Hematology

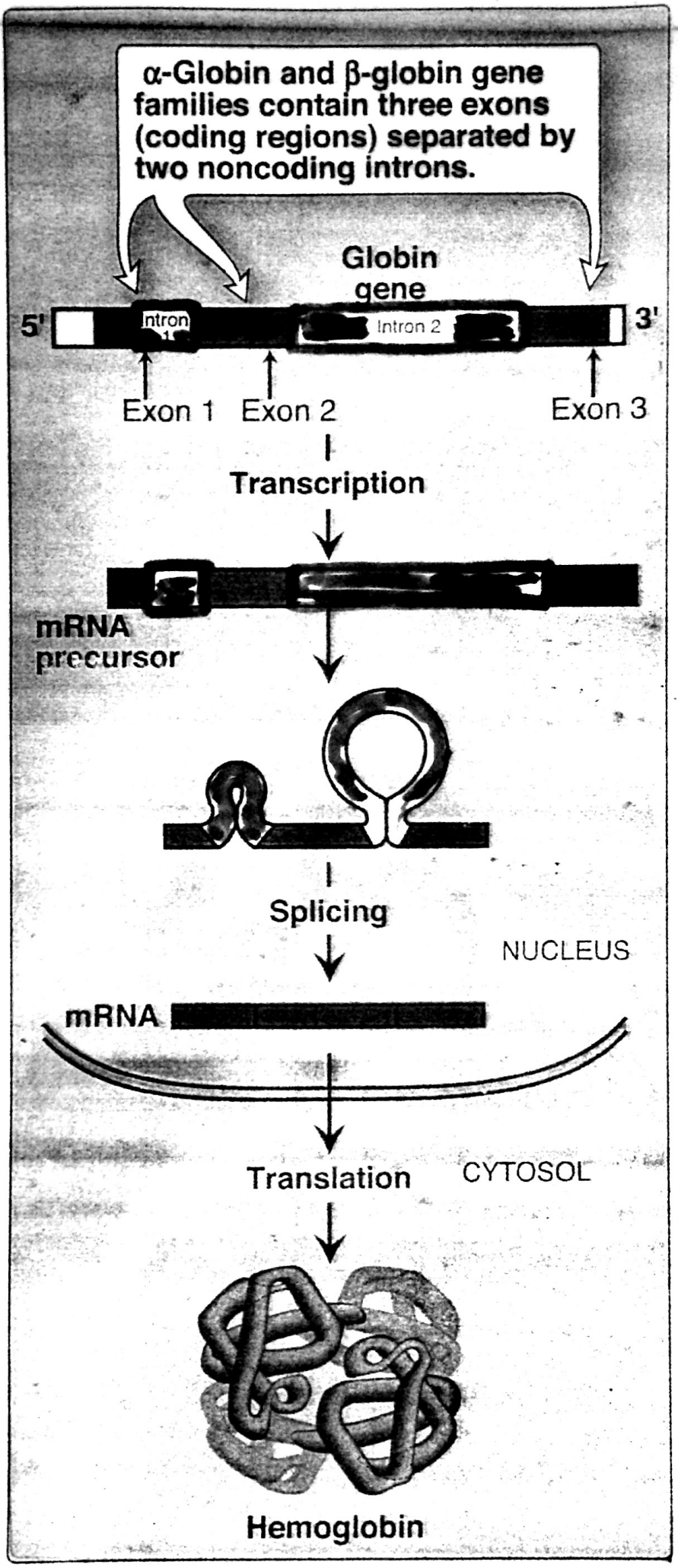
- Histology
- **Biochemistry**
- Pathology
- Pharmacology
- Physiology
- Microbiology

  - Handout
  - **Slide** 7
  - Sheet

- Dr. name : Dr Nayef karadsheh
- lecture number :
- Done BY :

By: Anas Abu Shokab

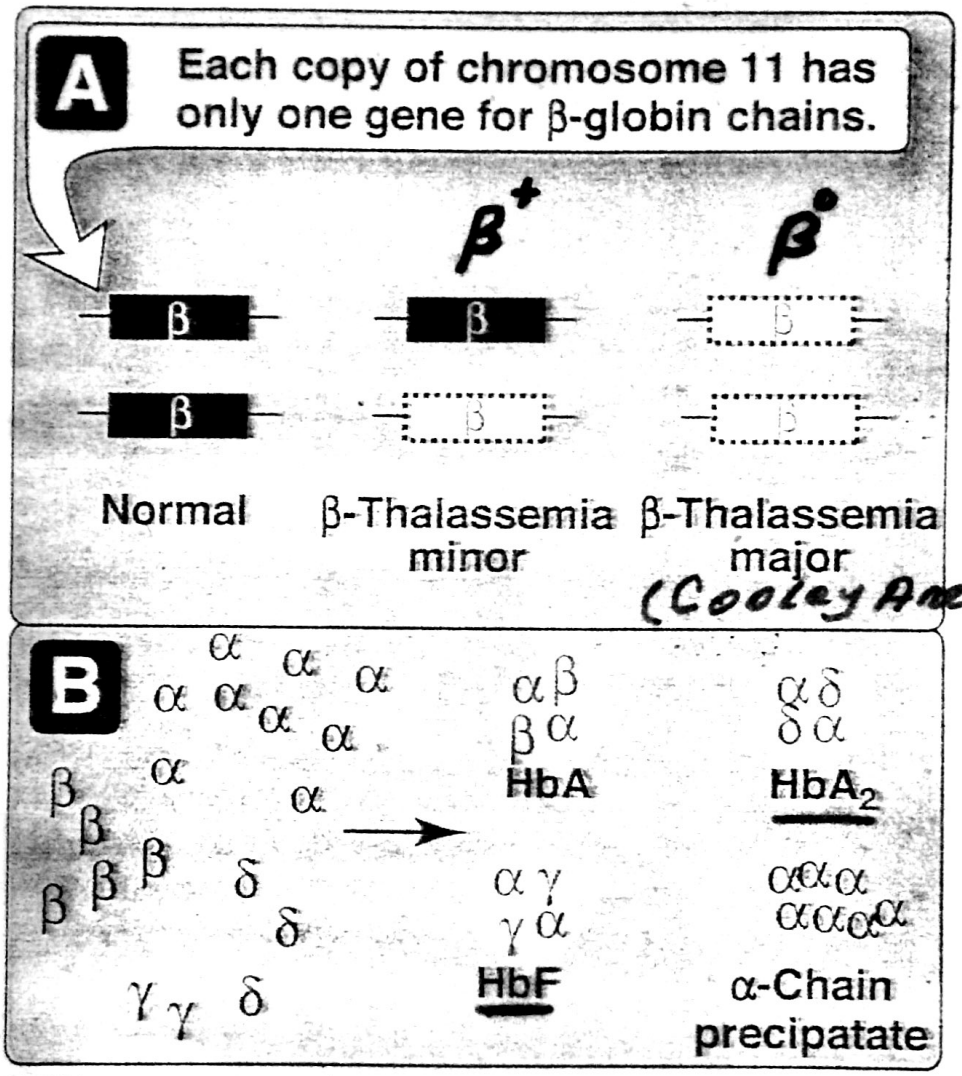
Globin chains Synthesis



THALASSEMIA

- Decreased Synthesis of α- or β-
  - → • Globin chain precipitate
  - → • Hemolysis
  - → • Hypochromic anemia

β-thalassemia

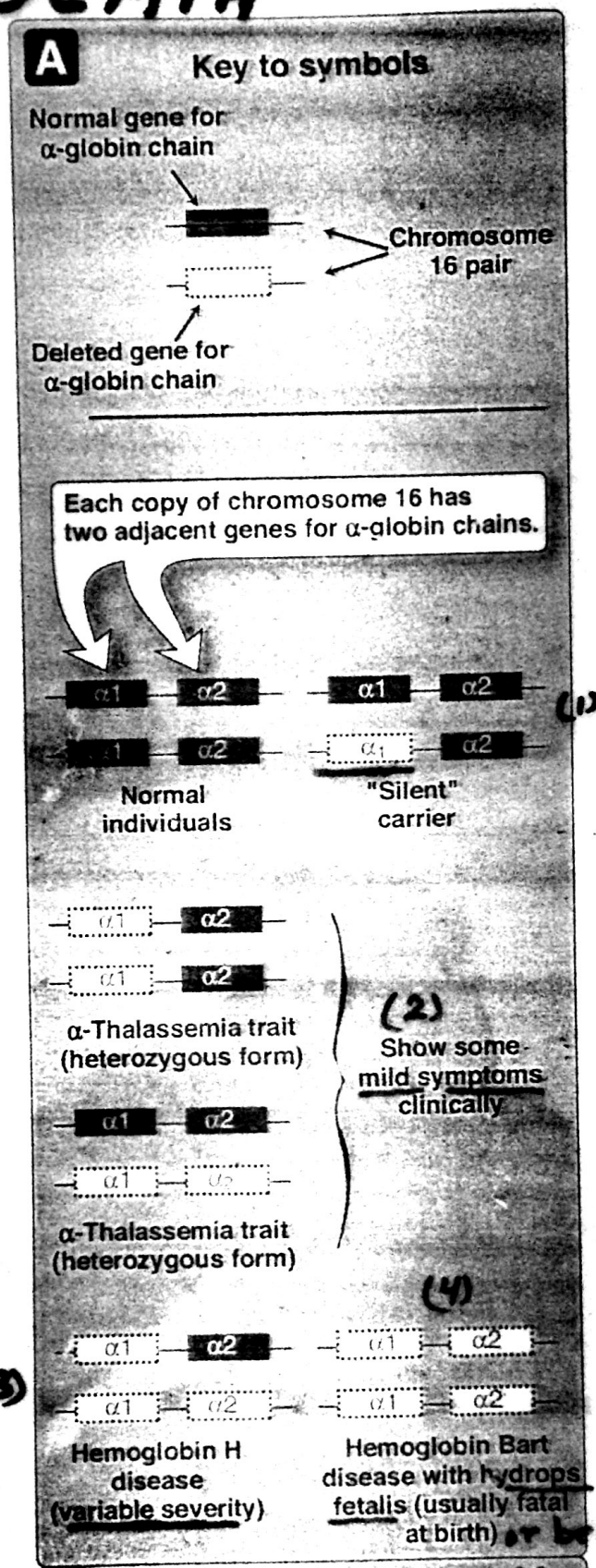


$Hb\,A_2 \uparrow$; $HbF \uparrow$

α chains ⟶ α_4 (Cooley's Hb)
- → • α_4 precipitates
- → Heinz bodies
- ⟶ Cell membrane damage ⟶ Premature death of erythrocyte

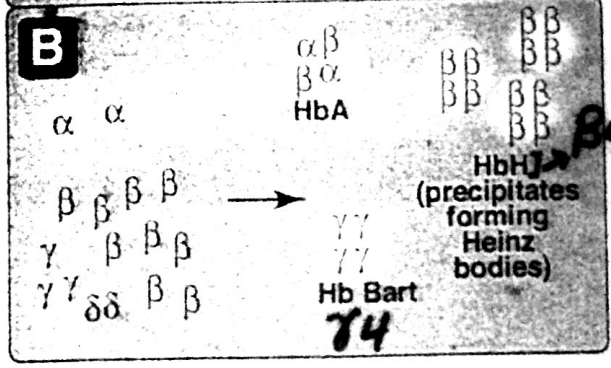
Manifestation of β-thalassemia appears only after birth (because of HbF); becomes severly anemic during the 1st or 2nd year of life.

α-THALASSEMIA



} in total loss of α-chain

or before



HbH ↑
Hb Bart ↑

Soluble Hb but without sigmoidal kinetics. Useless O_2 deliverer to tissues.

Primary Causes:

α- thalassemia :-
gene deletion

β- thalassemia :-
- Point mutation in the Promotor
- mutation in the translational initiation codon
- point mutation in the Poly-adenylation signal
- mutations → splicing abnormalities

- HbE $\beta^{26 Glu-lys}$
  structural and quantity abnormality
  60% of β-globin is made

- Hereditary persistance of fetal Hb (HPFH)
  → continue to make HbF in adult
  → Benign