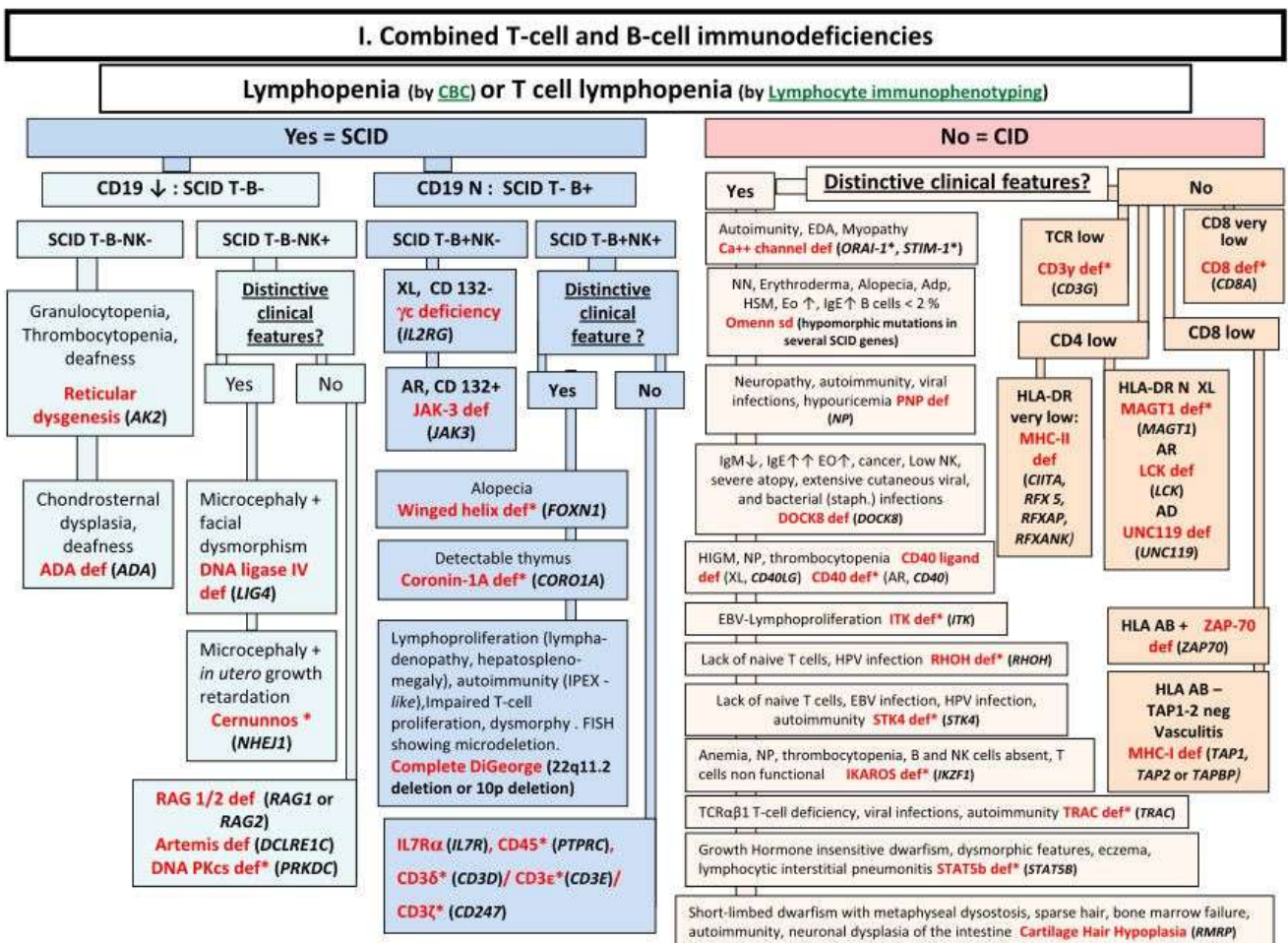
-If symptomatic: evaluate

-If asymptomatic, no prior symptoms: unlikely to be significant

2 years

-Classical SCID unlikely after 2 years

-Other combined immunodeficiencies (Di George syndrome), HIV possible



➤ **SCID and lymphopenia-**

- Age < 2 years • ALC (< 3000 cells/ μ L)
- Persistent diarrhea
- Respiratory symptoms
- Disseminated BCG infections
- Bacterial infections
- Failing to thrive
- Fatal unless treated early
- All patients with SCID are lymphopenic at birth
- Only exceptions
- Hypomorphic SCID mutations
- Engraftment of maternal lymphocytes