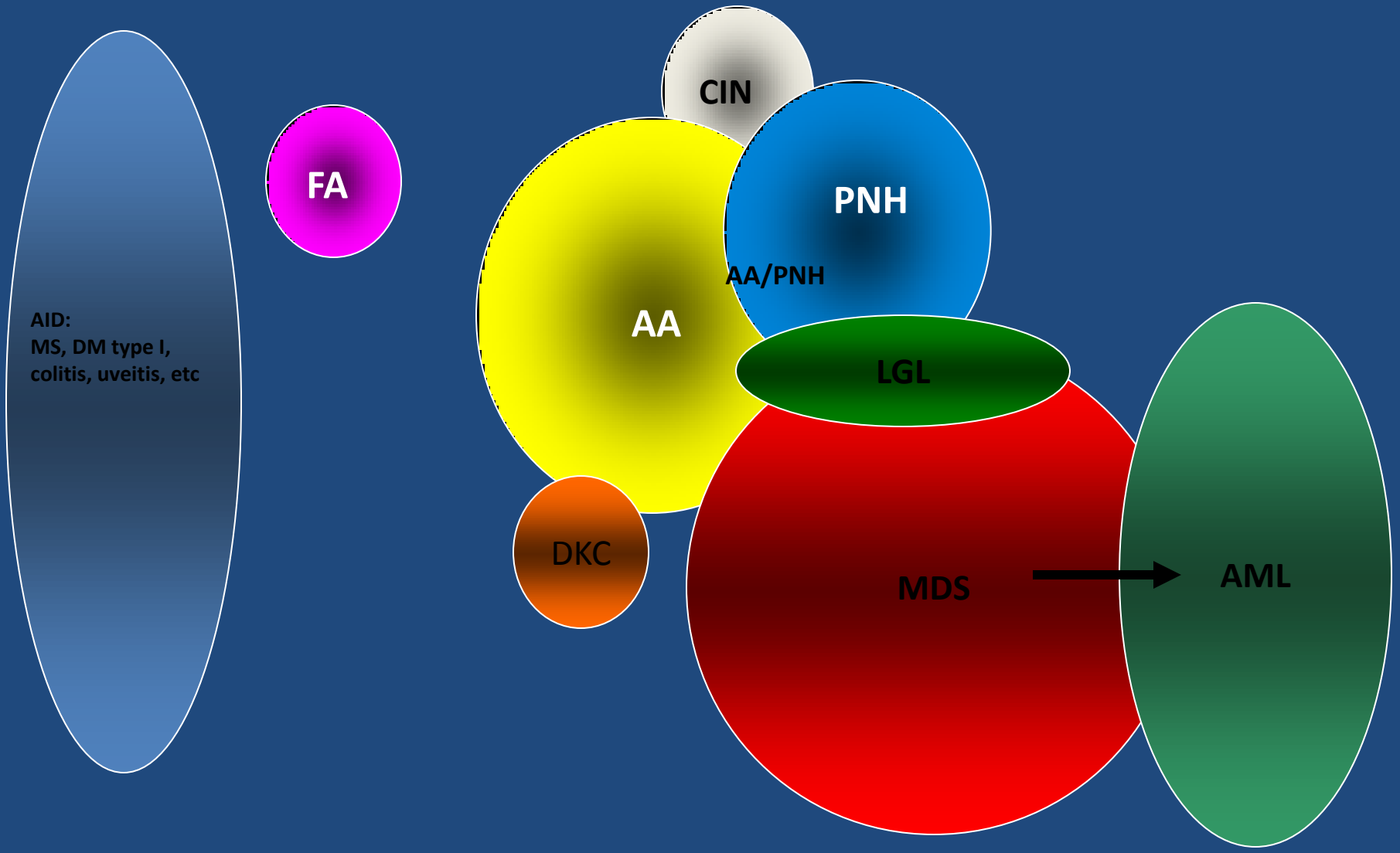
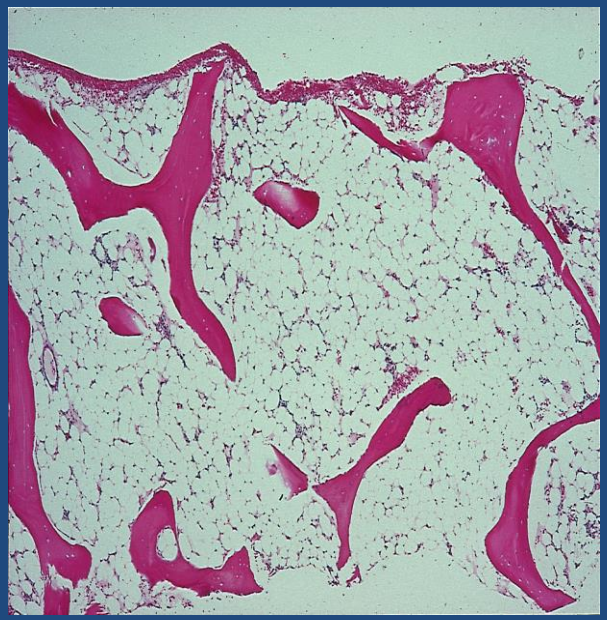
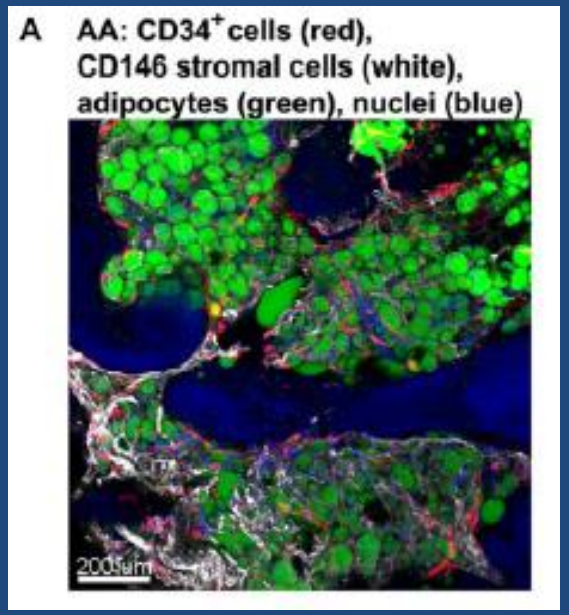
Απλαστική Αναιμία

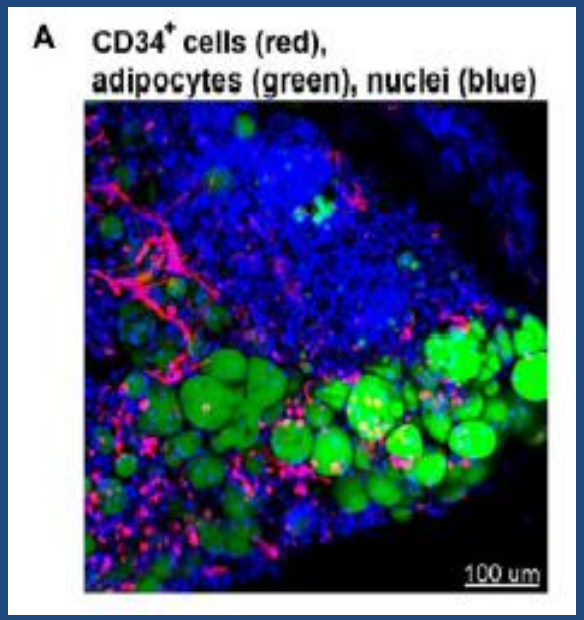
Bone Marrow Failure Syndromes



Η Απλαστικη αναιμία είναι σπάνιο αυτοάνοσο νόσημα που χαρακτηρίζεται από αντικατάσταση του αιμοποιητικού ιστου-μυελού- από λίπος. Το αποτελεσμα ειναι υποκυτταρικός μυελός και πανκυτταροπενια στην περιφέρεια

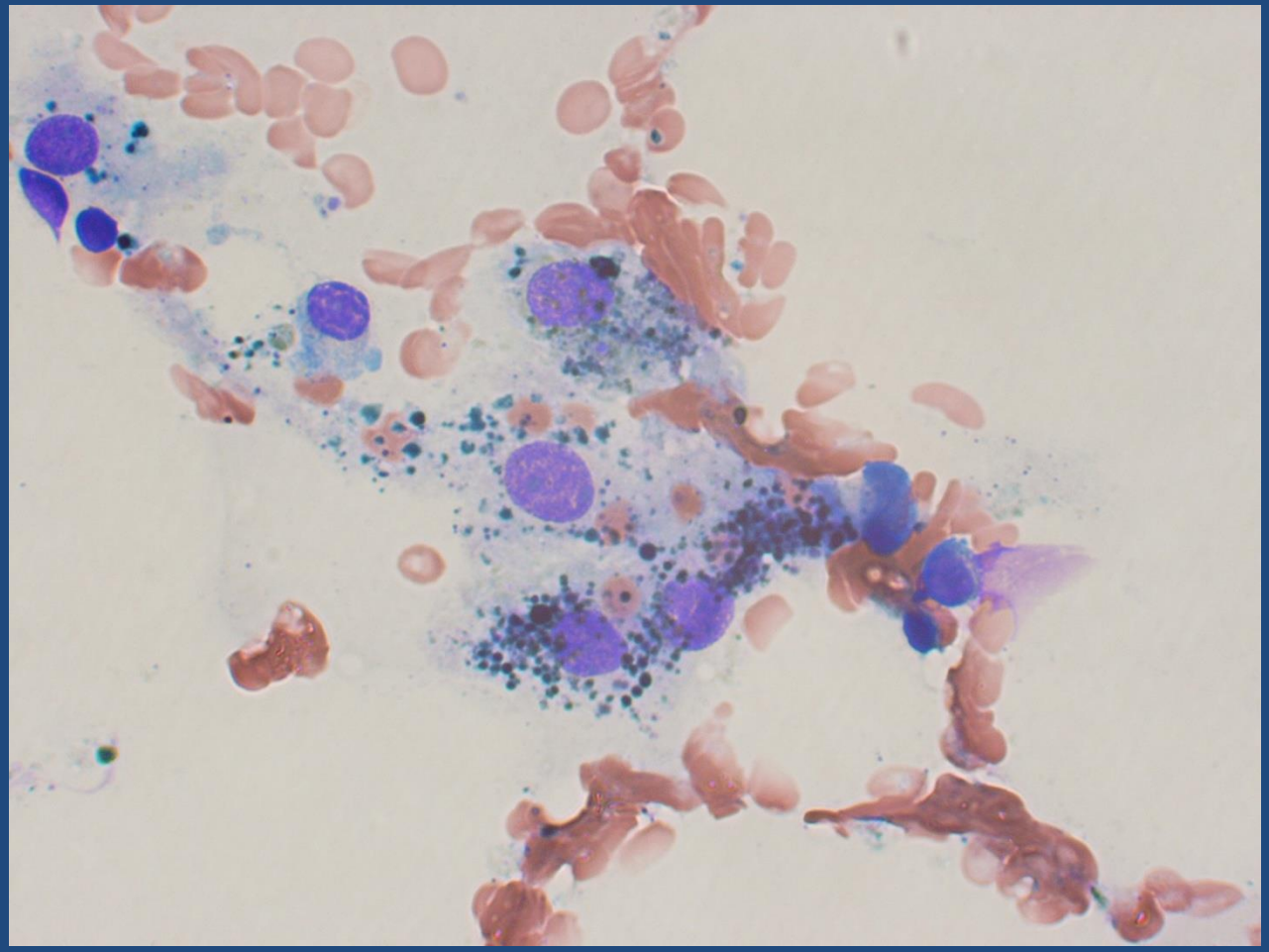






Takaku T et al, Blood 2010; 116: e41-e55

Increased hemophagocytosis in aplastic anemia



Αιτιολογική Ταξινόμηση Απλαστικής Αναιμίας

I. *Direct Toxicity*

radiation

cytotoxic chemotherapy

Benzene

Intermediate metabolites of some drugs

II. *Immune-Mediated*

Iatrogenic

transfusion-associated GVHD

Eosinophilic fasciitis

Hepatitis

Pregnancy

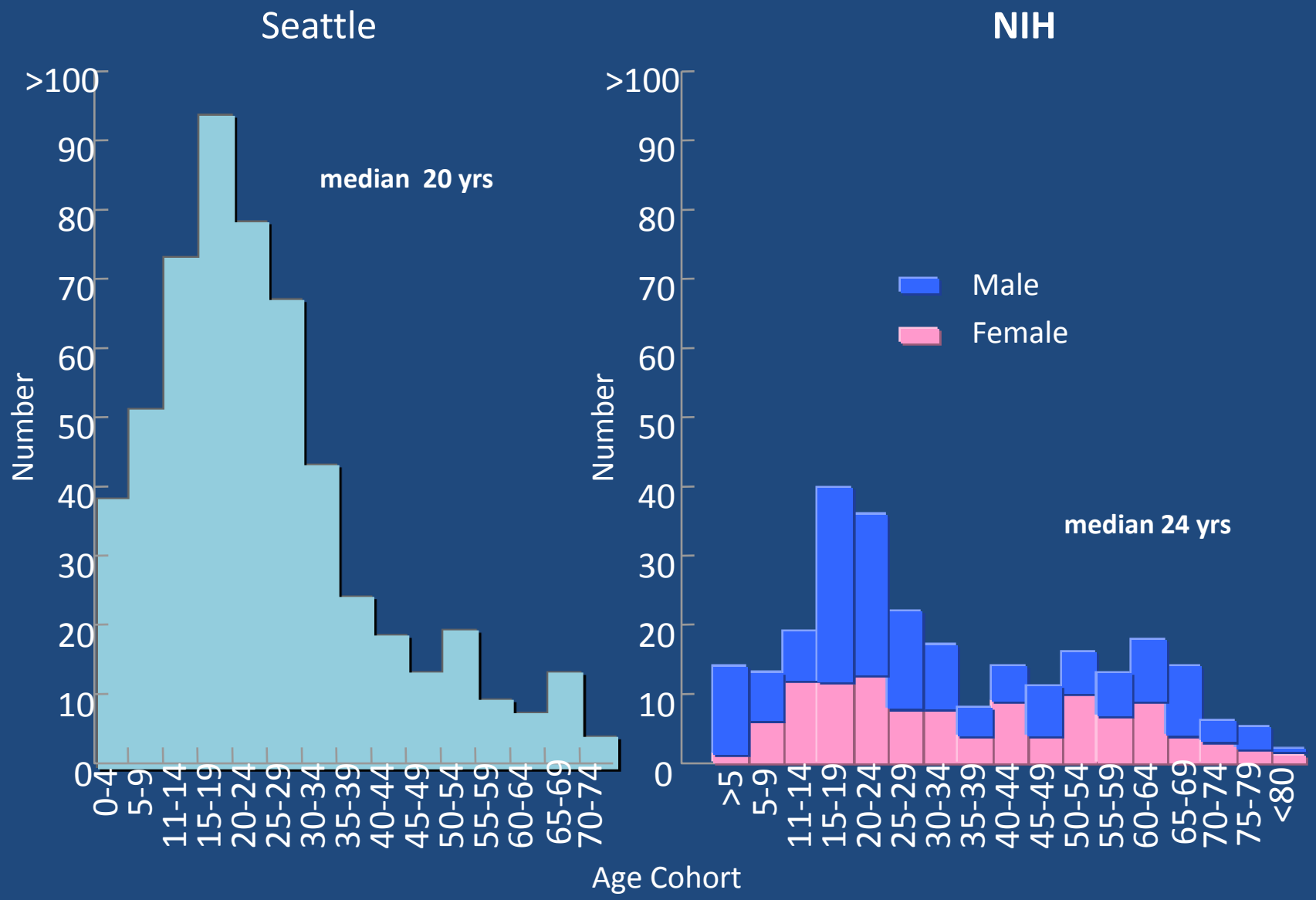
Intermediate metabolites of some drugs

Idiopathic aplastic anemia

III. *Constitutional* (Fanconi anemia, Dyskeratosis congenita)

Other syndromes

DEMOGRAPHICS OF APLASTIC ANEMIA



Διαβαθμιση Βαρύτητας Απλαστικής αναιμίας:

Βαριά απλαστική αναιμία: Peripheral Blood: two of three values:

ANC < 500

PLT < 20.000

Reticulocytes < 1% or <20.000 (absolute number)

Marrow cellularity < 30%

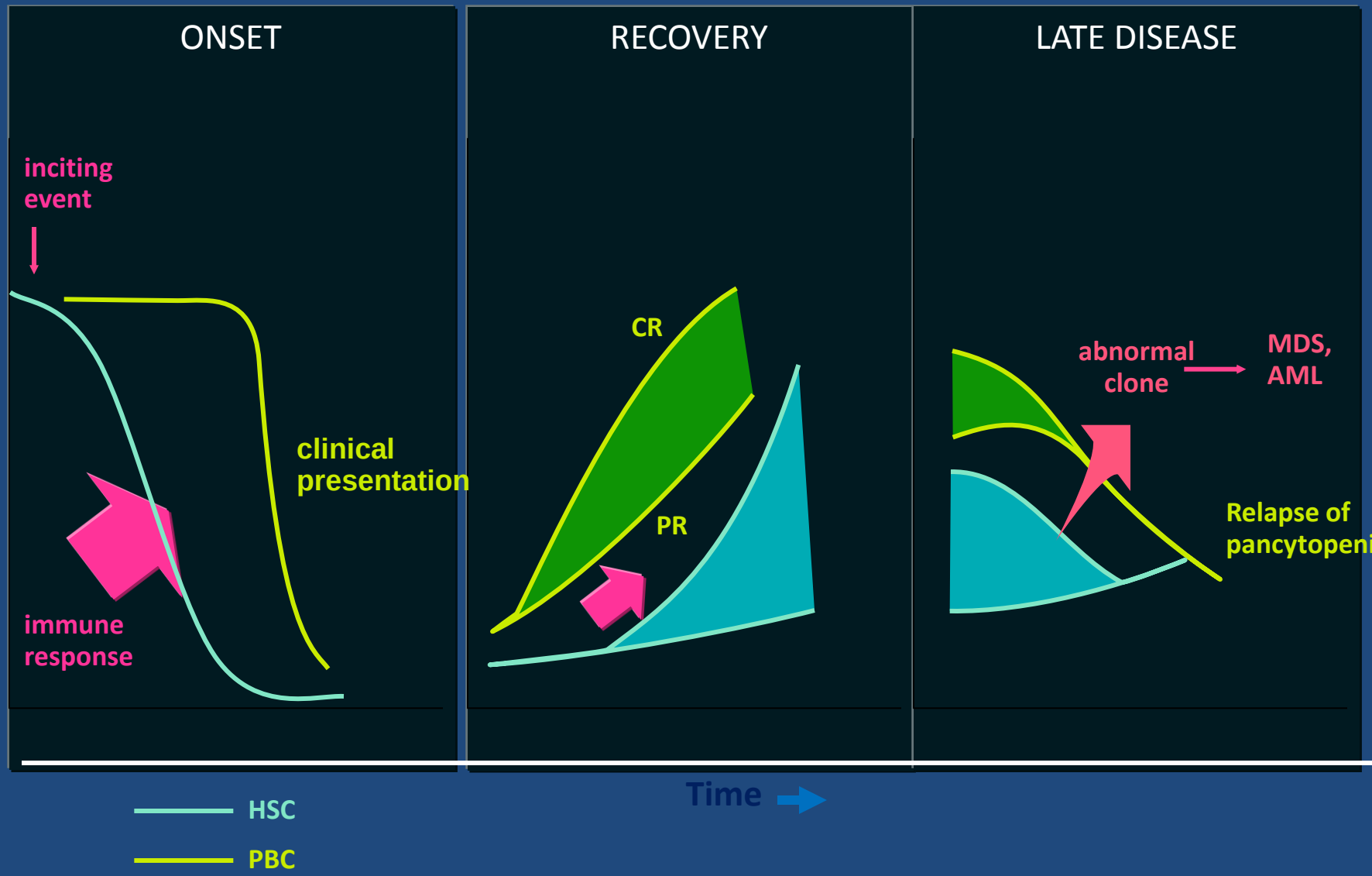
Πολύ Βαρια απλαστική αναιμία (very severe): As above but ANC < 200

Ηπια (moderate) απλαστικη αναιμια: Marrow cellularity < 30%

ANC > 500

RBC or PLT transfusion dependent

ACQUIRED APLASTIC ANEMIA



Aplastic anemia is an autoimmune disease

- Response to IST represents the best evidence of an immune-mediated pathogenesis
- T cell defects (CTL, IFN-γ, Tregs, Th17, ……)
- Auto-antibodies and cytokines
- Telomere dysfunction (shorter telomeres, mutations in TERT and TERC)
- Mesenchymal stem cell defects
- Increased apoptosis and decreased proliferation of HSC

Summary:

- SAA is a fatal disease that demands rapid recognition and institution of both Immediate tx of low blood counts and adequate hematopoiesis

- Most cases of AA is the result of T cell-mediated destruction of hematopoietic stem cells of the BM

- AA should be distinguished from other causes of pancytopenia

- BM examination is required for diagnosis: low cellularity and normal residual precursors, and signs of hemophagocytosis are common

- In some cases there is overlap with MDS and PNH

- Tx options: HLA identical sibling BMT in children and young adults, and IST with ATG-based regimens in pts >40yrs

- Relapse after successful IST is common. A minor proportion of the pts develop late clonal disease: abnormal cytogenetics, MDS, leukemia.