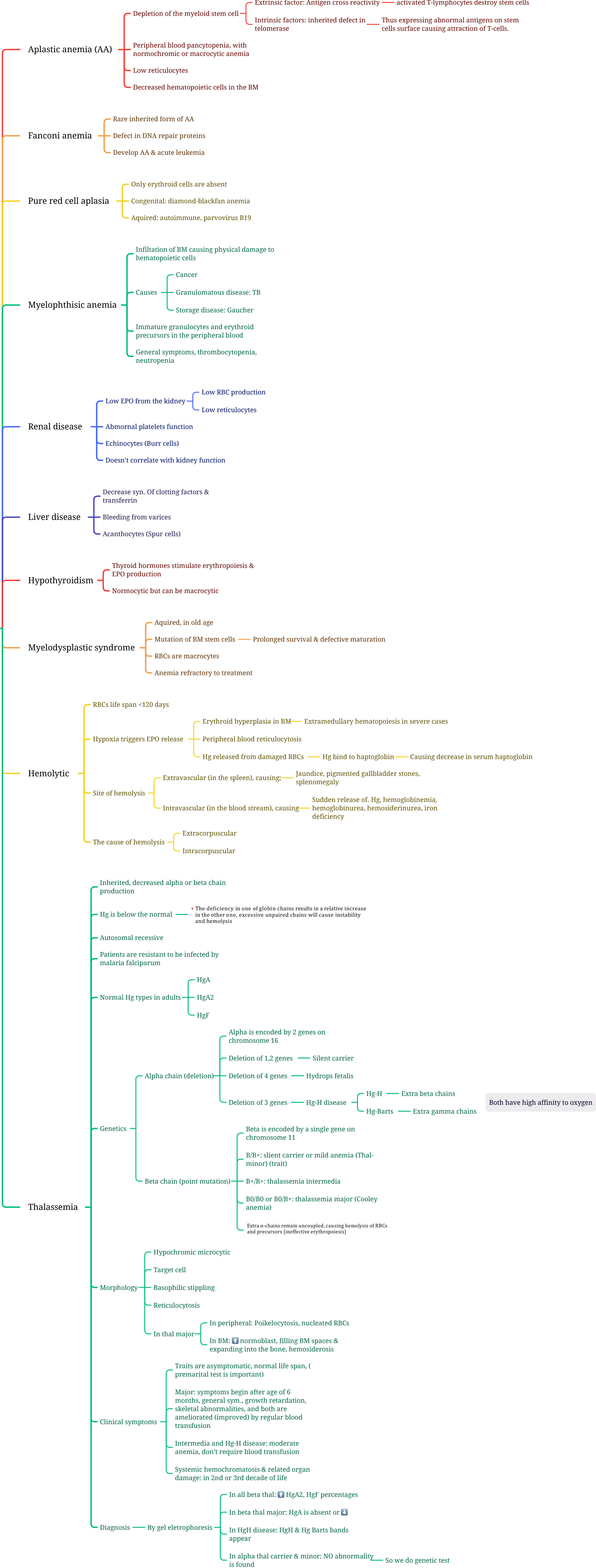


Anemia 3



# Sickle cell anemia

## General info.

- Most common hemolytic anemia worldwide
- Resistant to malaria falciparum
- Autosomal recessive
- Glutamic acid → valine in beta chain
- Hg electrophoresis

- Sickle cell disease (homozygous)
  - HgS is found, HgA is absent
- Sickle cell carrier (heterozygous)(asymptomatic)
  - Both HgS & HgA are presented

## Pathogenesis

- In deoxygenated case: HgS tends to polymerize in a longitudinal pattern
  - Causes distorting cell shape & creat sickle shape
- This change is reversible by reoxygenation
  - But with repeated sickling, the cell membrane is damaged and hemolysis occurs
- HgS polarization is inhibited in the presence of:
  - HgA
  - HgF in newborns
- When alpha thalassemia is presented too, the sickling is decreased
- Sickled RBCs take longer time to pass through capillaries
- They may adhere to the endothelial cells making a thrombus
- Sickled cells are removed by macrophages in spleen (extravascular hemolysis)

## Clinical symptoms

- Chronic moderate severe hemolytic anemia
  - Manifesting after the age of 6 months
- Vaso-occlusive crisis
  - Results in organ infarction
- Hand-foot syndrome
- Acute chest syndrome
- Stroke
- Myocardial infarction
- Retinopathy
- Autosplenectomy
- Aplastic crisis
  - Infection by parvovirus B19
- Susceptibility for encapsulated bacteria

## Diagnosis

- Blood smear
  - Adding hypoxic agent
- Hemoglobin electrophoresis
- DNA testing

