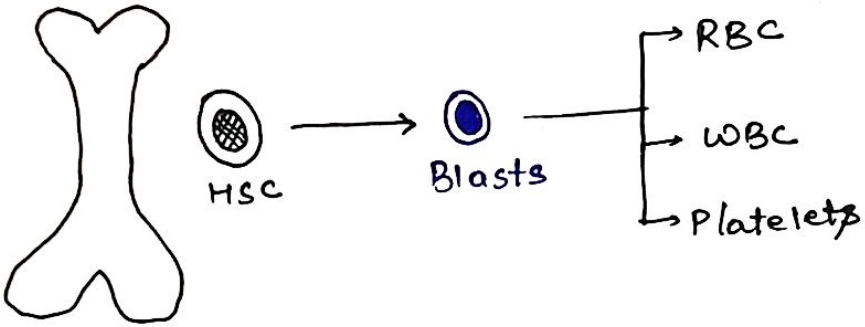
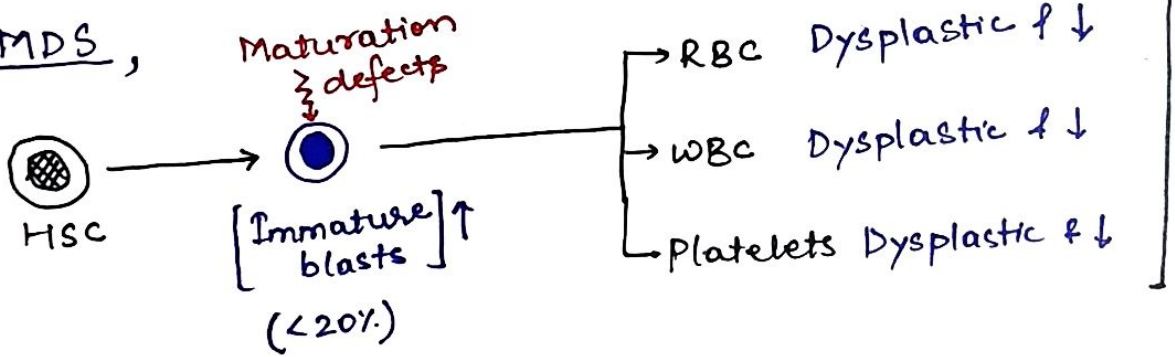
MYELODYSPLASTIC SYNDROME (MDS)

Normally,



MDS,



* ↑ risk of transformation to AML

Definat^n :- Group of clonal stem cell disorders with maturation defects

↓

ineffective hematopoiesis

↑

↑ risk of transformation to AML

(N) → <20% MDS → >20% AML

sification :-

MDS

- **Primary**
  - Cause :- Idiopathic
- **Secondary**
  - Cause :- Drug/therapy related.

Pathogenesis :-

- Epigenetic factors
- Transcription factors
- RNA splicing factors
- Others

Epigenetic factors :-

Mutation altering

- DNA methylation
- Histone modification
- Chromatin looping

RNA splicing factor :-

Mutation involving

- 3' RNA splicing machinery

Transcription factors :-

- t (8;21)
- t (15,17)
- inv (16)

→ can progress to AML

Others :-

Deletion

5q
7q
20q

→ m/c in adult MPS

Monosomy

5, 7

→ m/c in Childhood MDS

Trisomy

8

Person → (N) Blood count
→ 1/more pathogenic mutations associated ∈ MDS

⇒ CHIP [Clonal Hematopoiesis of Indeterminate Potential] (Asympto)

↓ to

MDS (Sympto)

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↓ to

MDS (Sympto)

Morphology :-

(full of dysplastic cells)

① 

- usually Hypercellular
- Sometimes Normocellular
- Less commonly Hypocellular

Myeloid blasts ↑
(But < 20%)

② Dysplastic differentiation of RBC, WBC, platelets

Erythroid series
- Ring sideroblast { Erythroblast ∈ iron-laden mitochondria visible as perinuclear granules in Prussian blue stain.
- nuclear bridging
- nuclear lobulation
- multiple nuclei
- cytoplasmic granules
- Nuclear budding abnormalities

Neutrophils - ↓ secondary granules
Dohles bodies
- Pseudo-Pelger-Huet cells → {Neutrophils ☹ only 2 lobes}

Megakaryocytes - Pawn ball megakaryocyte
{Multiple seperate nuclei}

C/F :-

-Adults

- ↓ RBC → pallor
→ Fatigue
→ Weakness

- ↓ WBC :- ↑ risk of infection
- ↓ platelet :- ↑ Bleeding tendencies

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