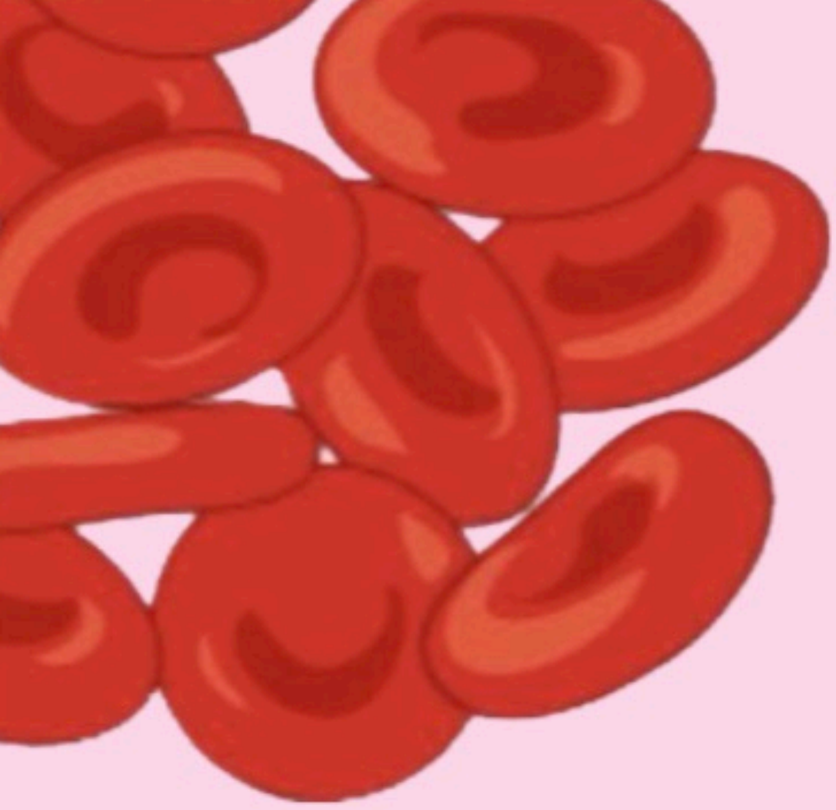


النادي الطبي

HEMATOPOIETIC & LYMPHATIC SYSTEM

-HAYAT BATCH-

SUBJECT : Biochemistry

LEC NO. : 3

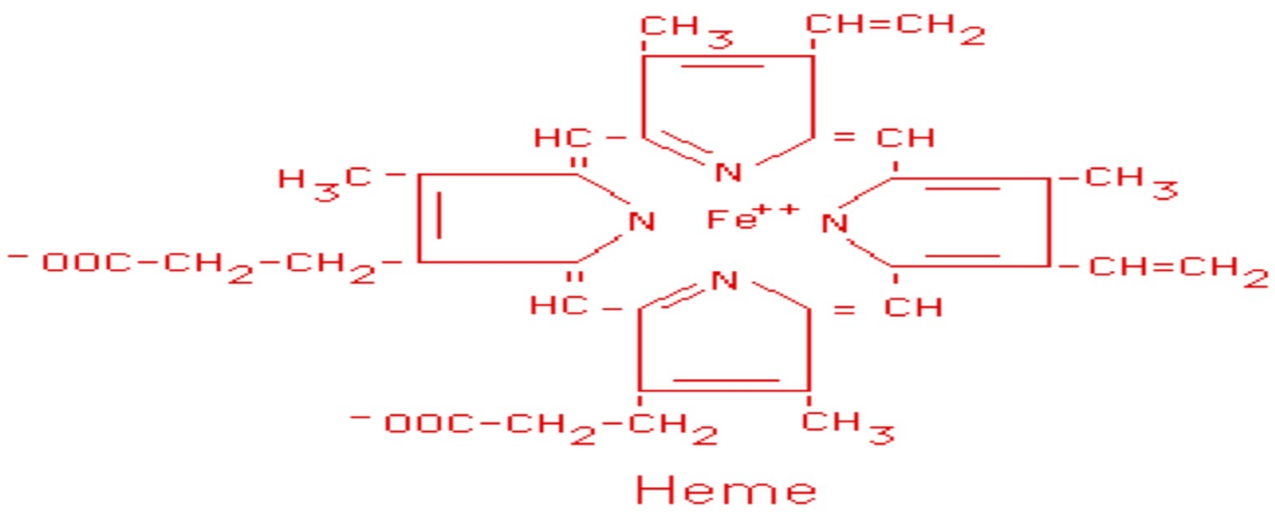
DONE BY : Esra'a Khaled

وَقُل رَّبِّ زِدْنِي عِلْمًا

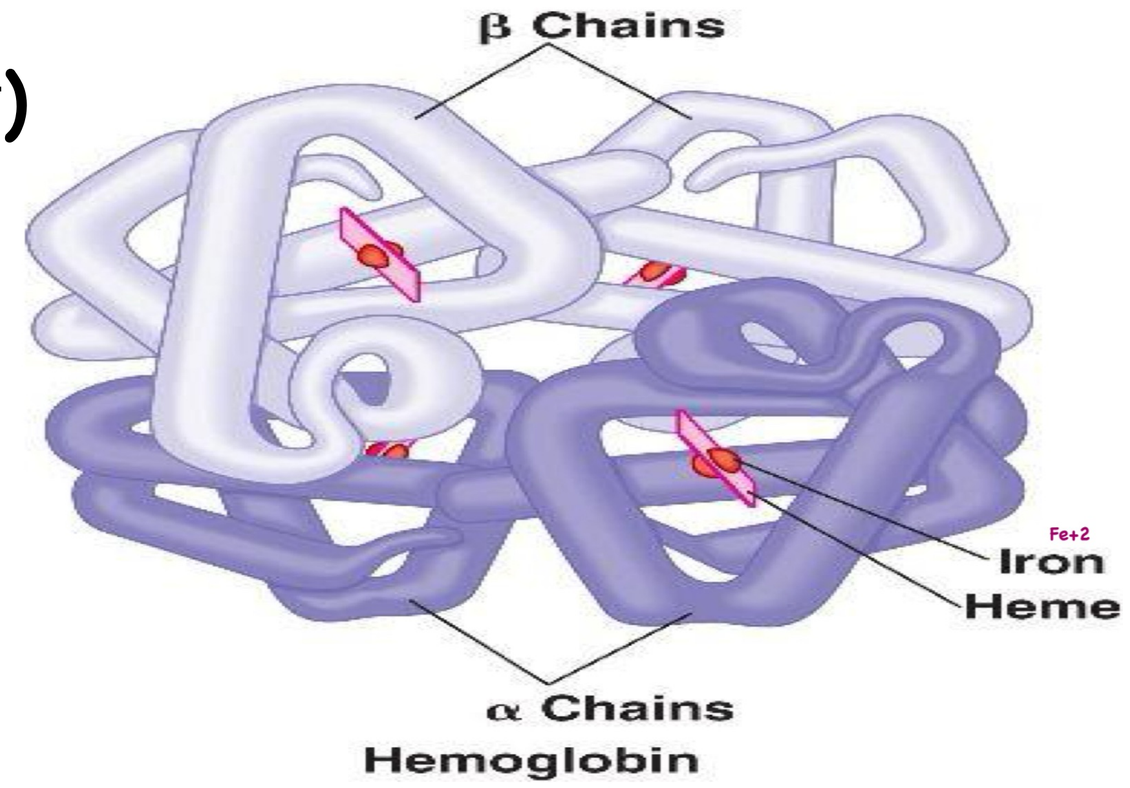
Hb & Hemoglobinopathies

*In Hb iron is in the ferrous state (Fe^{++})

And the ferrous iron has 6 valencies
Which means it has the ability to connect to 6 different atoms



Heme



Hemoglobin

- a chain gene is on chromosome 16 / 141 Amino Acids
- B , Y , & (Delta) chain genes are on chromosome 11 / 146 Amino Acids
- Hb is composed of 2 a and 2 either B , Y , &

Types of Hb

* Hb A
(2a , 2B)

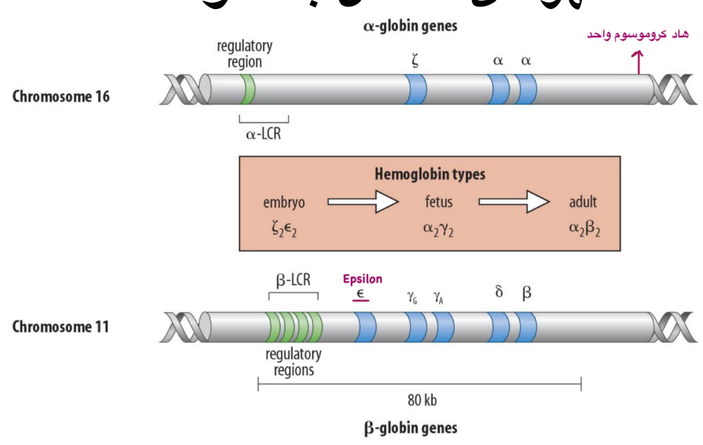
The Hb of normal adult forms about 97-98 % of adult Hb

Hb F
(2a , 2Y)

Present during fetal life and replaced by Hb A by the age of 6 months

- 2 a → (اصلهم) Zeta
- 2 B → (اصلهم) ∈-gene (Epsilon)

بعد ٣ اشهر من الحمل بختفو



* Hb A1 : Glycated Hb A

Hb A1c : normally forms from 4 - 5.6 % of total Hb so if the value is higher than 8% , indicate poorly controlled diabetes mellitus .
In this type glucose is linked to amino group of the N terminal valine of the B chain and This glycosylation depends on the blood glucose level

* Hb A2 (2a , 2&)

2-3 % of adult Hb , Appears in the blood at the age of 3 months

- It increases in B thalassemia

Hemoglobinopathies

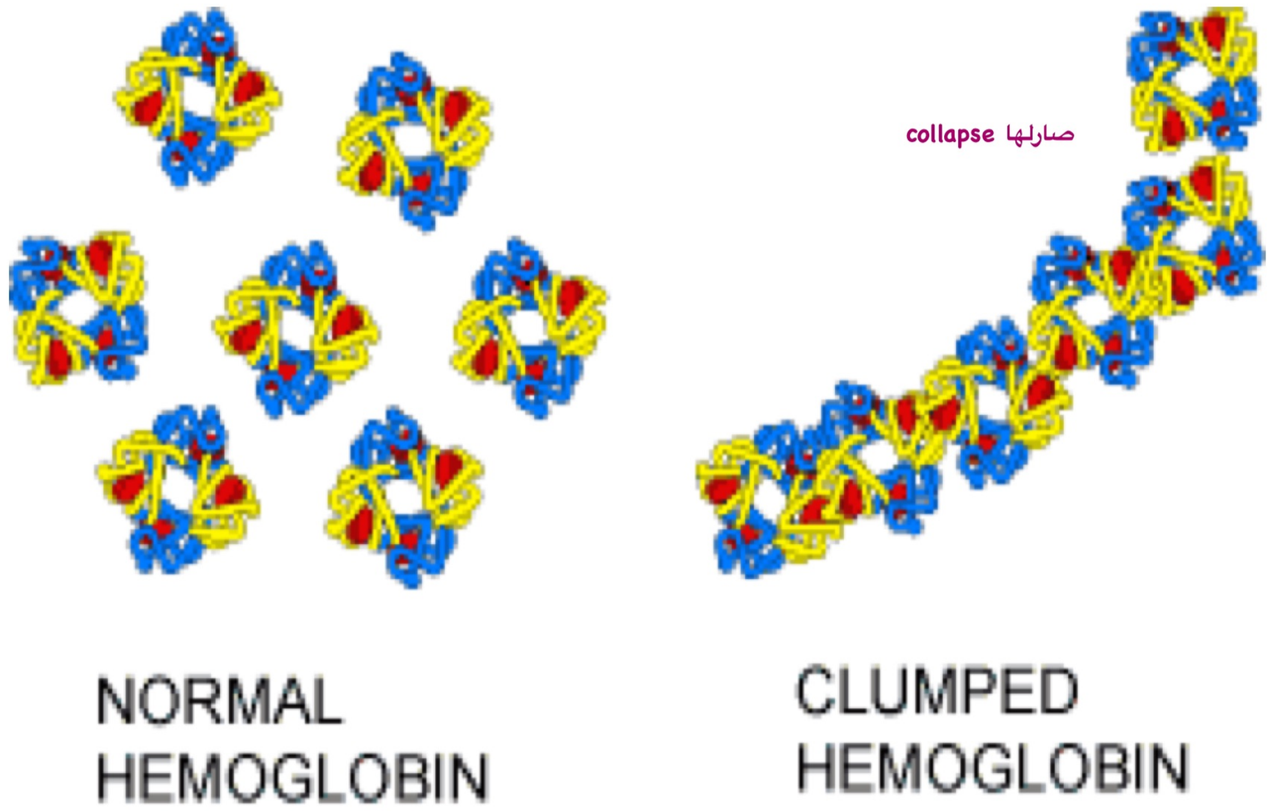
1.Hemoglobin S (HbS) / sickle cell hemoglobin

Genetic disease caused by replacement of glutamic acid in the 6th position of beta chain by valin

- Solubility of HbS in the deoxygenated form is 50 times less than oxygenated form leading to crystallization and formation of a fibrous precipitate in the RBCs which collapes and aquire the shape of a sickle

- The spleen removes sickle cells at a faster rate than normal cells leading to hemolytic anemia

- Individuals who are heterozygous for HbS (both HbA , HbS) in their blood cells , they are sickle cell (carriers) and they are resistant to parasites that causes malaria



α-Thalassemia

1-patients deficient in one α- globin gene are completely normal and are only carriers of α-thalassemia

2-patients deficient in 2 α- globin genes are said to have α-thalassemia trait with mild anemia

3-patients deficient in 3 α-globin genes are said to have α-thalassemia major with sever anemia that is present since birth due to deficient formation of HbF

4- patients deficient in the four α- globin genes are said to have homozygous α-thalassemia They usually die soon after birth or in the uterus as HbF can not be synthesized we get hydrops fetalis

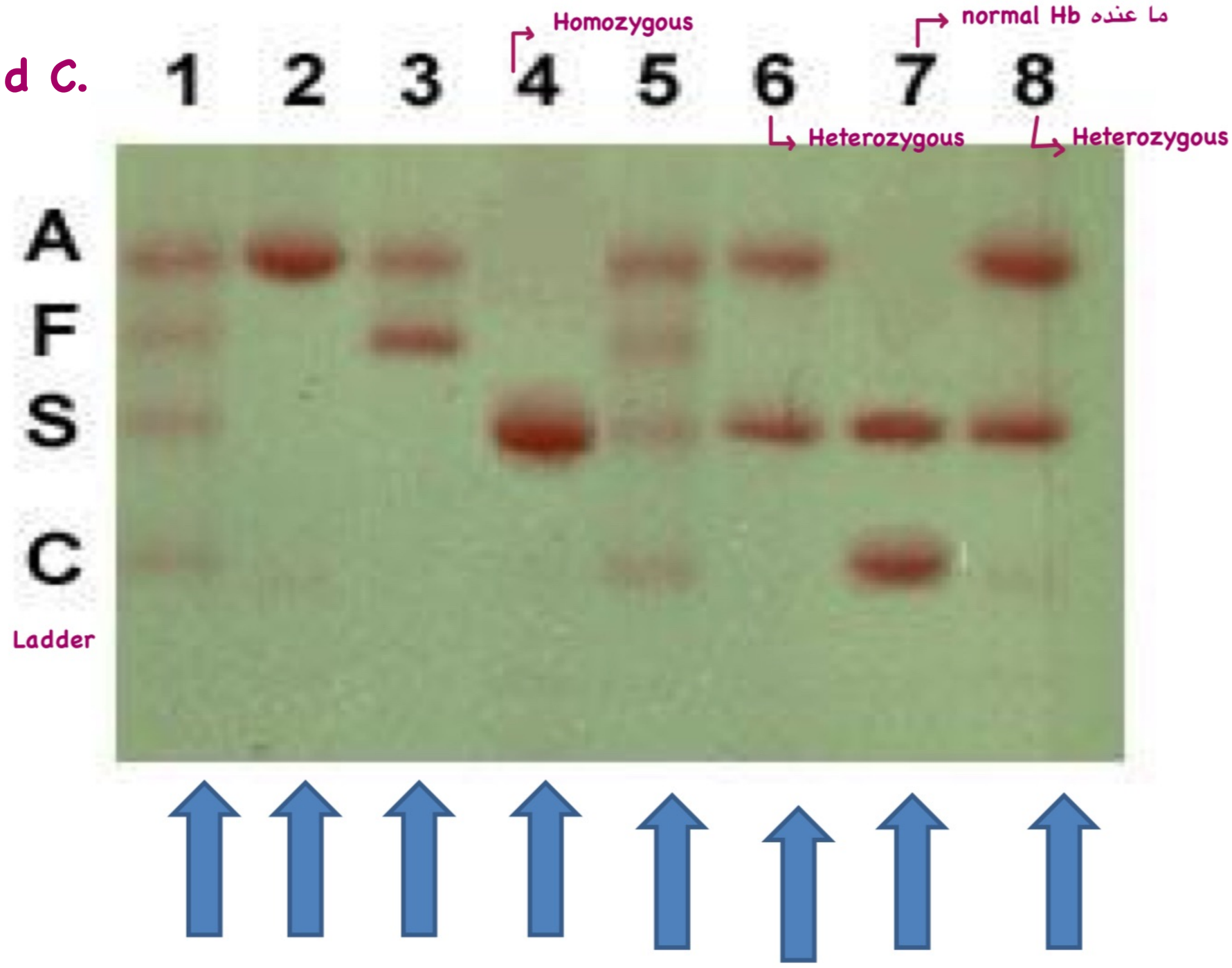
B-Thalassemia

1- If only one gene is defective we get β -thalassemia trait or β - thalassemia minor with mild anemia

2- If the 2 genes are defective we get β - thalassemia major with sever anemia , They rarely live to adulthood

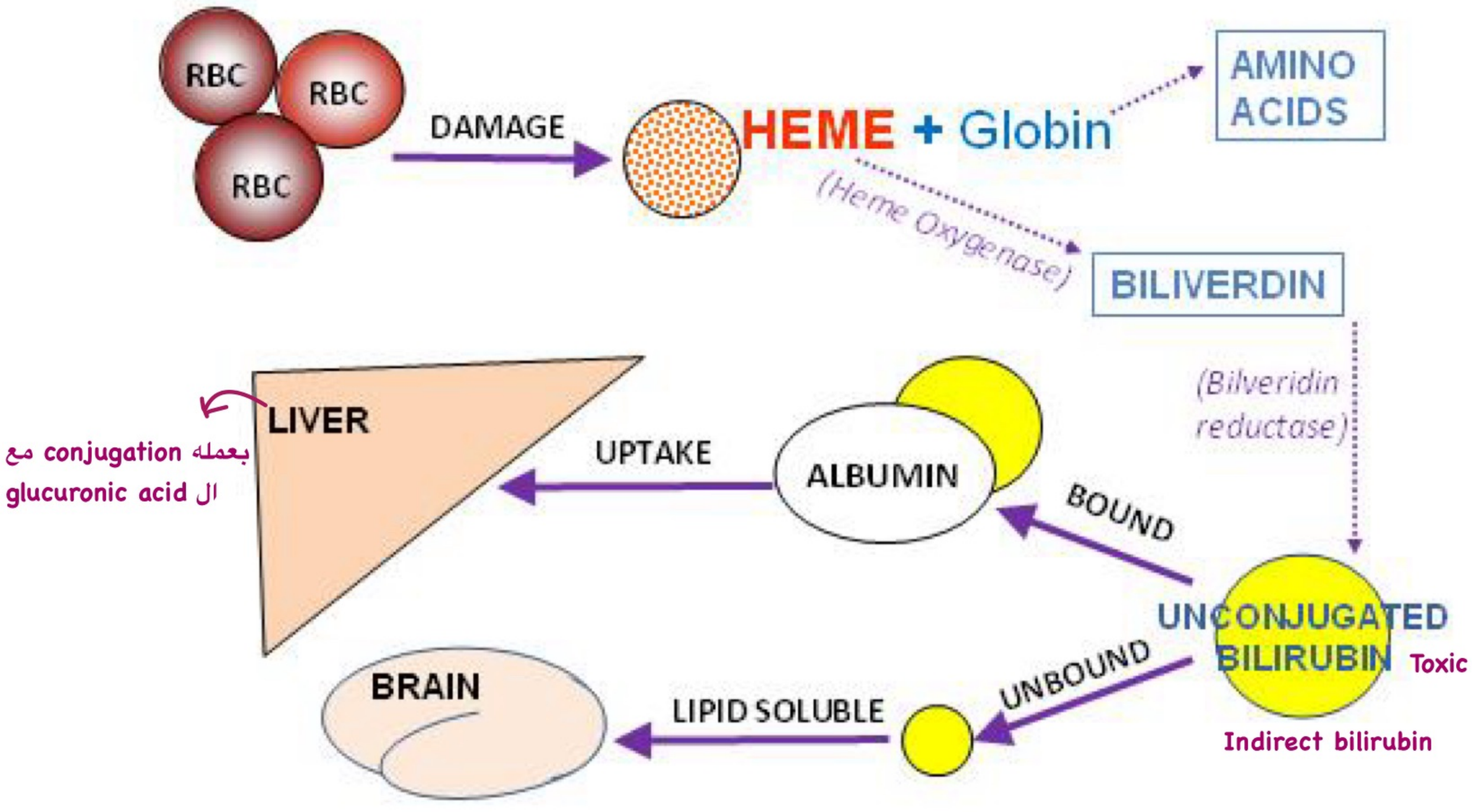
*The arrangement of hemoglobins , fastest to slowest , is A, F, S and C. Hb A2 runs with HbC

Hb electrophoresis



Very important

Bilirubin Metabolism



- لو زادت نسبته عند الأطفال خطر لأنه ممكن يدخل ال BBB و بعمللهم kernicterus