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Melanie Chang
Thalassemia

Described by Cooley in 1927

Autosomal recessive microcytic anemia

Decreased production of globin chain synthesis (alpha or beta)

Alpha thal- Southeast Asia, Middle East, China, African

Beta thal- Mediterranean
Imaging features

Skeletal: osteoporosis, marked bone marrow expansion, premature fusion of growth plates, predisposition to vertebral compression fractures

Non-skeletal: extramedullary hematopoiesis, cholelithiasis, hepatosplenomegaly
The Hemoglobinopathies: Sickle cell Disease and Thalassemia
www.isradiology.org/tropical_deseases/tmcr/chapter31/radiological9.htm
Treatment: Hypertransfusion

Reduces the extent of marrow expansion

Iron overload and hyperuricemia are potential consequences

High serum iron levels → synovium and articular cartilage abnormalities

Occasionally chondrocalcinosis

Treatment: Iron-chelation therapy - Deferoxamine

Dysplasia in ~1/3 of patients affecting growing long bones

Toxic effects on bone from zinc chelation, antiproliferative effect

Impaired metaphyseal collagen synthesis and inhibition of osteoblasts → growth retardation

Metaphyseal splaying, irregular widened physis


