HAEMOLYSIS & HAEMOGLOBINOPATHIES

DR. FATMA AL-QAHTANI

Consultant Haematologist

Hemolysis:

- Premature destruction of RBCs.
- Hemolysis could be due to:
  a. Defect in the RBCs (intra-corpuscular) as in congenital hemolytic Anaemia.
  b. Defect in the surrounding environment (extracorpuscular) as in acquired Anaemia.

Clinical Features of Hemolysis

Pallor, lethargy
Jaundice
Splenomegaly
Gall stones (Pigment – bilirubin)
Dark urine (urobilinogen)
Bone deformity (In some types of haemolytic anaemia)
Leg ulcers (in some types of haemolytic anaemia).

- **Laboratory Features of Hemolysis**

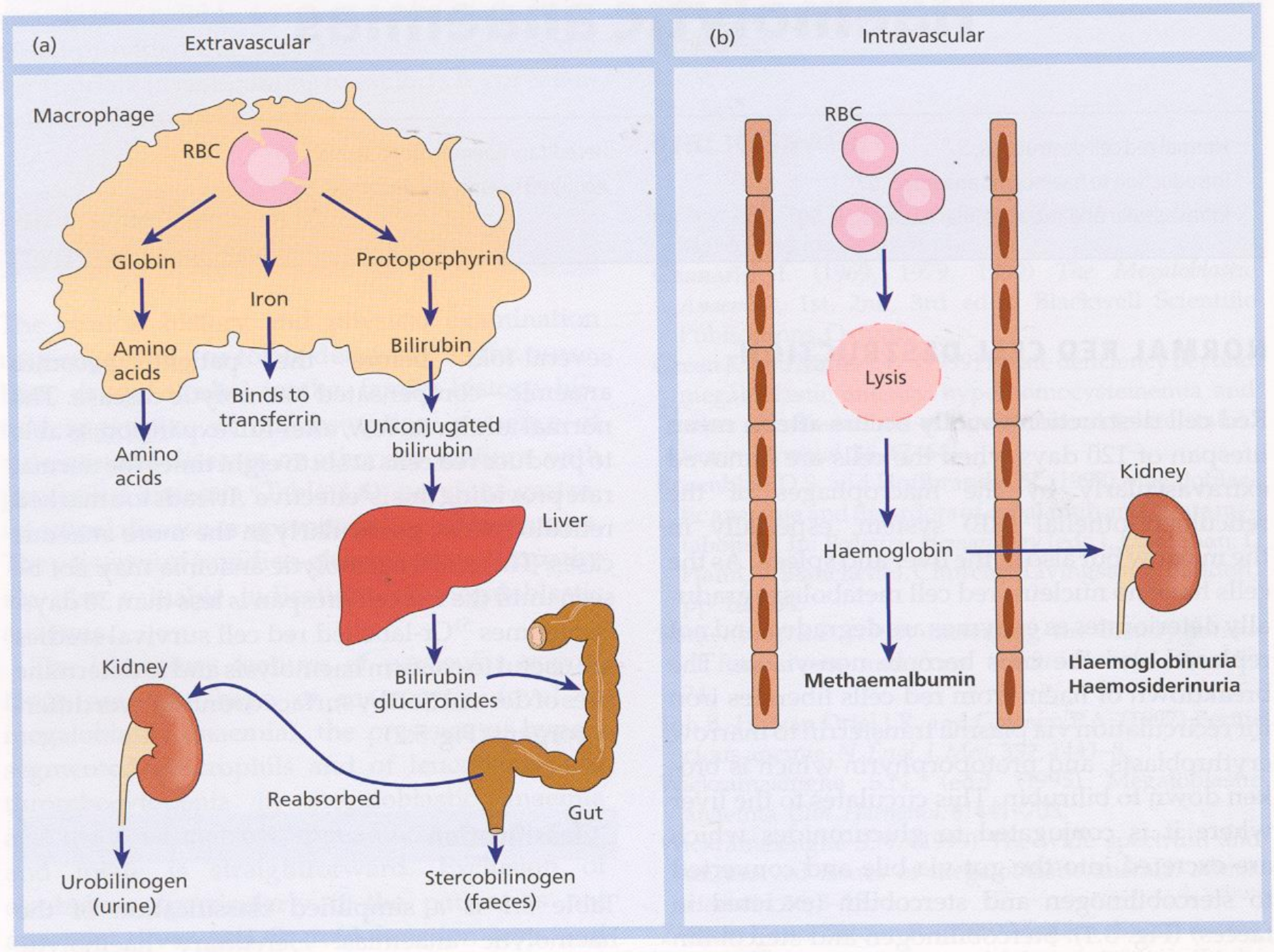
1. **Features of increased red cell breakdown.**
   - a. **↑ serum bilirubin is raised (unconjugated and bound to albumin).**
   - b. **↑ urine urobilinogen.**
   - c. **↑ faecal stercobilinogen.**
   - d. **Absent serum haptoglobins.**
   - e. **↑ lactate dehydrogenase (LDH)**

Laboratory Features of Hemolysis

2. Features of increased red cells production.
   a. Reticulocytosis
   b. Bone marrow erythroid hyperplasia.
3. Damaged red cells.
   a. Morphology (e.g. microspherocytes, elliptocytes, red cells fragmentation).
   b. Increased osmotic fragility, autohaemolysis etc).
   c. Shortened red cell survival (This can be shown by ^{51}Cr labeling with study of the sites of destruction.

Intravascular and extravascular haemolysis.

a. **Intravascular haemolysis, the process of breakdown of red cells directly in the circulation.**
b. **Extravascular haemolysis excessive removal of red cells by cells of RE system in the spleen and liver.**


(a) Extravascular
Macrophage
RBC
Globin
Iron
Protoporphyrin
Amino acids
Binds to transferrin
Bilirubin
Amino acids
Unconjugated bilirubin
Liver
Kidney
Bilirubin glucuronides
Reabsorbed
Gut
Urobilinogen (urine)
Stercobilinogen (faeces)
(b) Intravascular
RBC
Lysis
Haemoglobin
Methaemalbumin
Kidney
Haemoglobinuria
Haemosiderinuria

The main laboratory features of intravascular haemolysis are as follows:

1. **Haemoglobinaemia and haemoglobinuria.**
2. **Haemosiderinuria (Iron storage protein in the spun deposit of urine).**

Causes of intravascular haemolysis

Mismatched blood transfusion (usually ABO)

G6PD deficiency with oxidant stress

Red cell fragmentation syndromes

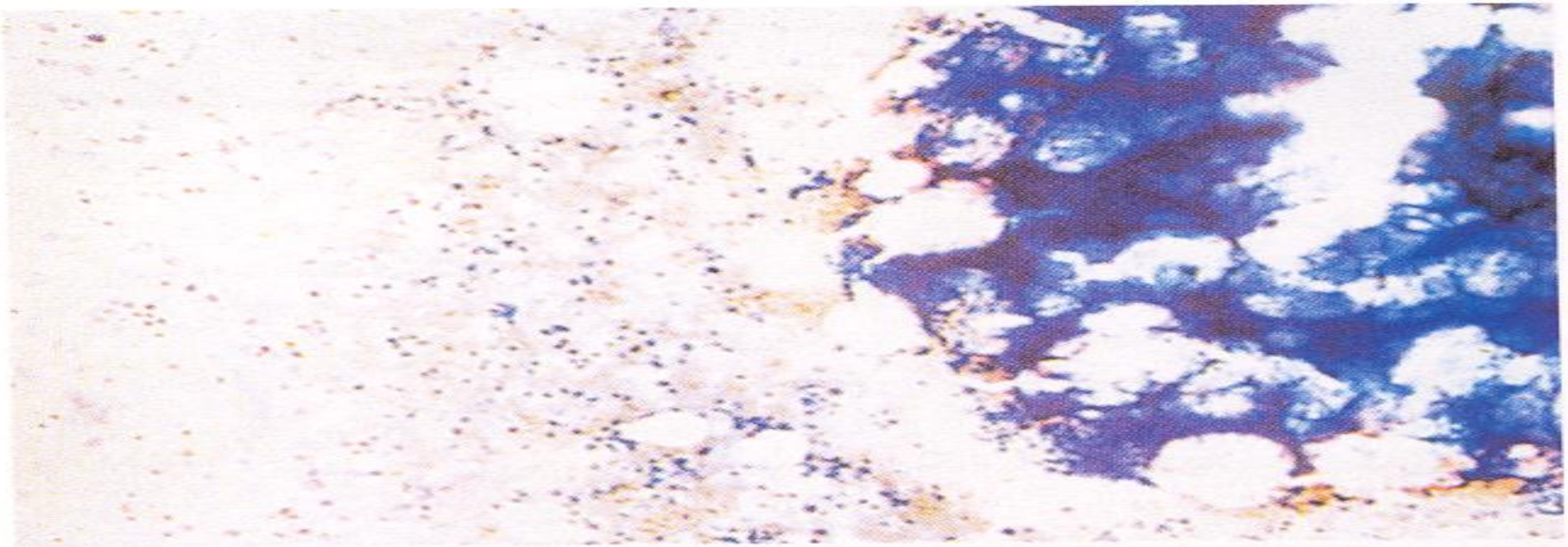
Some autoimmune haemolytic anaemias

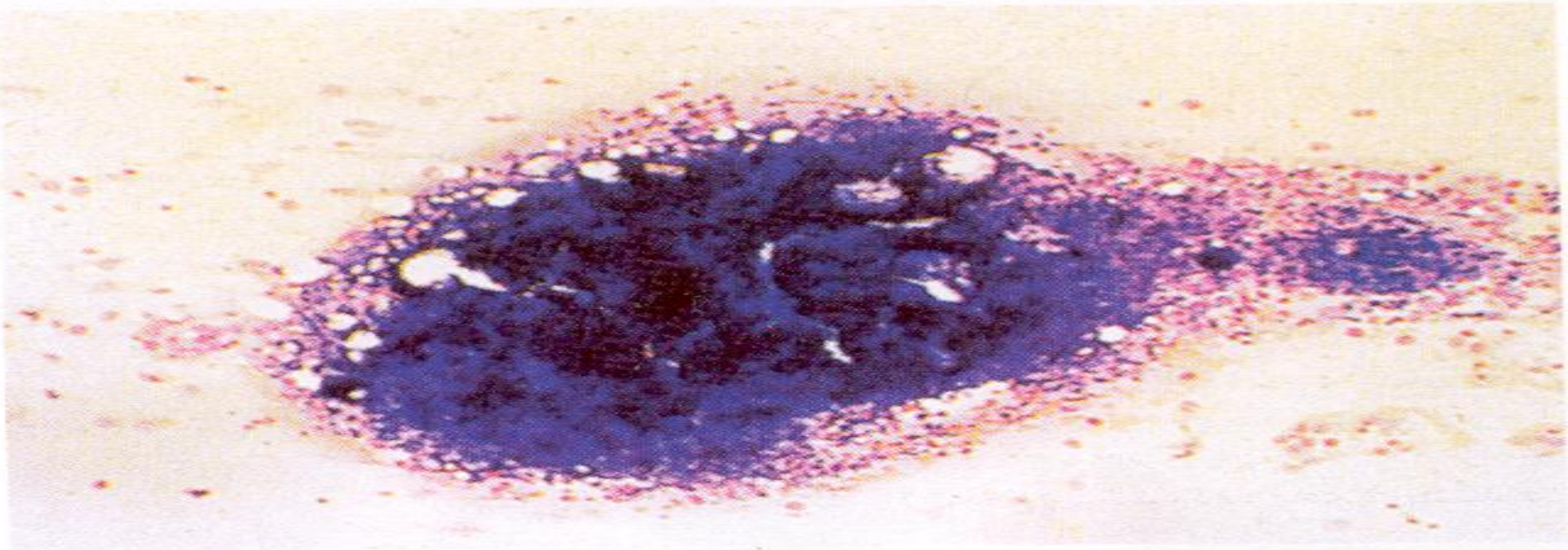
Some drug-and infection-induced haemolytic anaemias

Paroxysmal nocturnal haemoglobinuria

March haemoglobinuria

Unstable haemoglobin





HAEMOLYTIC ANAEMIA

A CONGENITAL

SICKLE CELL DISEASE & OTHER HAEMOGLOBIN DISORDERS

THALASSAEMIAS

ENZYMOPATHIES

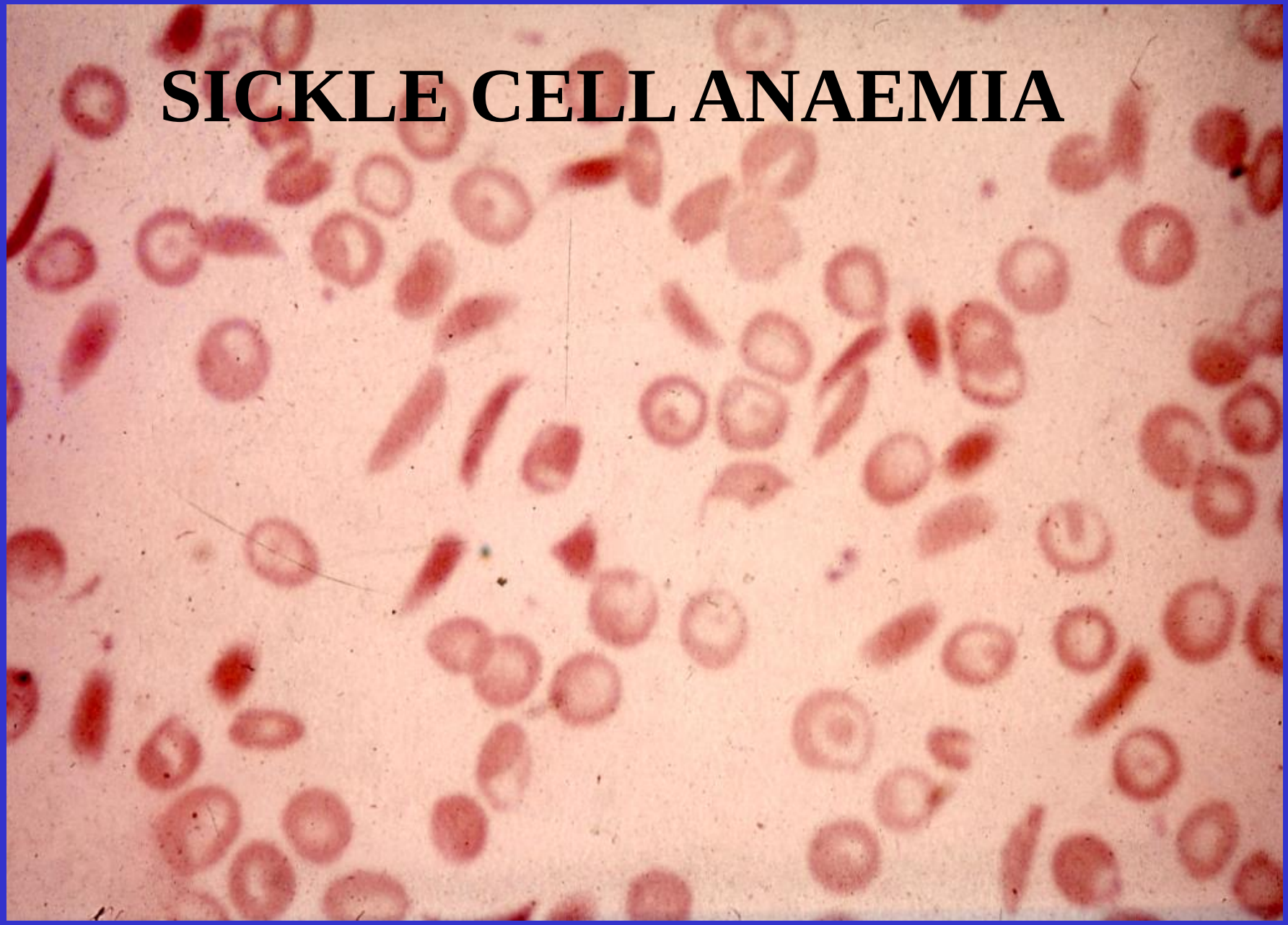
MEMBRANOPATHIES

B AQUIRED

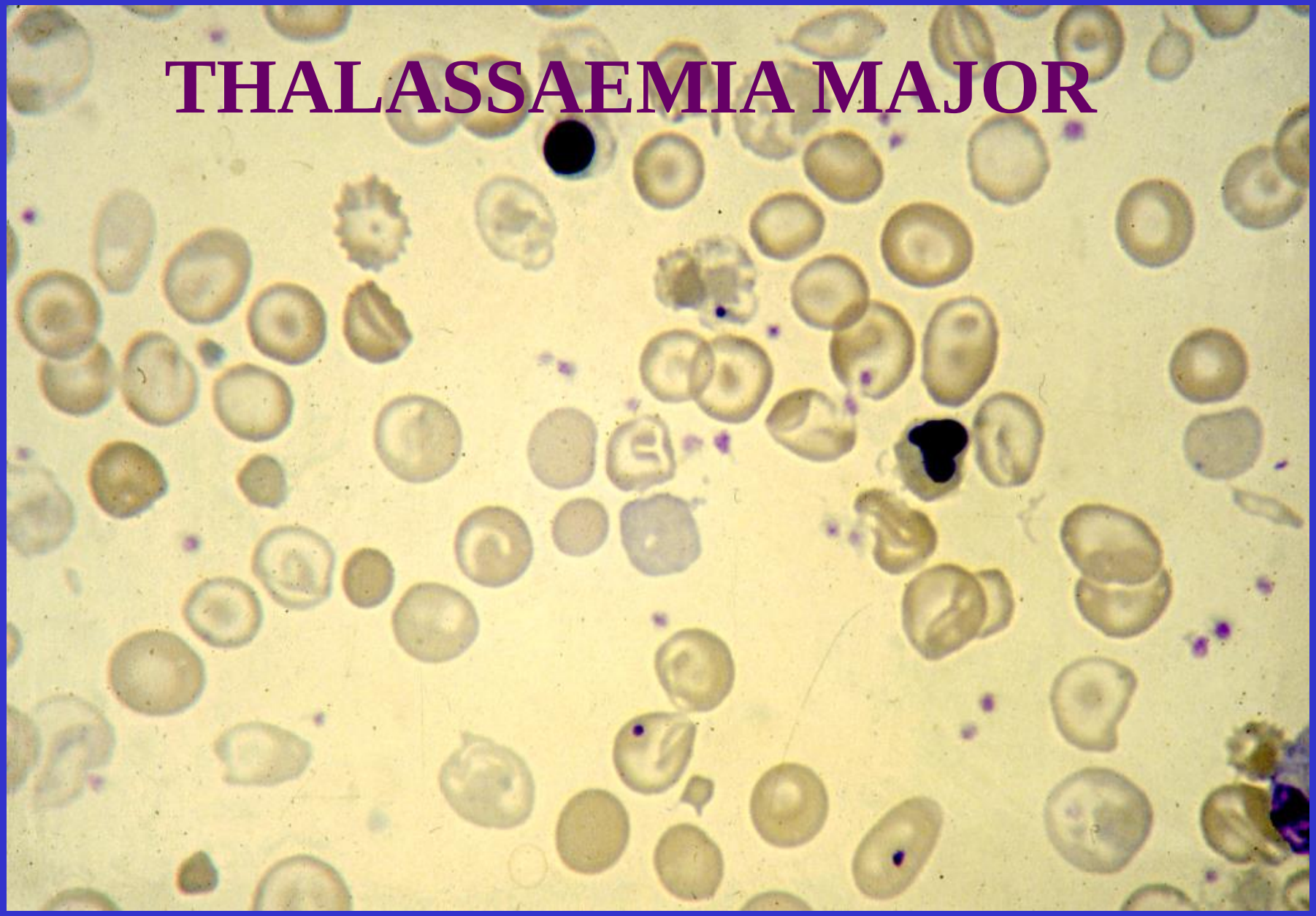
Classification Of Haemolytic Anaemias

Hereditary	Acquired
Haemoglobin	Allografts, especially marrow transplantation
Abnormal (Hb S, Hb C, unstable)	drug associated
Thalassaemia	**Red cell fragmentation syndrome**
Membranopathy	Arterial grafts, cardiac valves
Enzymopathy	Microangiopathic
	Thrombotic thrombocytopenic purpura
	Haemolytic uraemic syndrome
	Meningococcal sepsis
	Pre-eclampsia
	Disseminated intravascular coagulation
	March haemoglobinuria
	Infections
	Malaria, clostridia
	Chemical and physical agents
	Especially drugs, inductrial/domestic substances, burns
	Secondary
	Liver and renal disease
	Paroxysmal nocturnal haemoglobinuria

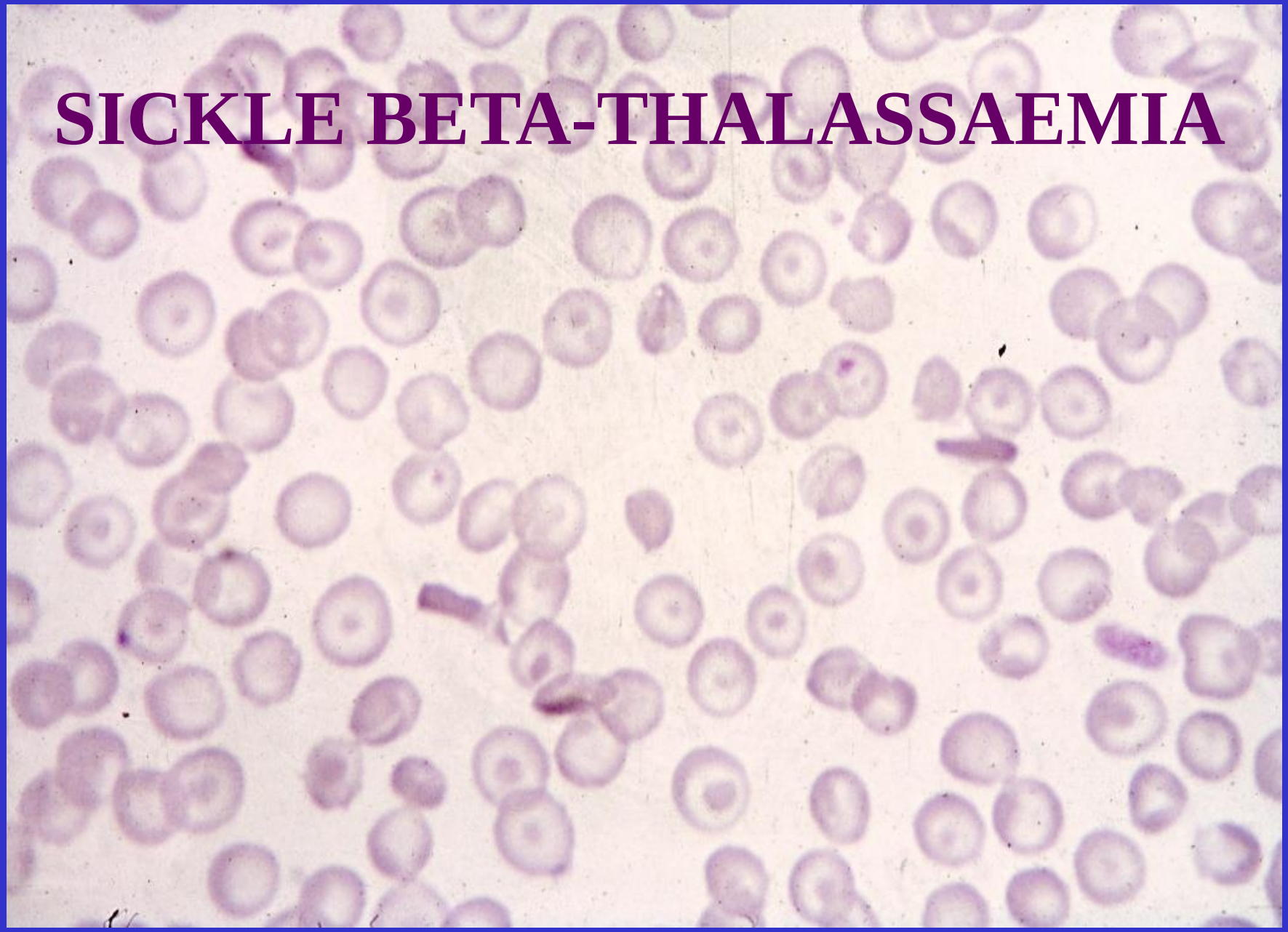
SICKLE CELL ANAEMIA



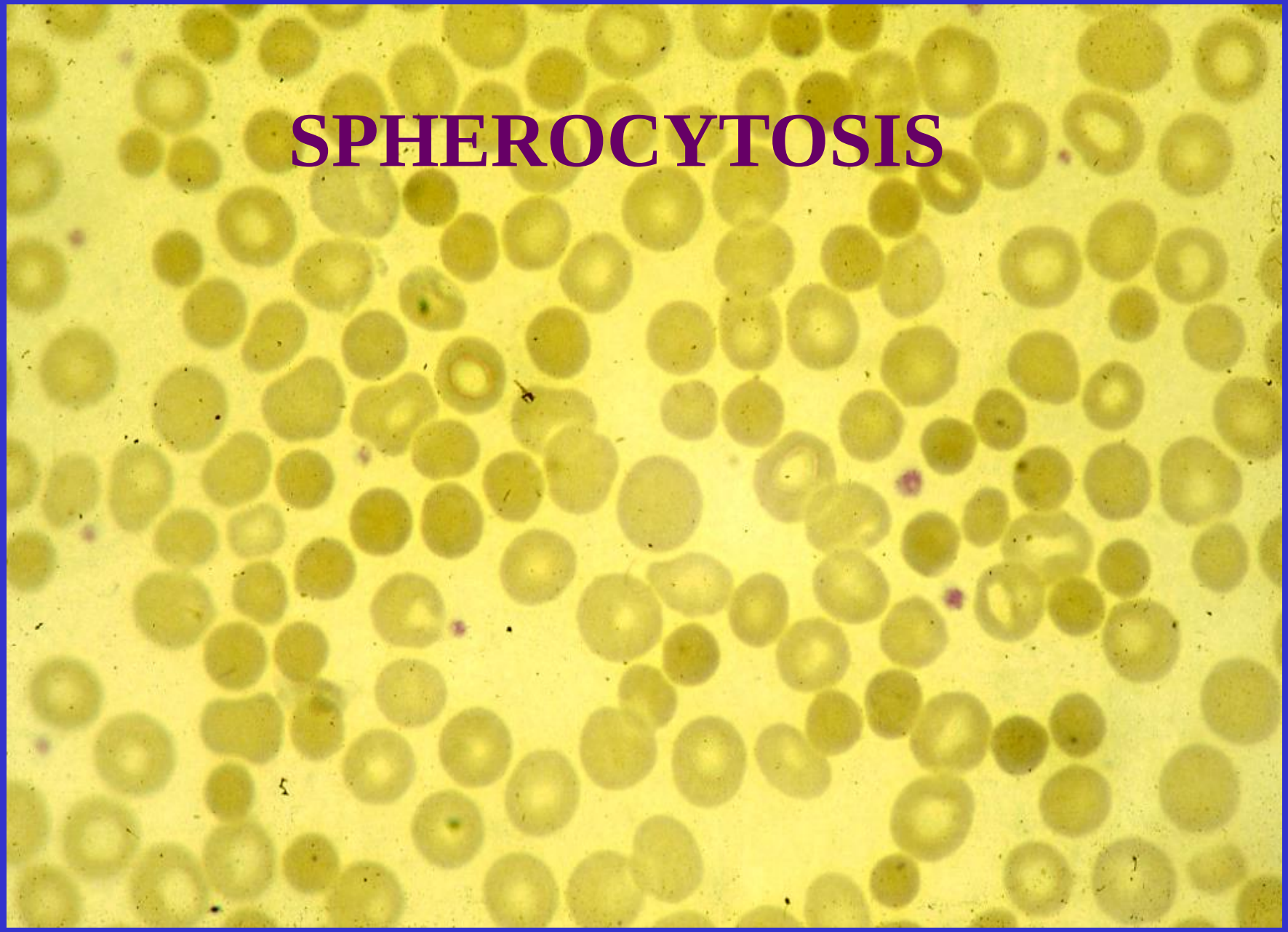
THALASSAEMIA MAJOR



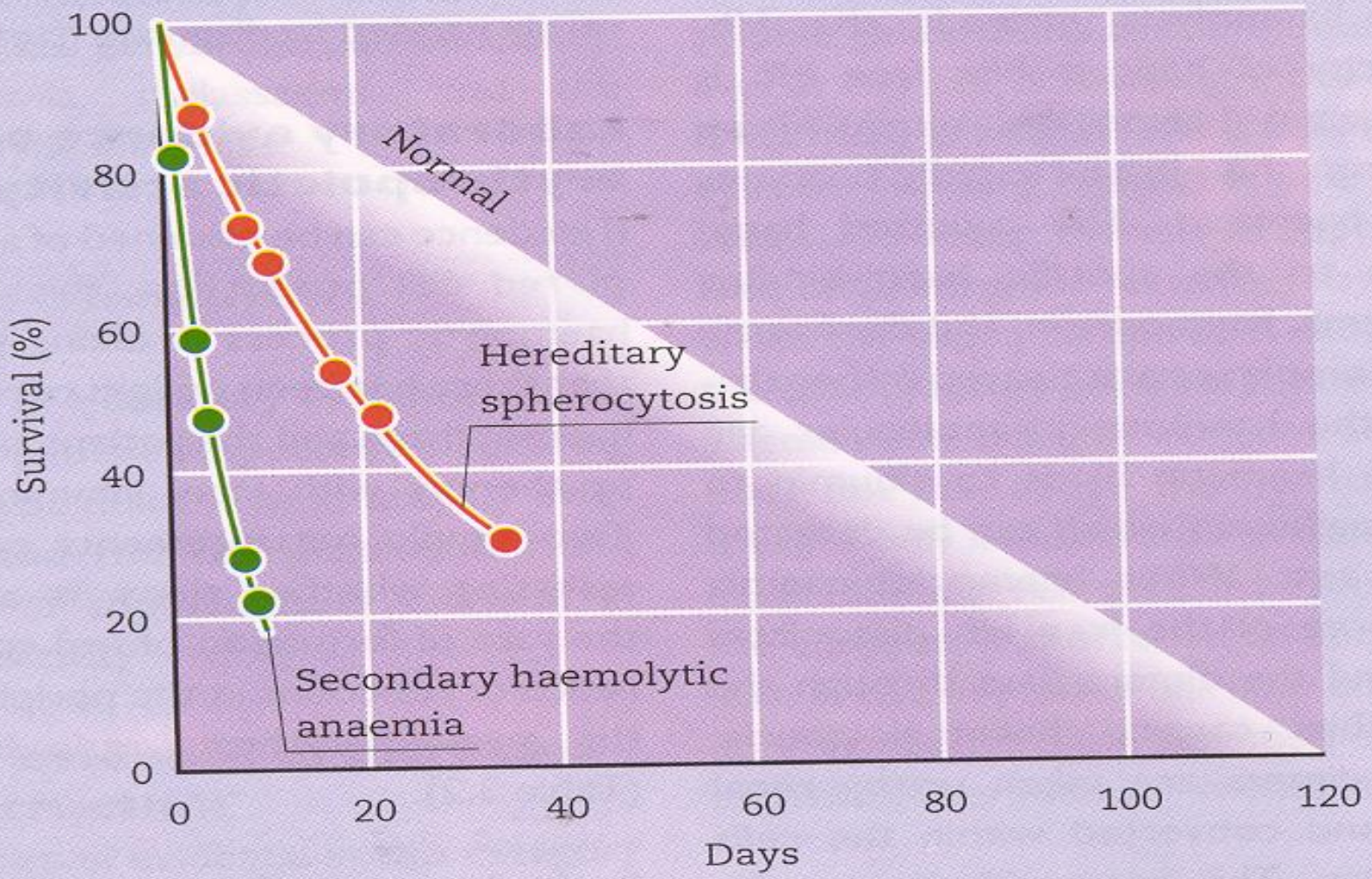
SICKLE BETA-THALASSAEMIA

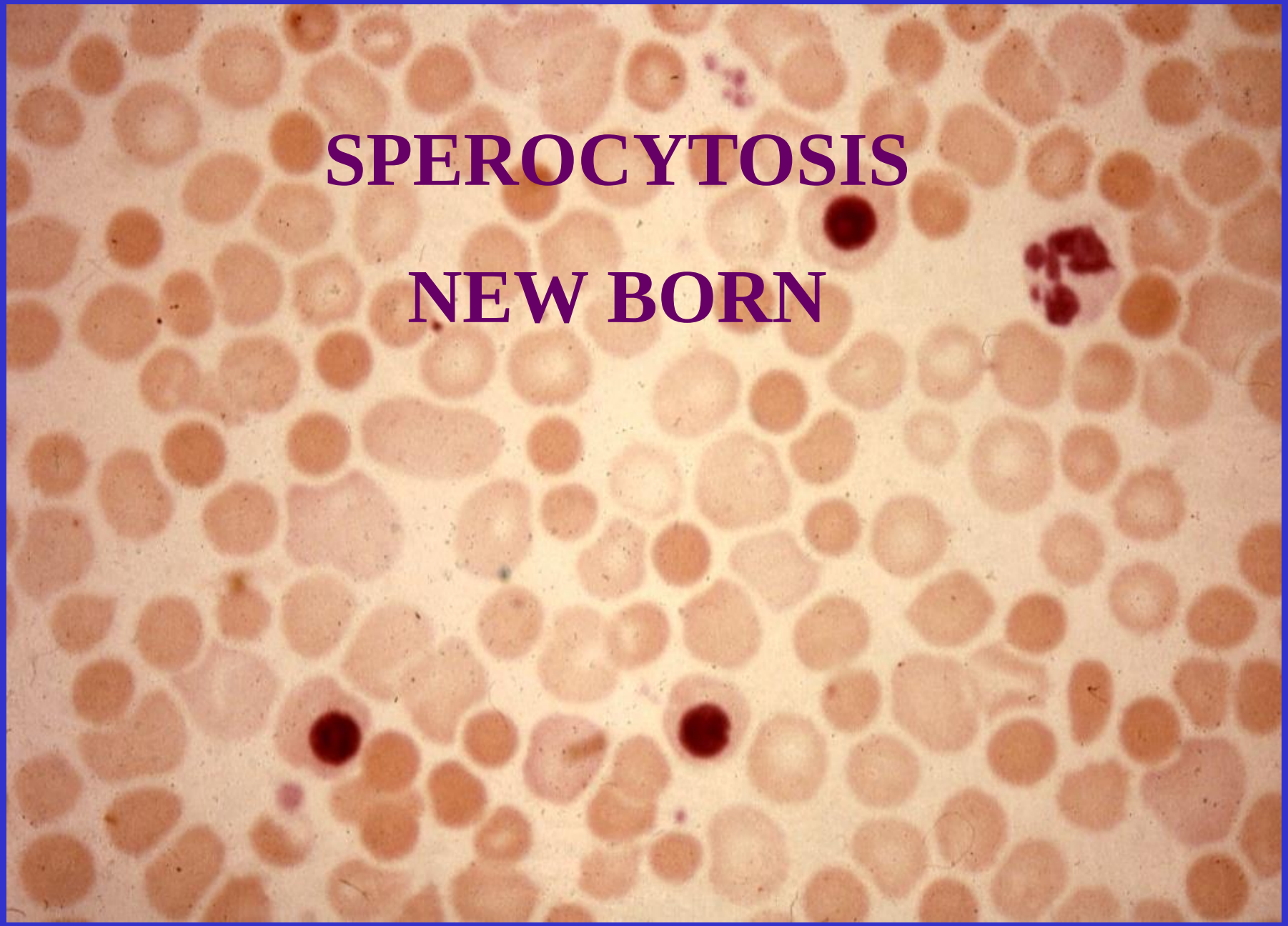


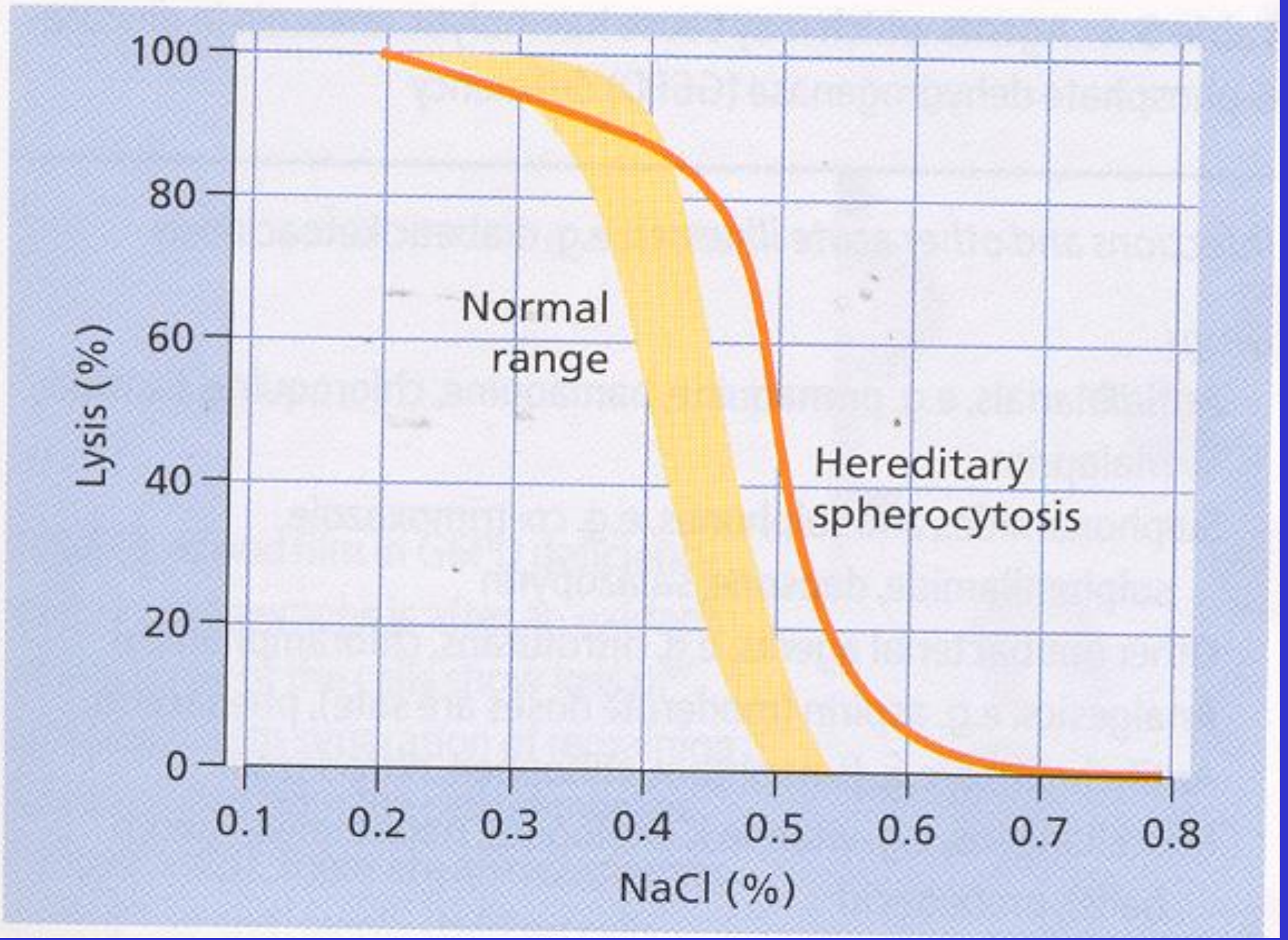
SPHEROCYTOSIS



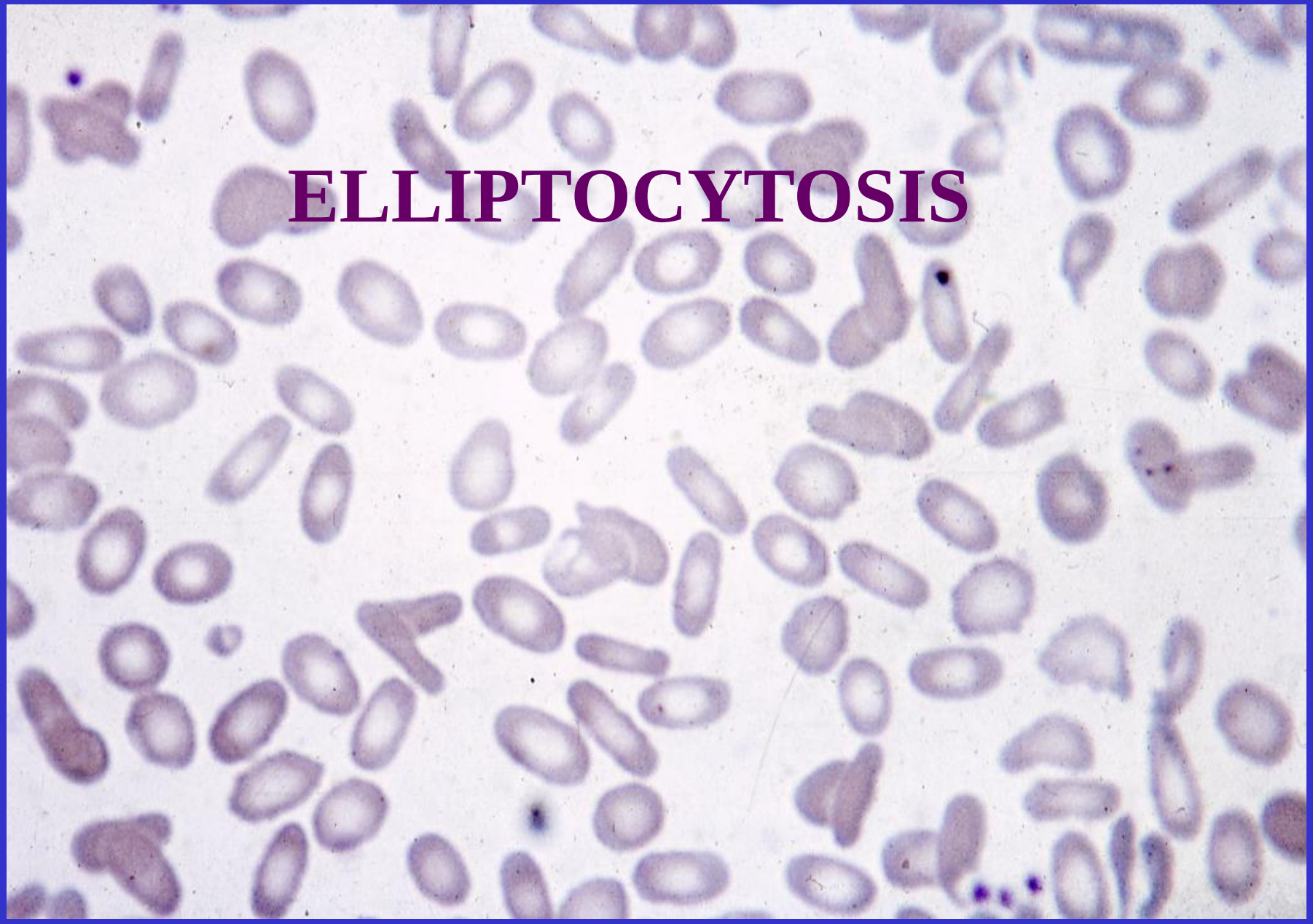
RED CELL SURVIVAL MEASUREMENTS



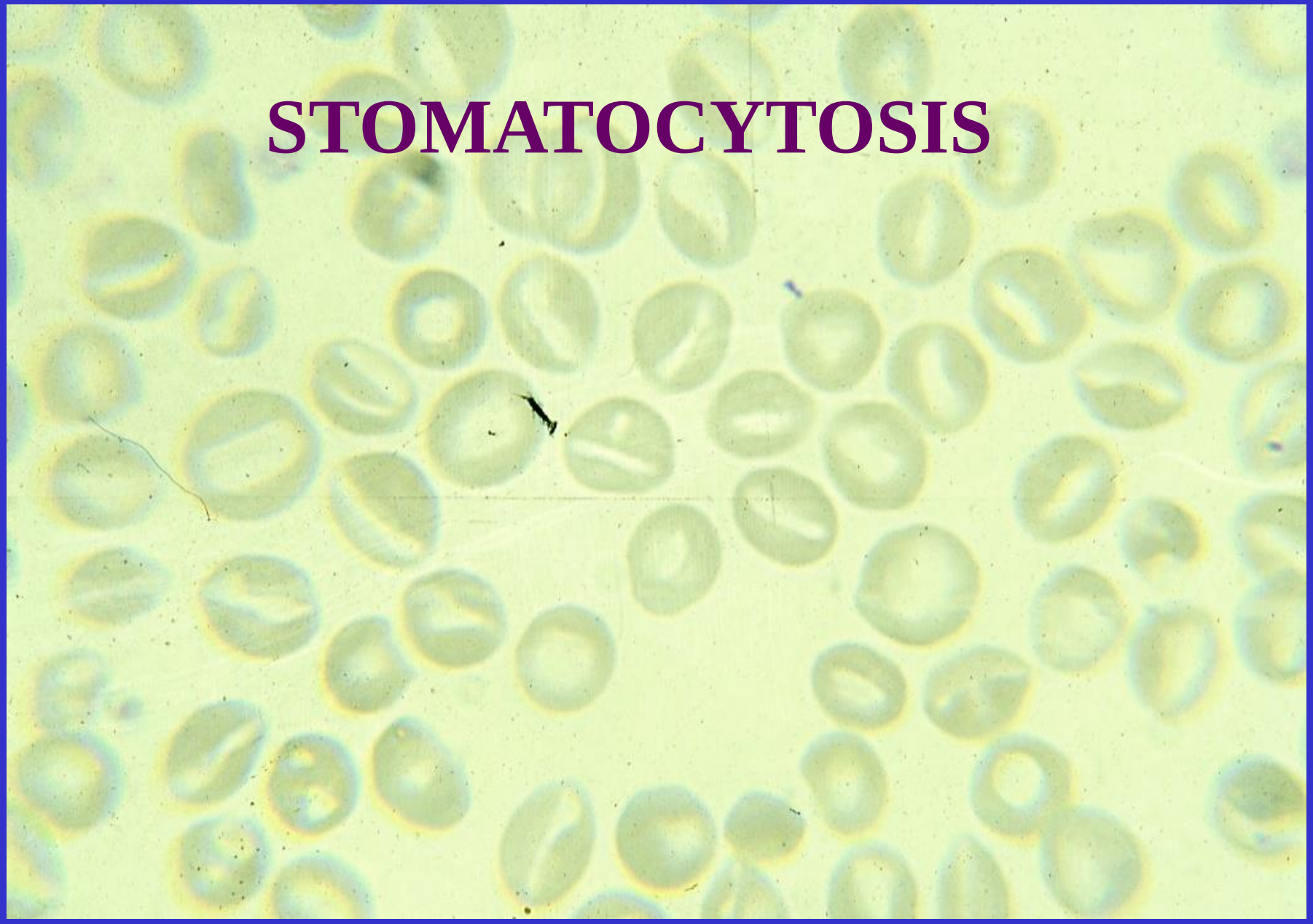

SPEROCYTOSIS
NEW BORN

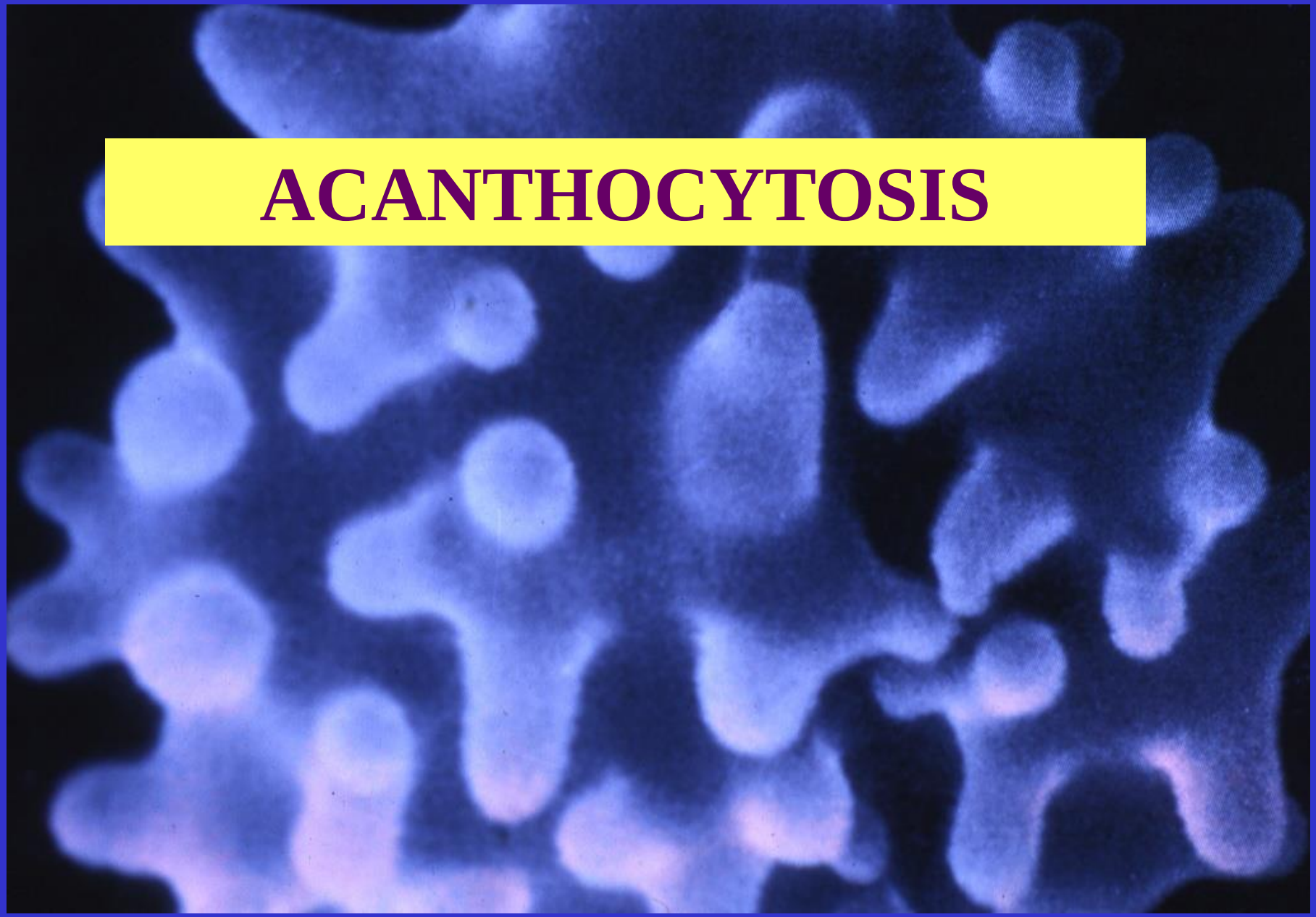

100
80
60
40
20
0
Lysis (%)
Normal
range
Hereditary
spherocytosis
0.1
0.2
0.3
0.4
0.5
0.6
0.7
0.8
NaCl (%)

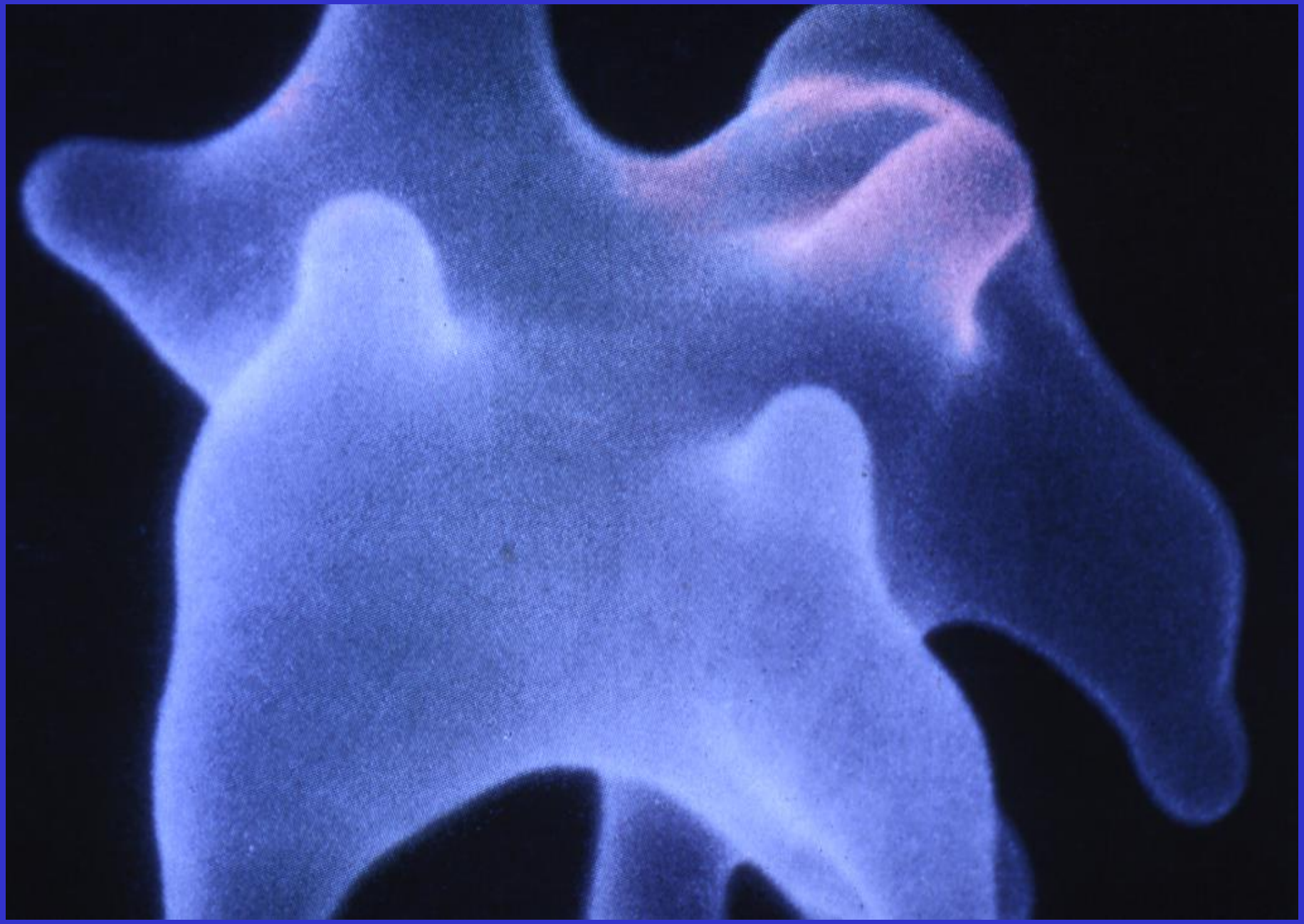
ELLIPTOCYTOSIS



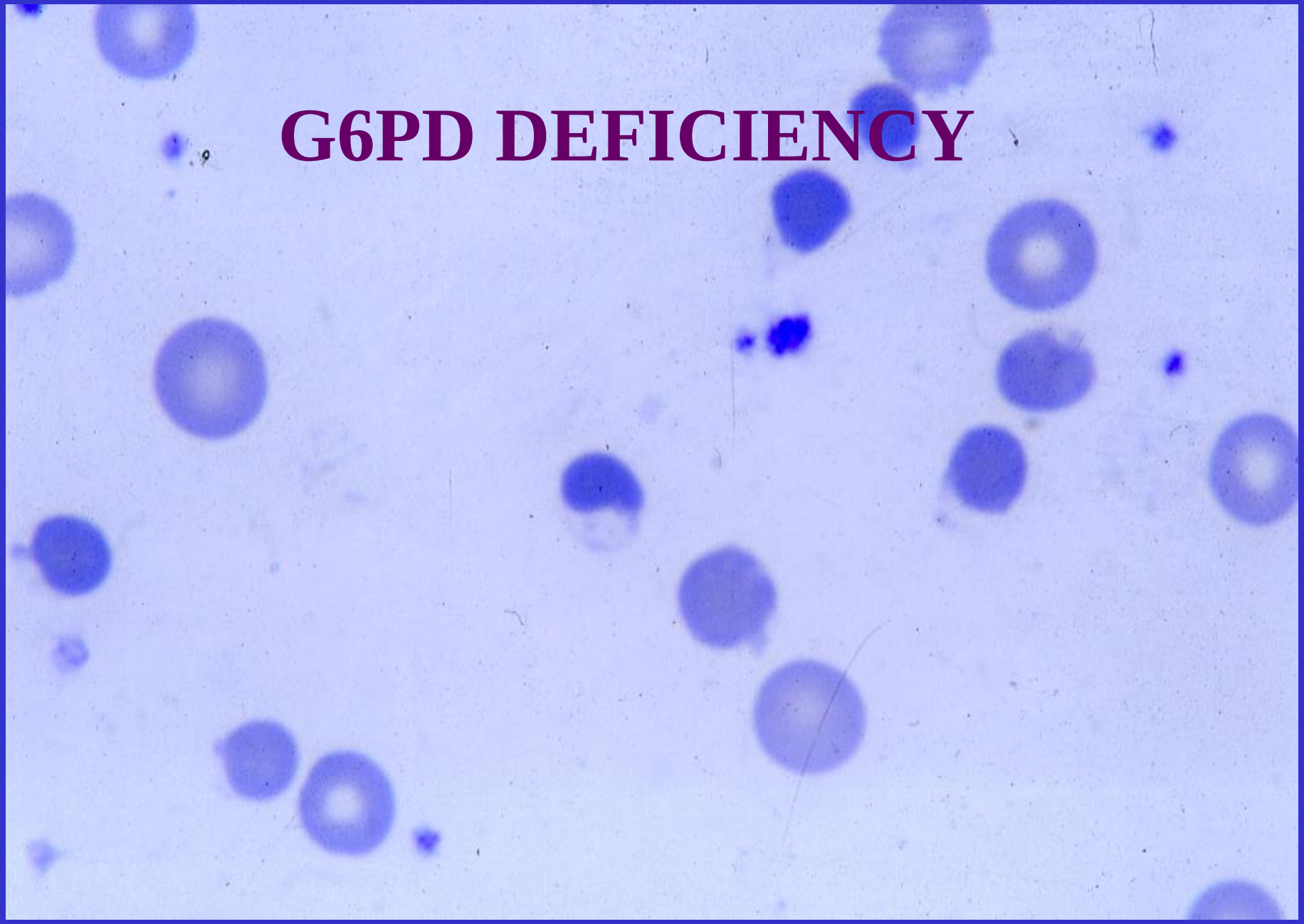
STOMATOCYTOSIS

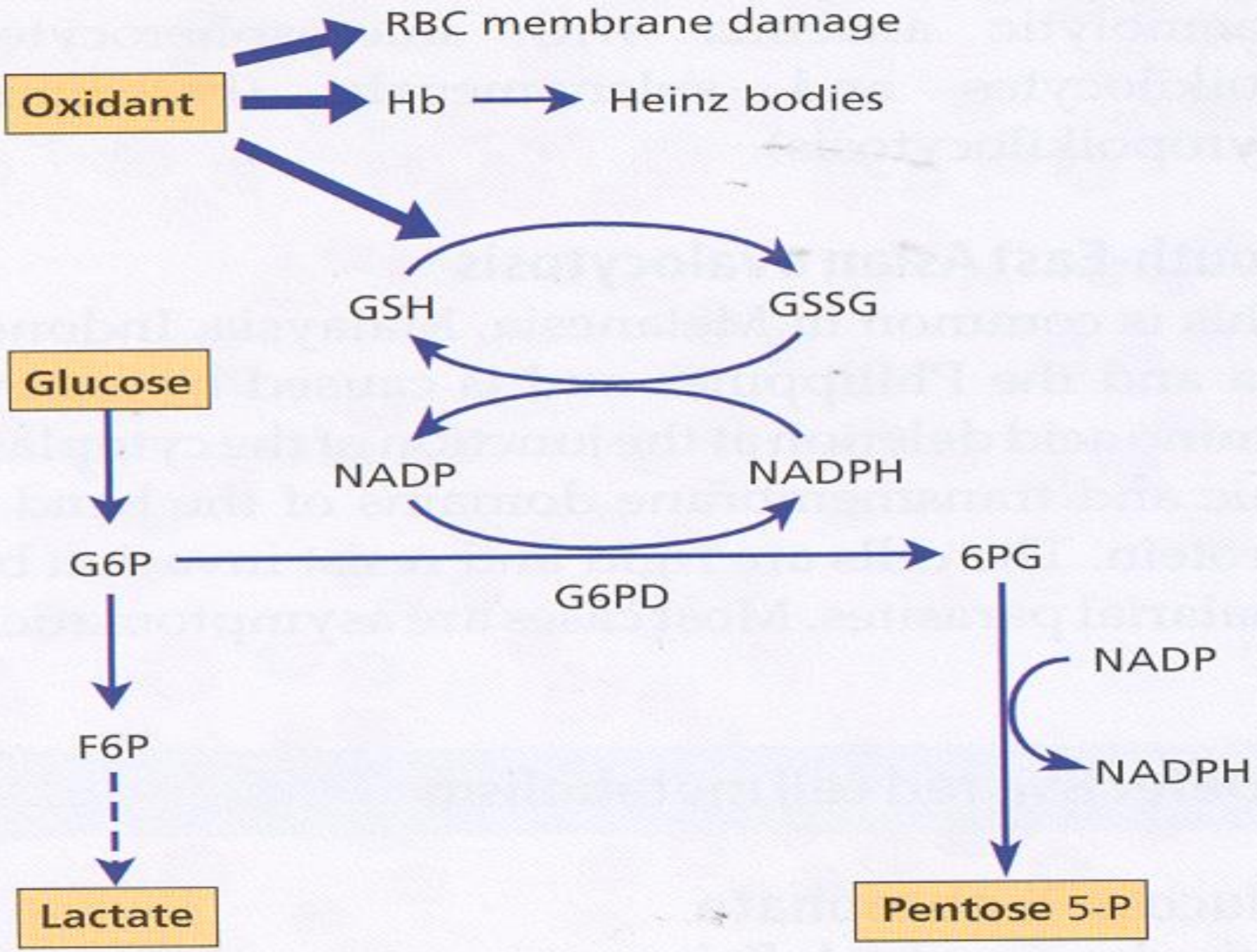



ACANTHOCYTOSIS

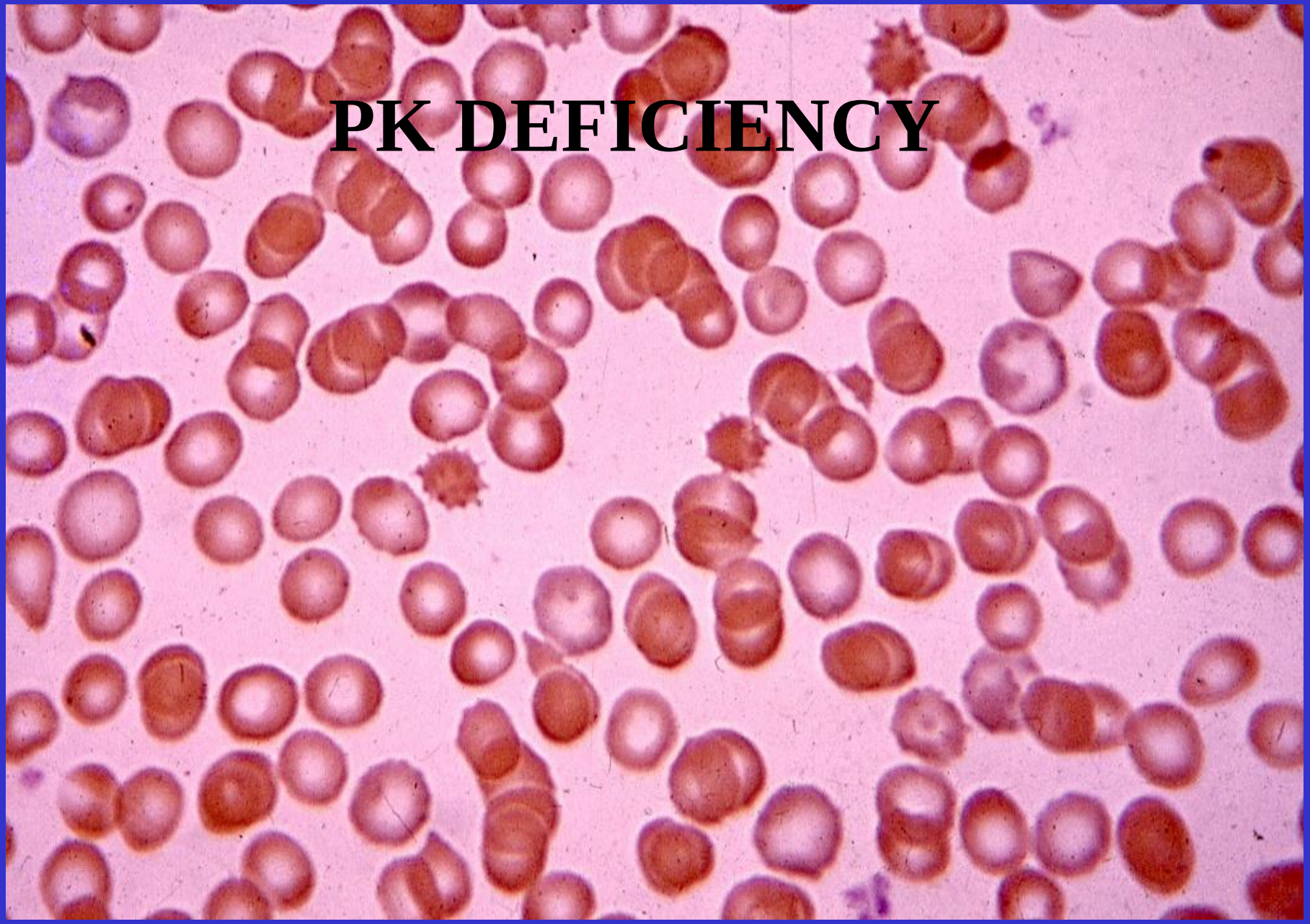


G6PD DEFICIENCY




RBC membrane damage
Oxidant
Hb
Heinz bodies
GSH
GSSG
Glucose
NADP
NADPH
G6P
6PG
G6PD
NADP
NADPH
F6P
Lactate
Pentose 5-P

PK DEFICIENCY



Abnormal haemoglobins (Haemoglobinopathies)

```
 1   2   3   4   5   6   7   8   9   10  11  12  13  14  15
VAL-HIS-LEU-THR-PRO-GLU-GLU-LYS-SER-ALA-VAL-THR-ALA-LEU-TRY

 16  17  18  19  20  21  22  23  24  25  26  27  28  29  30
GLY-LYS-VAL-ASN-VAL-ASP-GLU-VAL-GLY-GLY-GLU-ALA-LEU-GLY-ARG

 31  32  33  34  35  36  37  38  39  40  41  42  43  44  45
LEU-LEU-VAL-VAL-TYR-PRO-TRY-THR-GLN-ARG-PHE-PHE-GLU-SER-PHE

 46  47  48  49  50  51  52  53  54  55  56  57  58  59  60
GLY-ASP-LEU-SER-THR-PRO-ASP-ALA-VAL-MET-GLY-ASN-PRO-LYS-VAL

 61  62  63  64  65  66  67  68  69  70  71  72  73  74  75
LYS-ALA-HIS-GLY-LYS-LYS-VAL-LEU-GLY-ALA-PHE-SER-ASP-GLY-LEU

 76  77  78  79  80  81  82  83  84  85  86  87  88  89  90
ALA-HIS-LEU-ASP-ASN-LEU-LYS-GLY-THR-PHE-ALA-THR-LEU-SER-GLU

 91  92  93  94  95  96  97  98  99 100 101 102 103 104 105
LEU-HIS-CYS-ASP-LYS-LEU-HIS-VAL-ASP-PRO-GLU-ASN-PHE-ARG-LEU

106 107 108 109 110 111 112 113 114 115 116 117 118 119 120
LEU-GLY-ASN-VAL-LEU-VAL-CYS-VAL-LEU-ALA-HIS-HIS-PHE-GLY-LYS

121 122 123 124 125 126 127 128 129 130 131 132 133 134 135
GLU-PHE-THR-PRO-PRO-VAL-GLN-ALA-ALA-TYR-GLN-LYS-VAL-VAL-ALA

136 137 138 139 140 141 142 143 144 145 146
GLY-VAL-ALA-ASN-ALA-LEU-ALA-HIS-LYS-TYR-HIS
```

```
 1   2   3   4   5   6   7   8   9   10  11  12  13  14  15
VAL-LEU-SER-PRO-ALA-ASP-LYS-THR-ASN-VAL-LYS-ALA-ALA-TRY-GLY

 16  17  18  19  20  21  22  23  24  25  26  27  28  29  30
LYS-VAL-GLY-ALA-HIS-ALA-GLY-GLU-TYR-GLY-ALA-GLU-ALA-LEU-GLU

 31  32  33  34  35  36  37  38  39  40  41  42  43  44  45
ARG-MET-PHE-LEU-SER-PHE-PRO-THR-THR-LYS-THR-TYR-PHE-PRO-HIS

 46  47  48  49  50  51  52  53  54  55  56  57  58  59  60
PHE-ASP-LEU-SER-HIS-GLY-SER-ALA-GLN-VAL-LYS-GLY-HIS-GLY-LYS

 61  62  63  64  65  66  67  68  69  70  71  72  73  74  75
LYS-VAL-ALA-ASP-ALA-LEU-THR-ASN-ALA-VAL-ALA-HIS-VAL-ASP-ASP

 76  77  78  79  80  81  82  83  84  85  86  87  88  89  90
MET-PRO-ASN-ALA-LEU-SER-ALA-LEU-SER-ASP-LEU-HIS-ALA-HIS-LYS

 91  92  93  94  95  96  97  98  99 100 101 102 103 104 105
LEU-ARG-VAL-ASP-PRO-VAL-ASN-PHE-LYS-LEU-LEU-SER-HIS-CYS-LEU

106 107 108 109 110 111 112 113 114 115 116 117 118 119 120
LEU-VAL-THR-LEU-ALA-ALA-HIS-LEU-PRO-ALA-GLU-PHE-THR-PRO-ALA

121 122 123 124 125 126 127 128 129 130 131 132 133 134 135
VAL-HIS-ALA-SER-LEU-ASP-LYS-PHE-LEU-ALA-SER-VAL-SER-THR-VAL

136 137 138 139 140 141
LEU-THR-SER-LYS-TYR-ARG
```

Some Known Haemoglobin Mutants

NAME	SUBSTITUTION
Hb. S	α2 β2 6 GLU → VAL
Hb. C	α2 β2 6 GLU → LYS
Hb. E	α2 β2 26 GLU → LYS
Hb. O ARAB	α2 β2 121 GLU → LYS
Hb. D PUNJAB	α2 β2 121 GLU →GLN
Hb RIYADH	α2 β2 120 LYS → ASN
Hb. HAMMERSMITH	α2 β2 42 PHE → SER
Hb. N. BALTIMORE	α2 β2 95 LYS → GLU
Hb. KORLE-BU	α2 β2 73 ASP → ASN
Hb. K. WOOLWICH	α2 β2 132 LYS → GLN
Hb. K. IBADAN	α2 β2 46 GLY → GLU
Hb. KÖ LN	α2 β2 98 VAL → MET
Hb. J. BALTIMORE	α2 β2 16 GLY → ASP

Some Known Haemoglobin Mutants

NAME	SUBSTITUTION
Hb. G. PHILADELPHIA	α2 68 ASN → LYS β2
Hb. ZAMBIA	α2 60 LYS → ASN β2
Hb. G. CHINESE	α2 30 GLU → GLN β2
Hb. HASHARON	α2 47 ASP → HIS β2
Hb. J. TONGARIKI	α2 115 ALA → ASP β2
Hb. J. OXFORD	α2 15 GLY → ASP β2
Hb. NORFOLK	α2 57 GLY → ASP β2

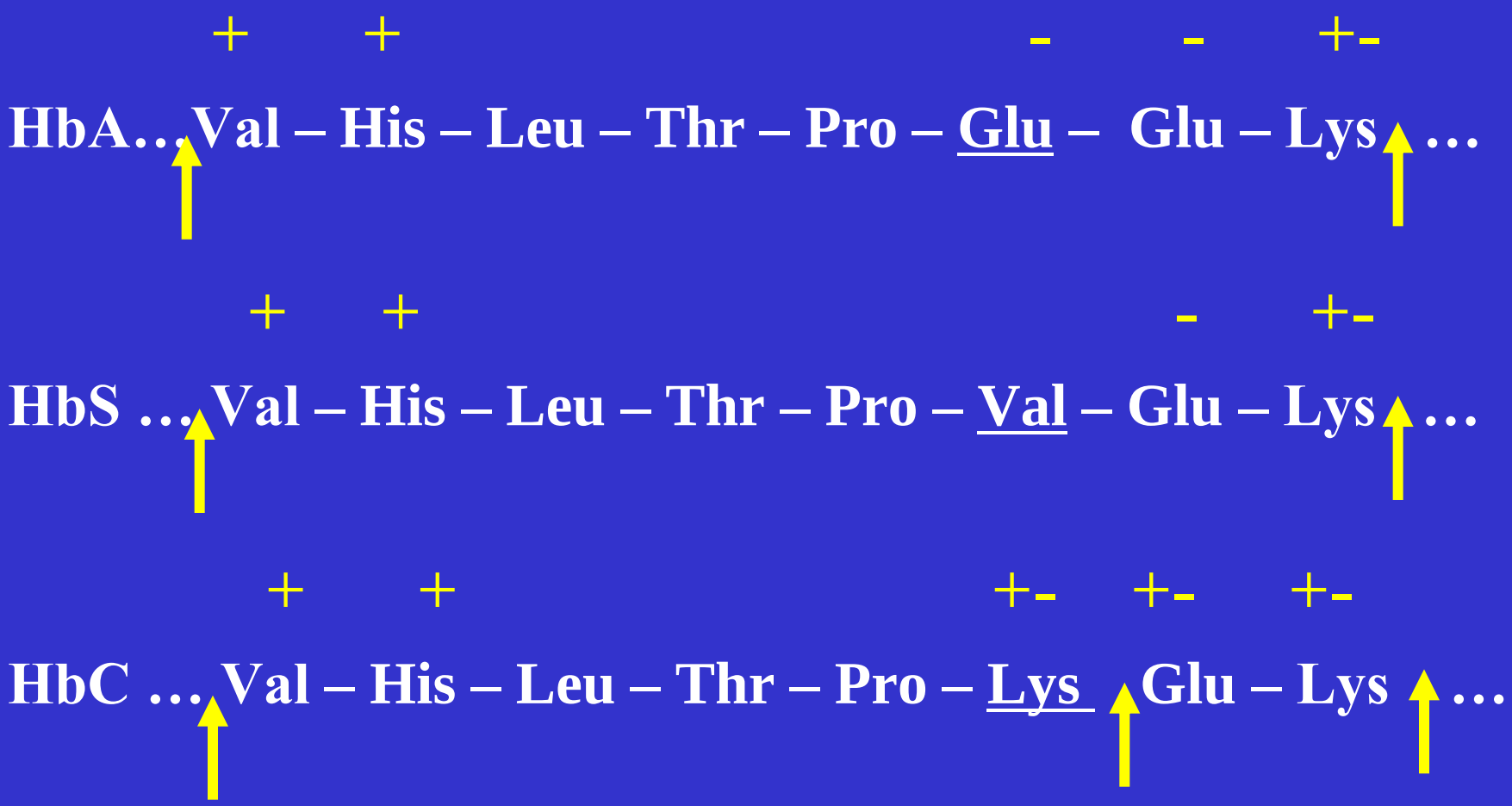
DNA Coding for the Amino-Acid in the sixth position in the β-chain

Normal

	5	6	7
Amino Acid	pro	glu	glu
DNA Base Composition	CCT	G A G	G A G

Sickle

DNA Base composition	CCT	G T G	G A G
Amino Acid	pro	val	glu
	5	6	7



Amino acid sequences of the peptides 4 in haemoglobins A, S and C.

HAEMOGLOBIN VARIANTS: GENE DISTRIBUTION

SICKLE CELL DISEASE

```
 1   2   3   4   5   6   7   8   9   10  11  12  13  14  15
VAL-LEU-SER-PRO-ALA-ASP-LYS-THR-ASN-VAL-LYS-ALA-ALA-TRY-GLY

 16  17  18  19  20  21  22  23  24  25  26  27  28  29  30
LYS-VAL-GLY-ALA-HIS-ALA-GLY-GLU-TYR-GLY-ALA-GLU-ALA-LEU-GLU

 31  32  33  34  35  36  37  38  39  40  41  42  43  44  45
ARG-MET-PHE-LEU-SER-PHE-PRO-THR-THR-LYS-THR-TYR-PHE-PRO-HIS

 46  47  48  49  50  51  52  53  54  55  56  57  58  59  60
PHE-ASP-LEU-SER-HIS-GLY-SER-ALA-GLN-VAL-LYS-GLY-HIS-GLY-LYS

 61  62  63  64  65  66  67  68  69  70  71  72  73  74  75
LYS-VAL-ALA-ASP-ALA-LEU-THR-ASN-ALA-VAL-ALA-HIS-VAL-ASP-ASP

 76  77  78  79  80  81  82  83  84  85  86  87  88  89  90
MET-PRO-ASN-ALA-LEU-SER-ALA-LEU-SER-ASP-LEU-HIS-ALA-HIS-LYS

 91  92  93  94  95  96  97  98  99 100 101 102 103 104 105
LEU-ARG-VAL-ASP-PRO-VAL-ASN-PHE-LYS-LEU-LEU-SER-HIS-CYS-LEU

106 107 108 109 110 111 112 113 114 115 116 117 118 119 120
LEU-VAL-THR-LEU-ALA-ALA-HIS-LEU-PRO-ALA-GLU-PHE-THR-PRO-ALA

121 122 123 124 125 126 127 128 129 130 131 132 133 134 135
VAL-HIS-ALA-SER-LEU-ASP-LYS-PHE-LEU-ALA-SER-VAL-SER-THR-VAL

136 137 138 139 140 141
LEU-THR-SER-LYS-TYR-ARG
```

```
 1   2   3   4   5   6   7   8   9   10  11  12  13  14  15
VAL-HIS-LEU-THR-PRO-GLU-GLU-LYS-SER-ALA-VAL-THR-ALA-LEU-TRY

 16  17  18  19  20  21  22  23  24  25  26  27  28  29  30
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GLY-ASP-LEU-SER-THR-PRO-ASP-ALA-VAL-MET-GLY-ASN-PRO-LYS-VAL

 61  62  63  64  65  66  67  68  69  70  71  72  73  74  75
LYS-ALA-HIS-GLY-LYS-LYS-VAL-LEU-GLY-ALA-PHE-SER-ASP-GLY-LEU

 76  77  78  79  80  81  82  83  84  85  86  87  88  89  90
ALA-HIS-LEU-ASP-ASN-LEU-LYS-GLY-THR-PHE-ALA-THR-LEU-SER-GLU

 91  92  93  94  95  96  97  98  99 100 101 102 103 104 105
LEU-HIS-CYS-ASP-LYS-LEU-HIS-VAL-ASP-PRO-GLU-ASN-PHE-ARG-LEU

106 107 108 109 110 111 112 113 114 115 116 117 118 119 120
LEU-GLY-ASN-VAL-LEU-VAL-CYS-VAL-LEU-ALA-HIS-HIS-PHE-GLY-LYS

121 122 123 124 125 126 127 128 129 130 131 132 133 134 135
GLU-PHE-THR-PRO-PRO-VAL-GLN-ALA-ALA-TYR-GLN-LYS-VAL-VAL-ALA

136 137 138 139 140 141 142 143 144 145 146
GLY-VAL-ALA-ASN-ALA-LEU-ALA-HIS-LYS-TYR-HIS
```

DNA Coding for the Amino-Acid in the sixth position in the β-chain

Normal

	5	6	7
Amino Acid	pro	glu	glu
DNA Base Composition	CCT	G A G	G A G

Sickle

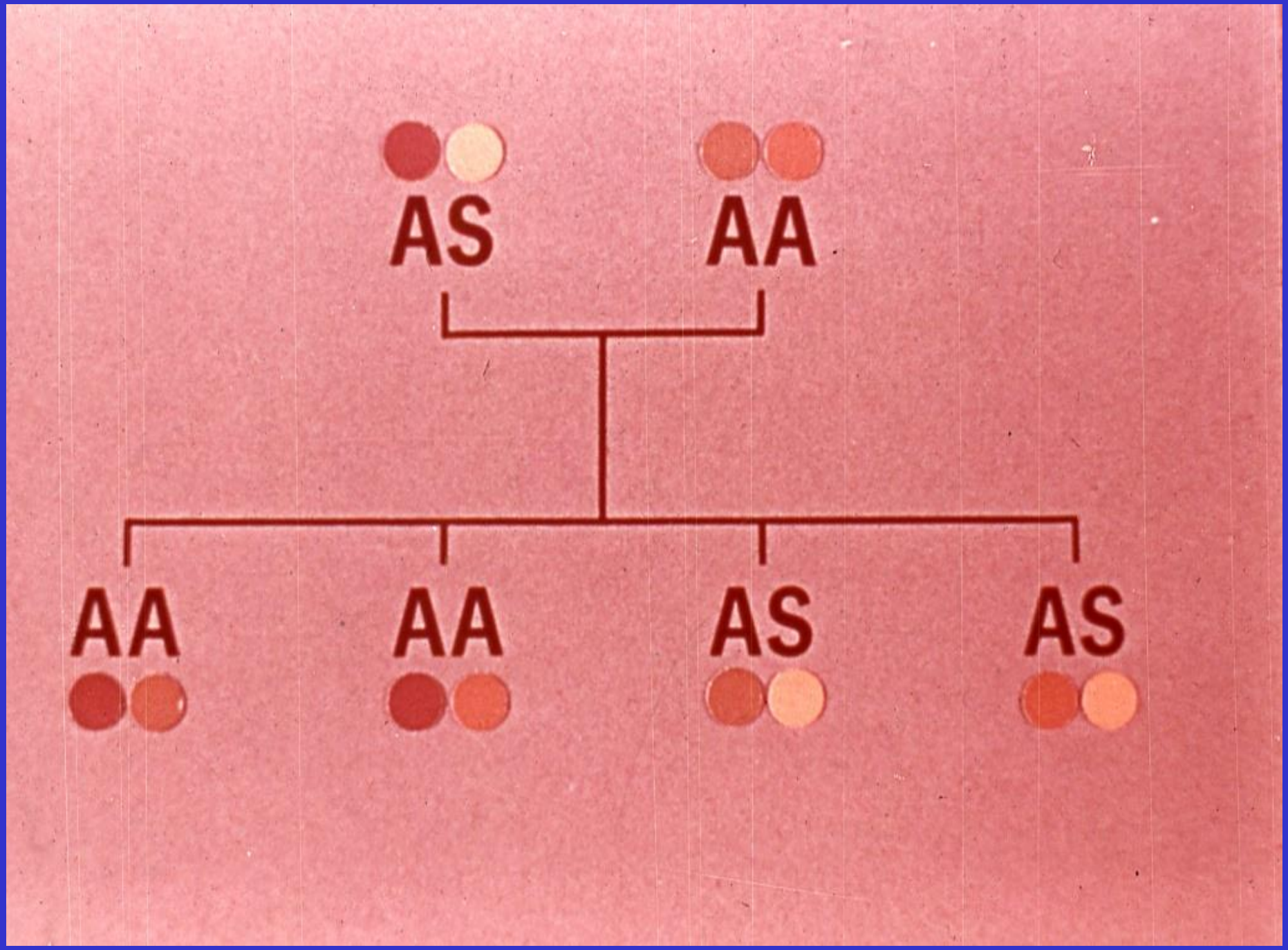
	5	6	7
DNA Base composition	CCT	G T G	G A G
Amino Acid	pro	val	glu

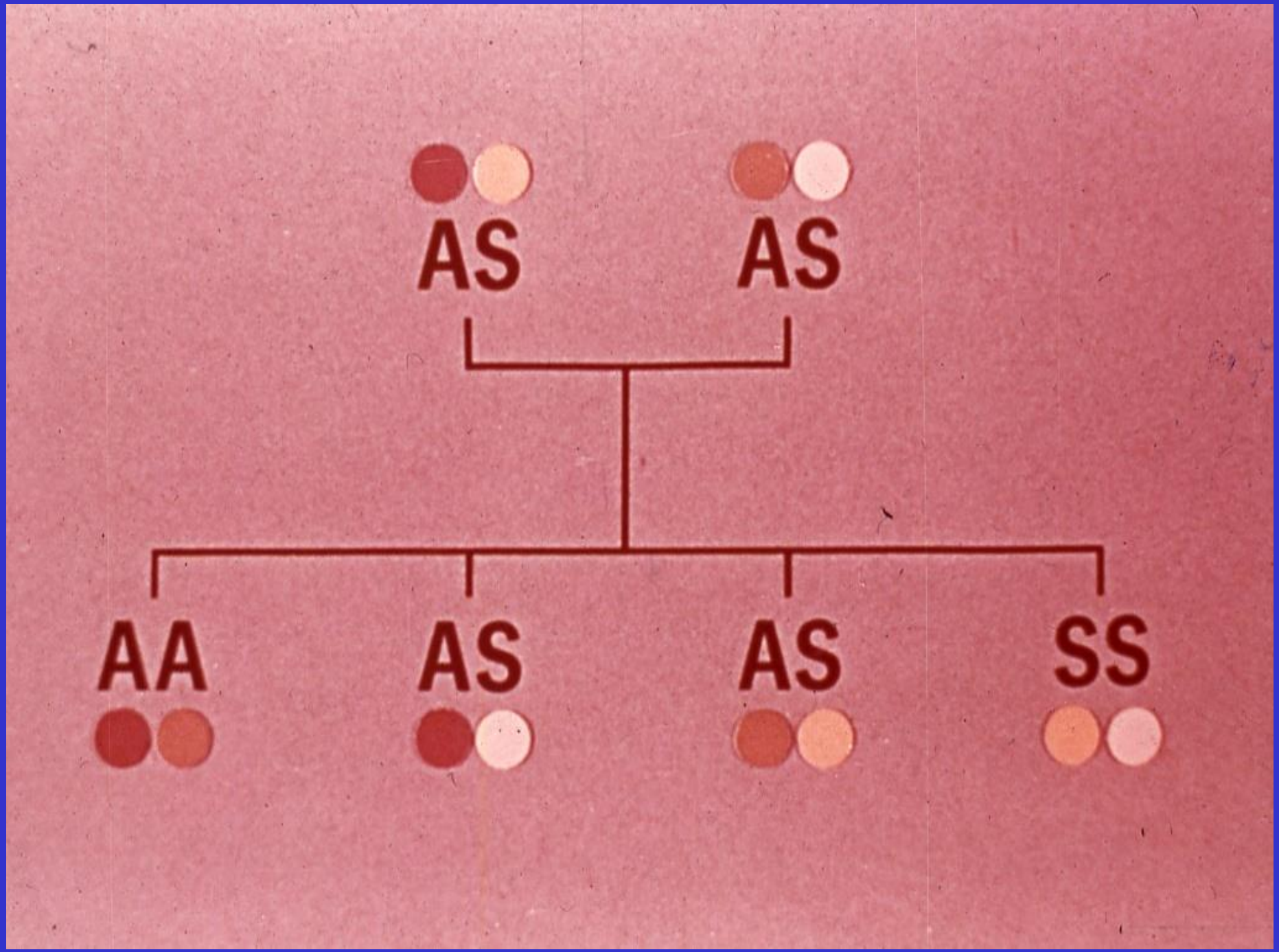
1910 1st published report of sickle cell anaemia (Herrick)

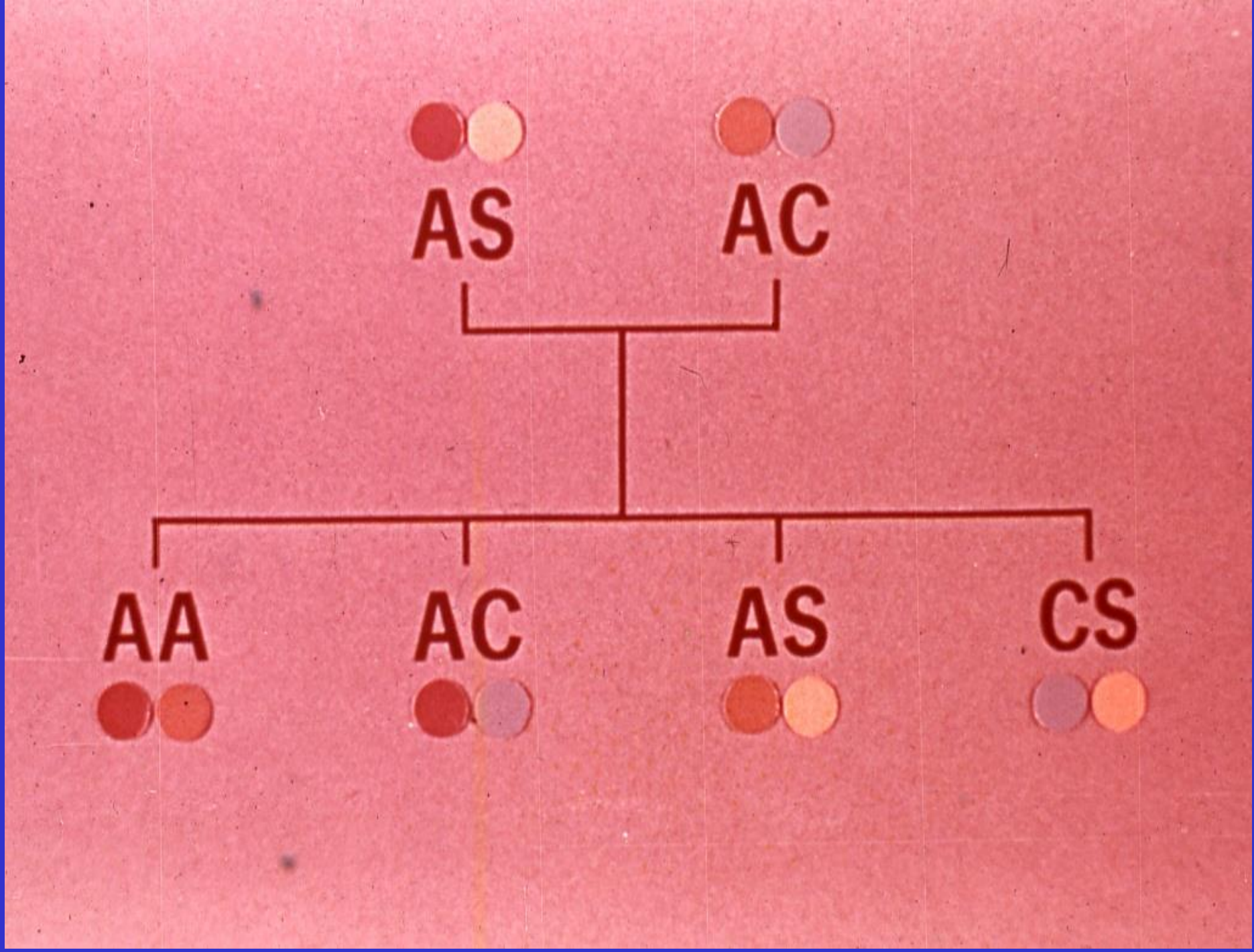
1949 Pauling et al : chemical difference between HbA and HbS

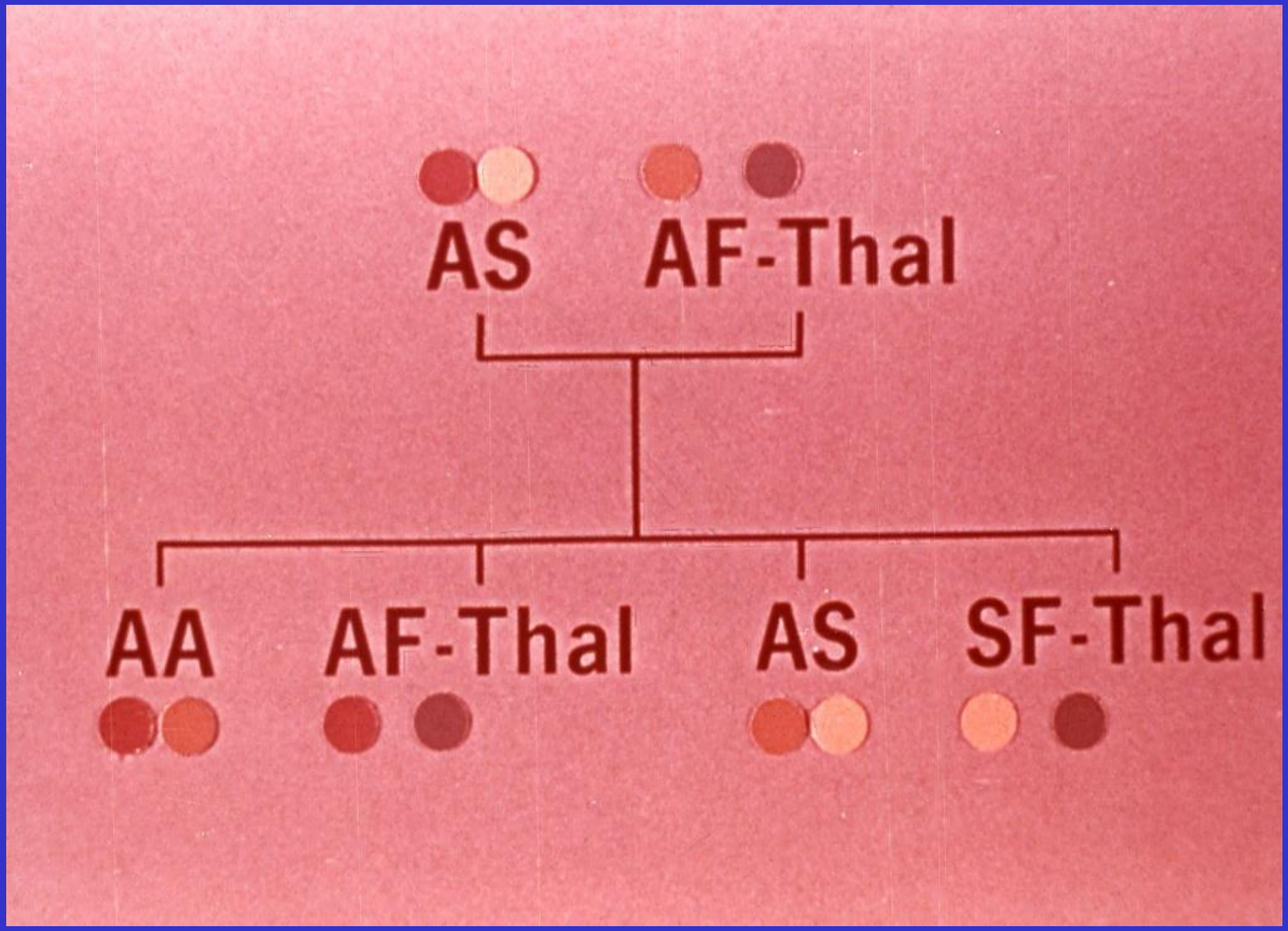
1956 Ingram: Fingerprinting

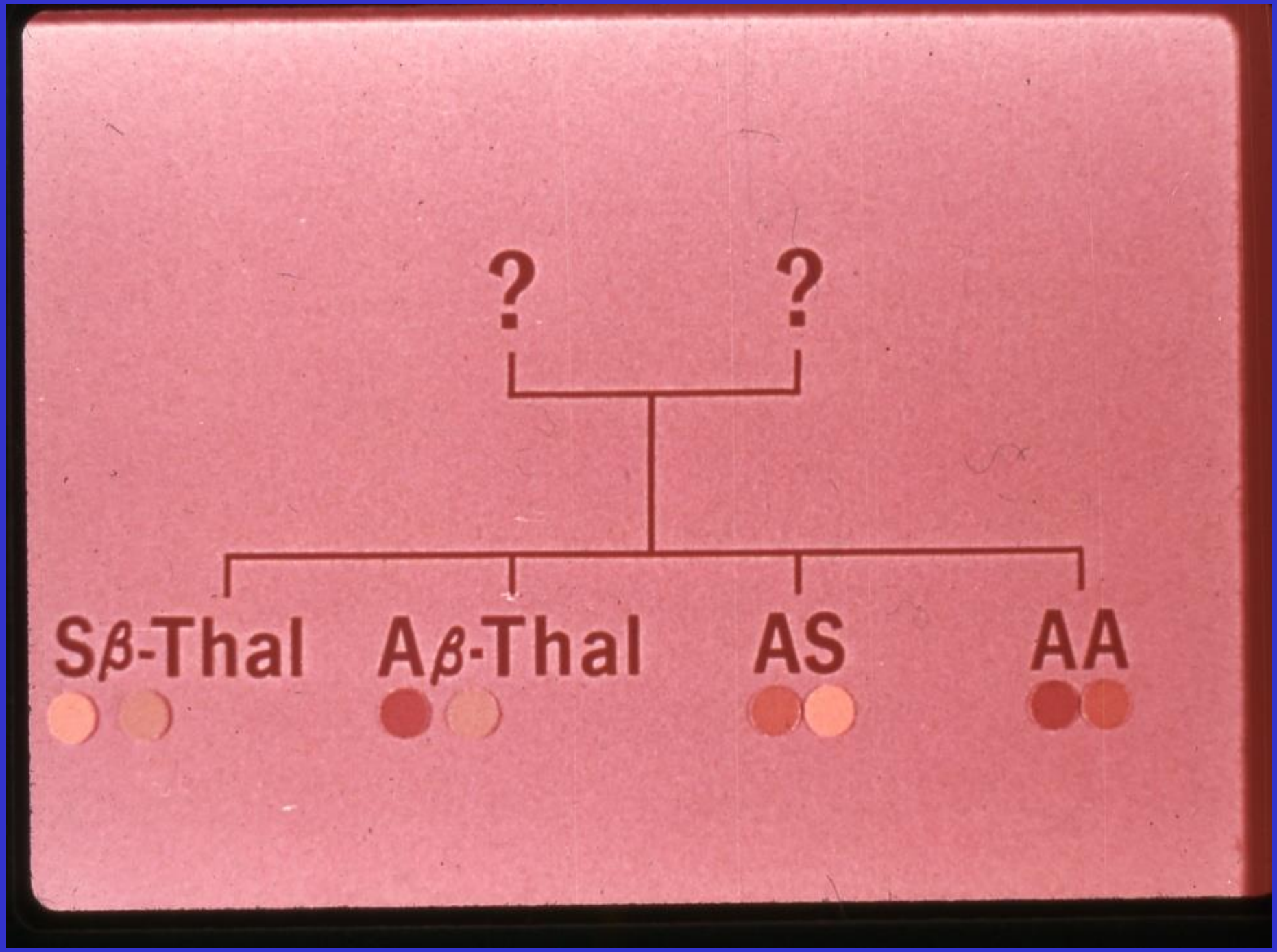
βglu → val

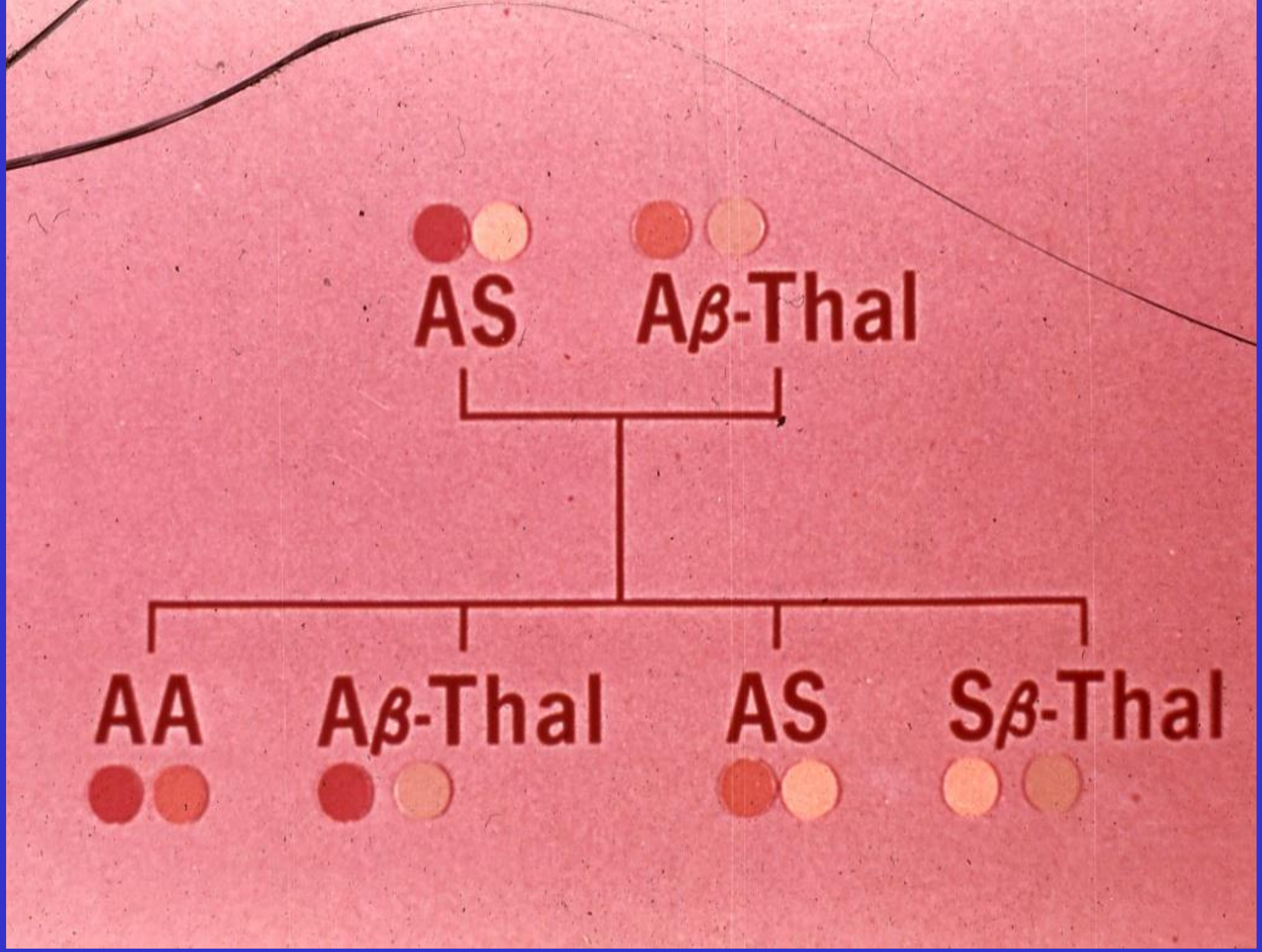

AS
AA
AA
AA
AS
AS


AS
AS
AA
AS
AS
SS


AS
AC
AA
AC
AS
CS


AS
AF-Thal
AA
AF-Thal
AS
SF-Thal


? ?
Sβ-Thal
Aβ-Thal
AS
AA


AS
Aβ-Thal
AA
Aβ-Thal
AS
Sβ-Thal

SICKLE CELL DISEASE

THE SICKLE CELL TRAIT

HOMOZYGOUS SICKLE CELL DISEASE (SS)

Sickle cell anaemia

DOUBLY HETEROZYGOUS SICKLE CELL DISEASE

Sickle cell / haemoglobin C disease

Sickle cell / thalassaemia

PROPERTIES OF HbS

Solubility ↓

Conformational changes – "tactoid formation"
→ sickled cells
→ irreversibly sickled cells

↑ mechanical fragility → haemolysis

↑ viscosity → organ infarction

FACTORS AFFECTING SICKLING

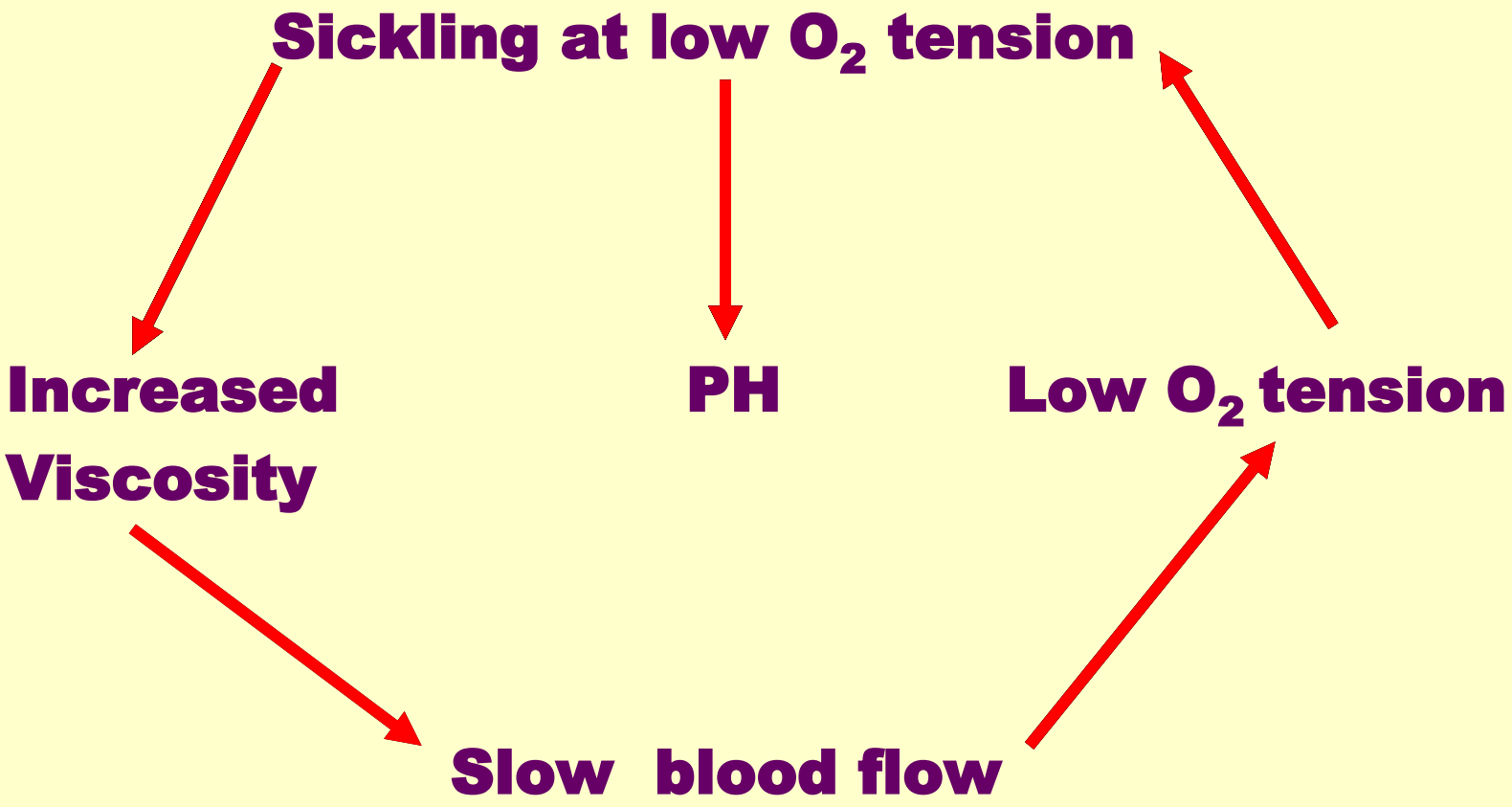
Oxygen tension — 50–60 mm Hg for SS
20–30 mm Hg for AS

pH — inhibited at alkaline pH
exacerbated by acidification

Concentration of Hb S

Presence of other haemoglobins

polymerisation : S > D > C > J = A > F


Sickling at low O_2 tension
Increased Viscosity
PH
Low O_2 tension
Slow blood flow

FACTORS PRECIPITATING CRISES

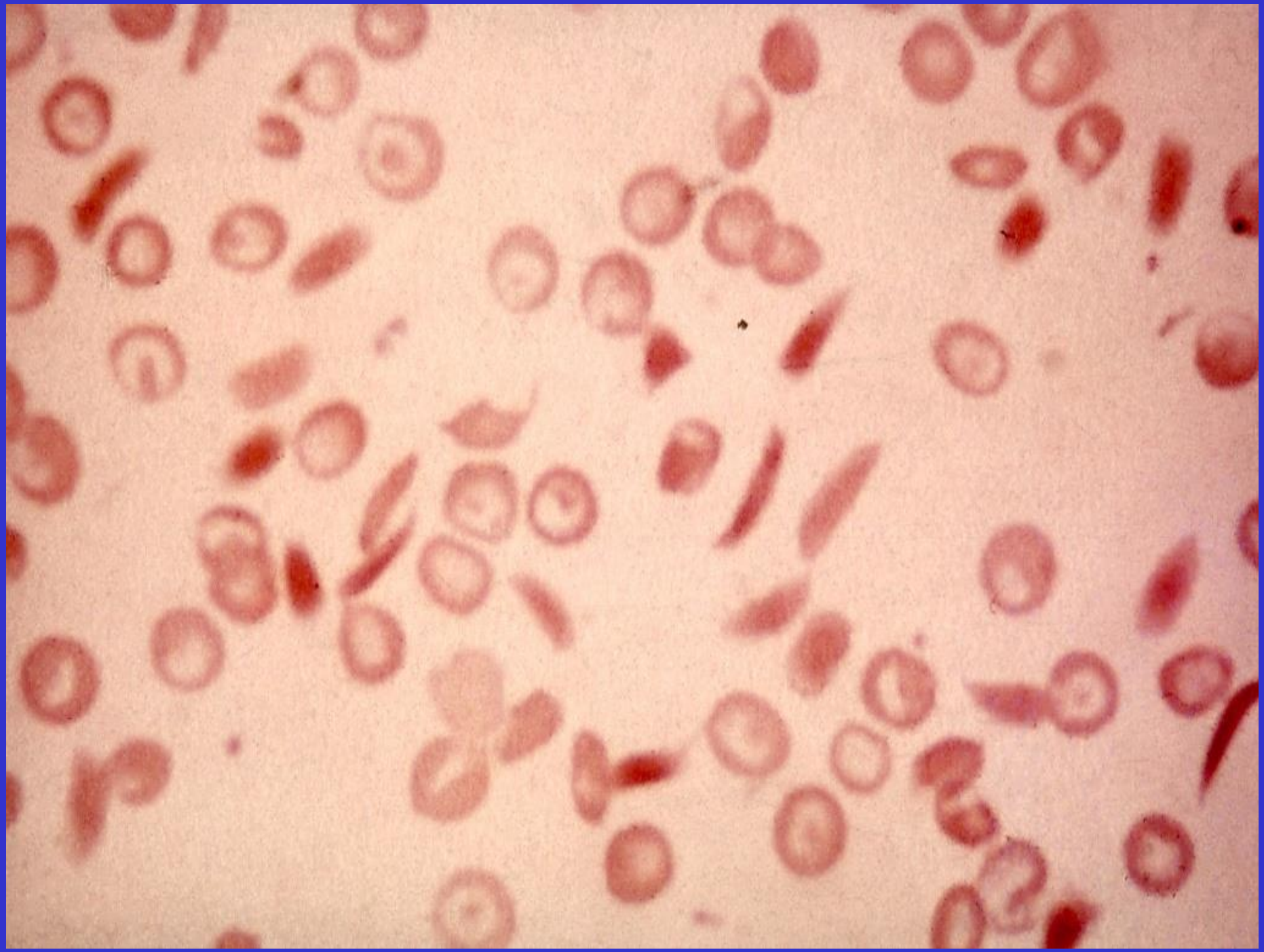
INFECTIONS (especially Malaria)

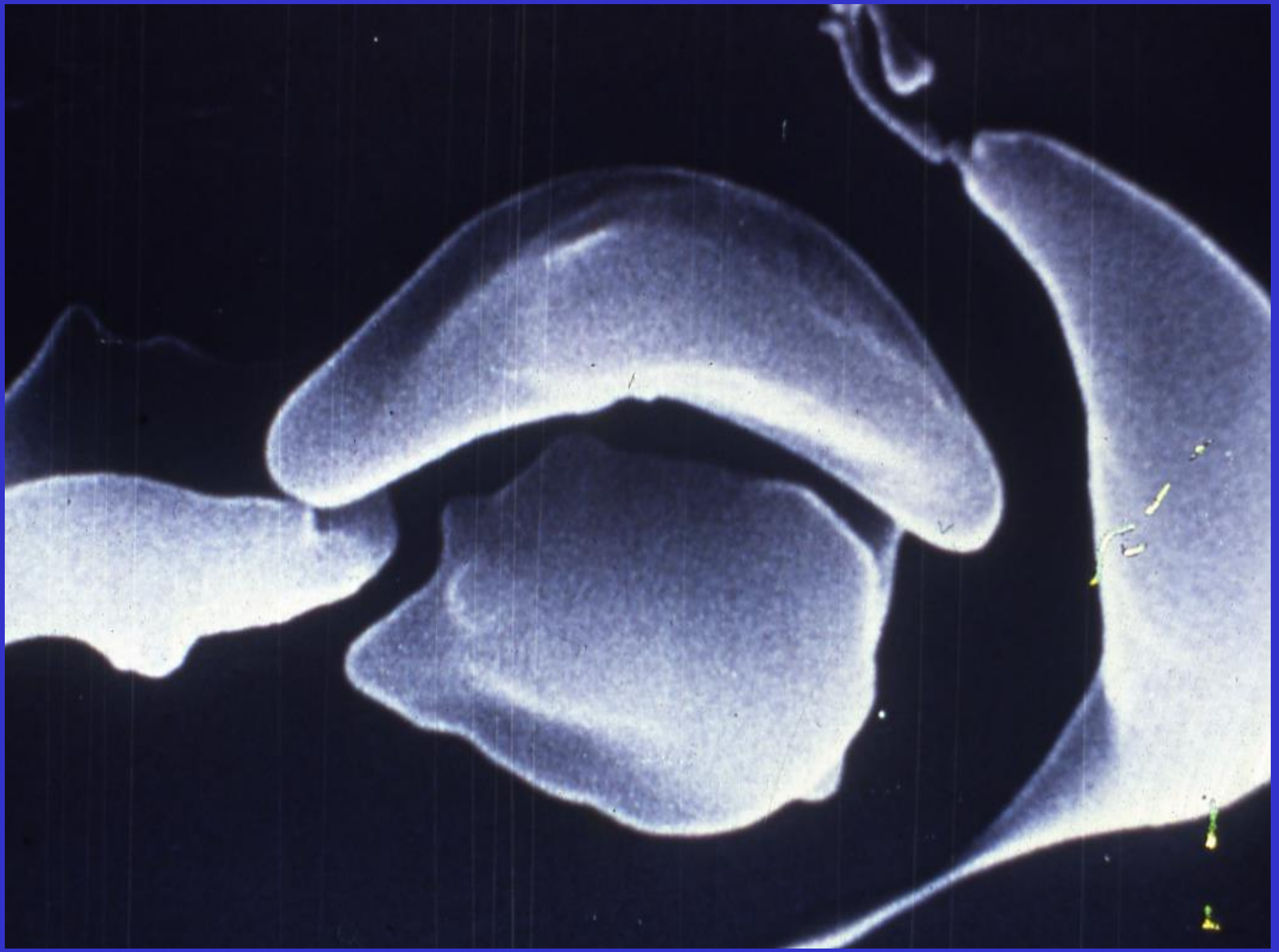
PYREXIA

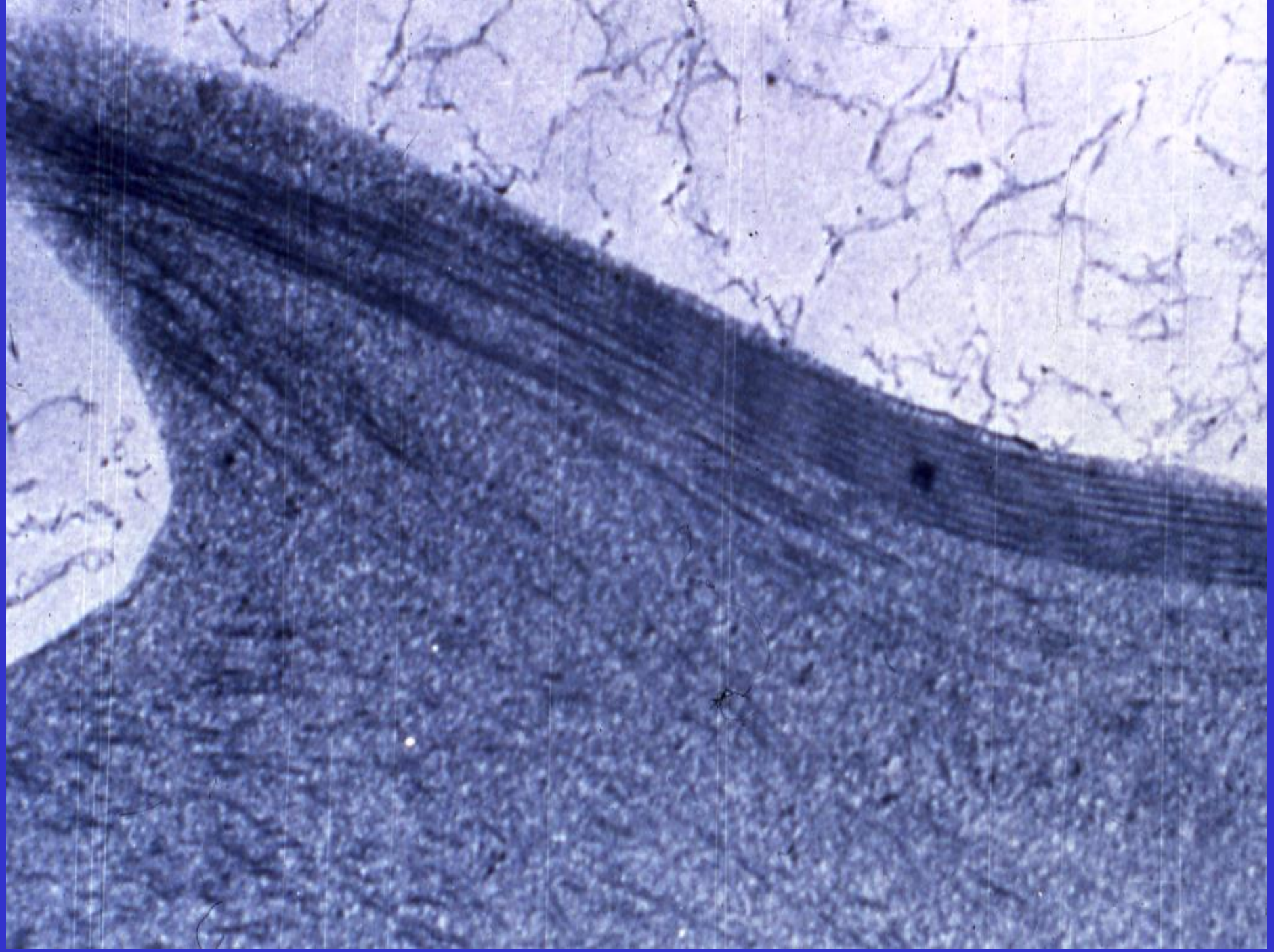
EXPOSURE TO COLD

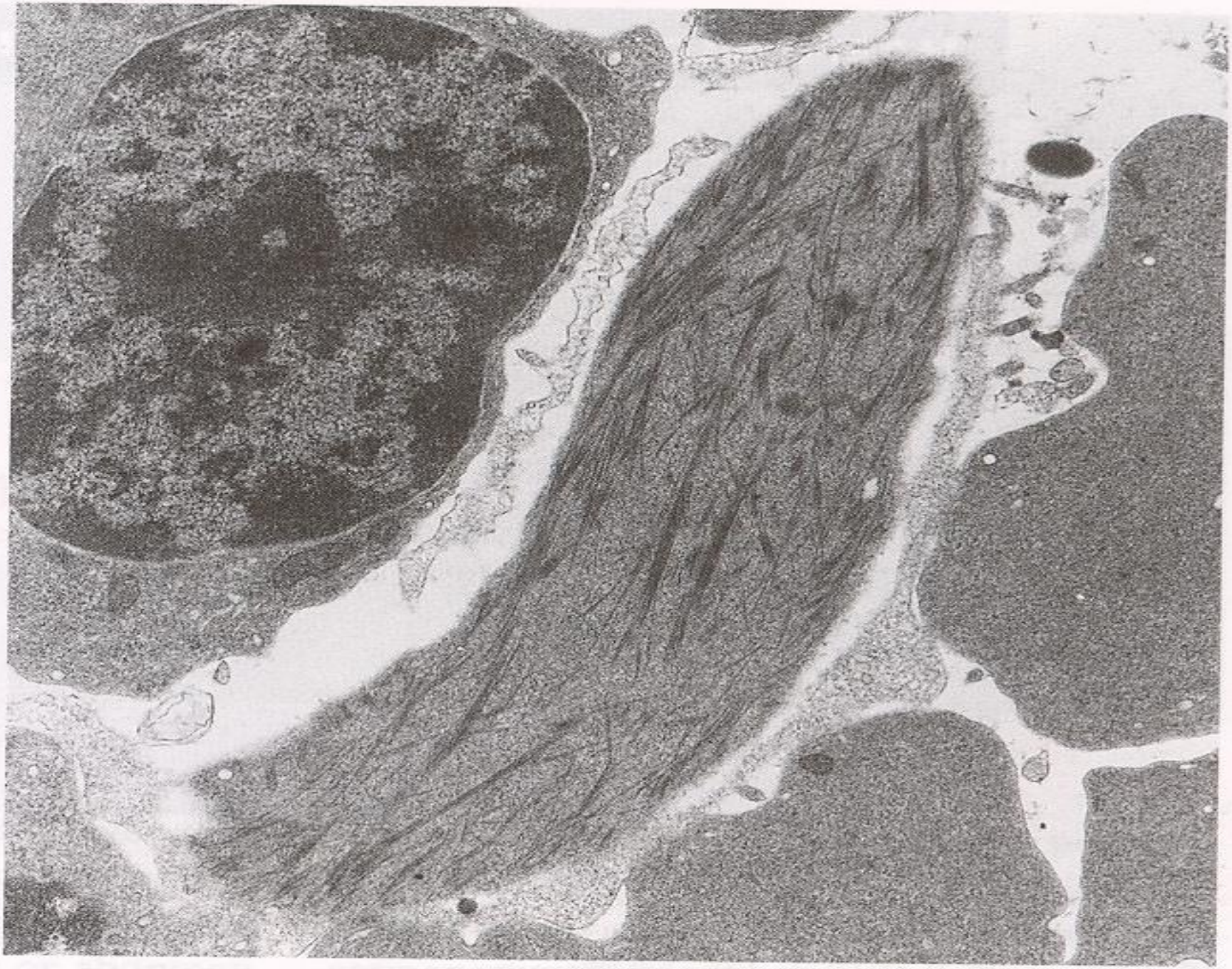
DEHYDRATION

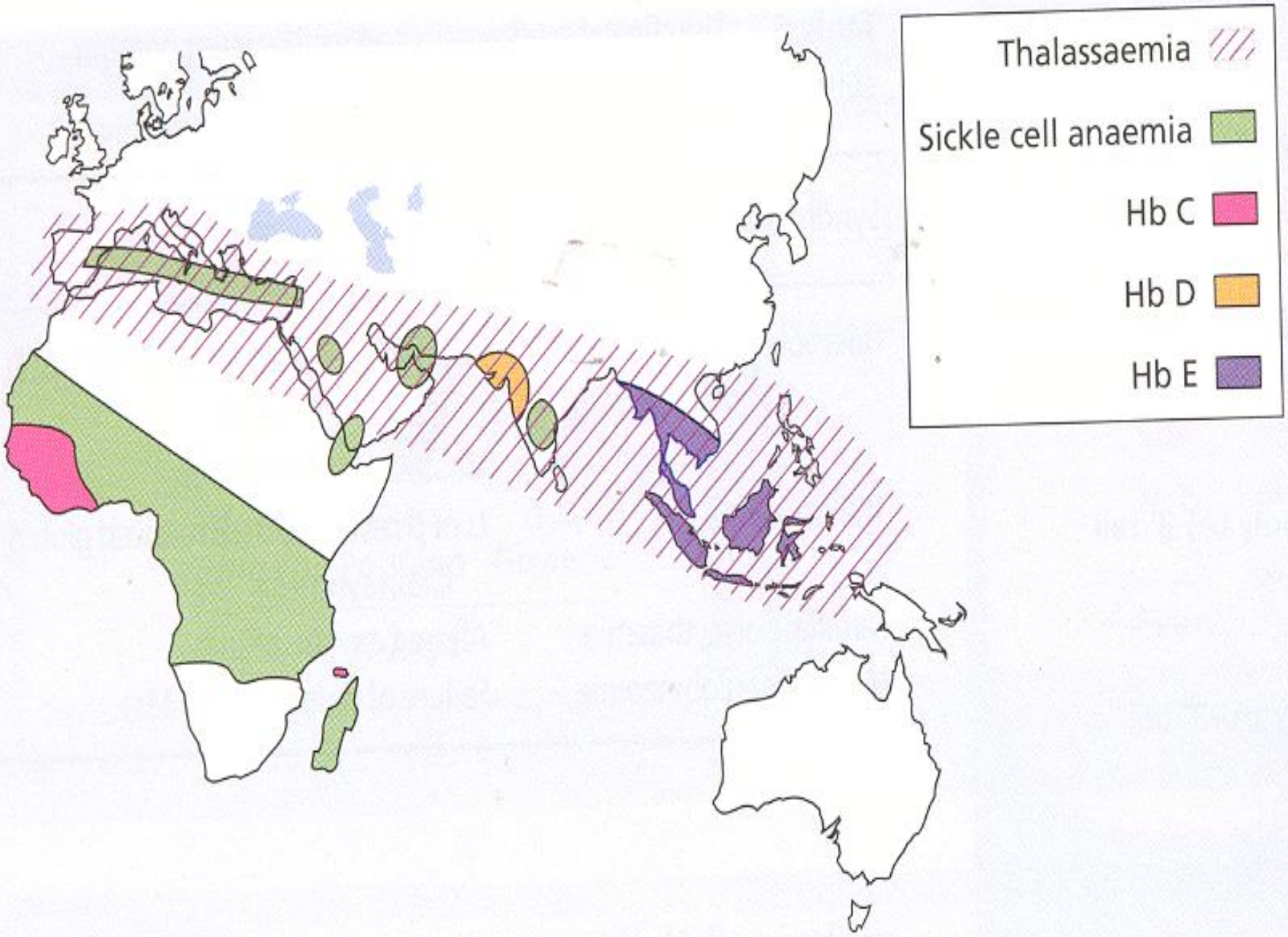
PREGNANCY

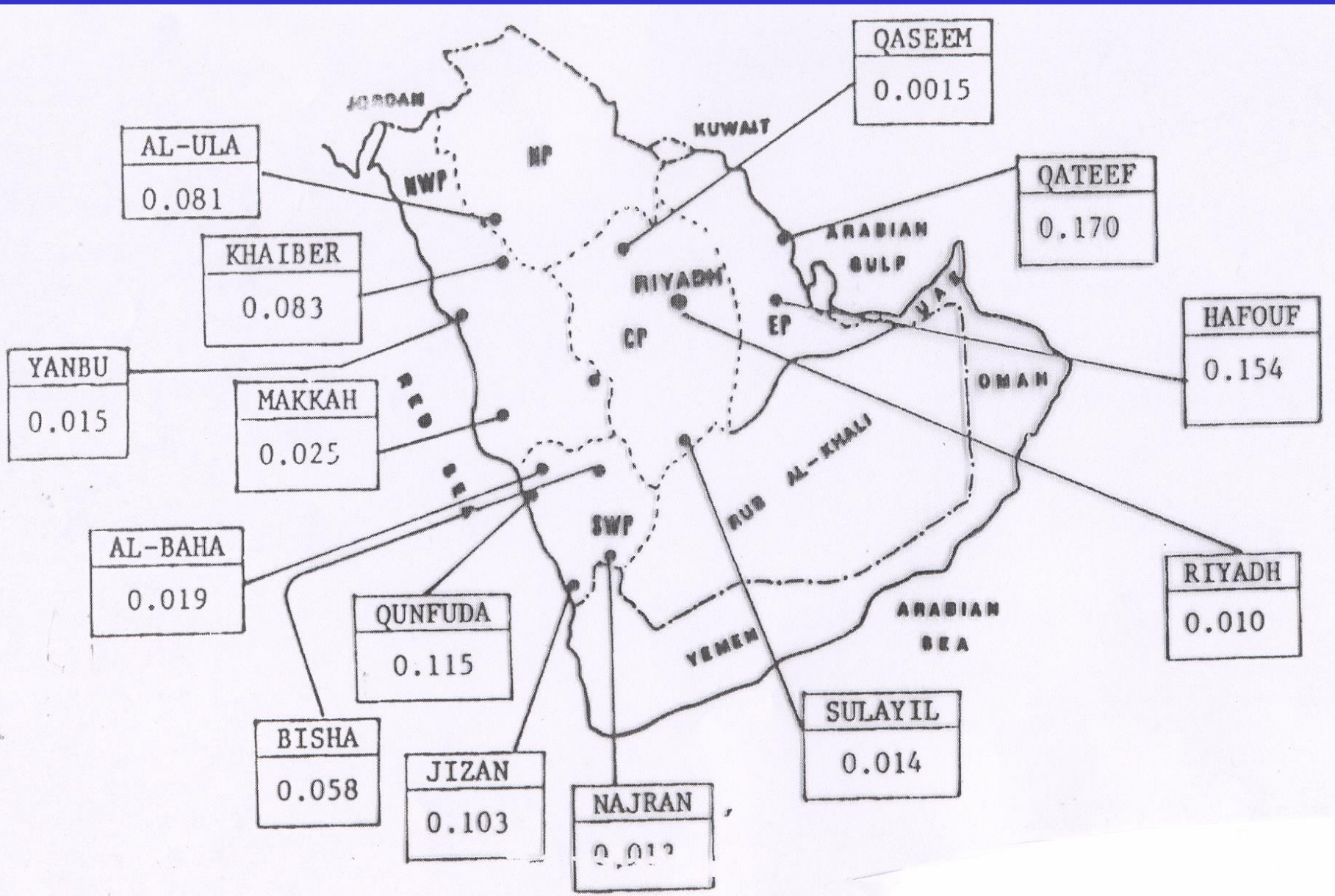









Thalassaemia
Sickle cell anaemia
Hb C
Hb D
Hb E



Frequency of sickle cell (Hb S) gene in different regions of Saudi Arabia

CRISES IN SICKLE CELL DISEASE

HYPERHAEMOLYTIC

AREGENERATIVE OR APLASTIC

SMALL VESSEL OCCLUSION

CLINICAL MANIFESTATIONS OF SICKLE CELL DISEASE

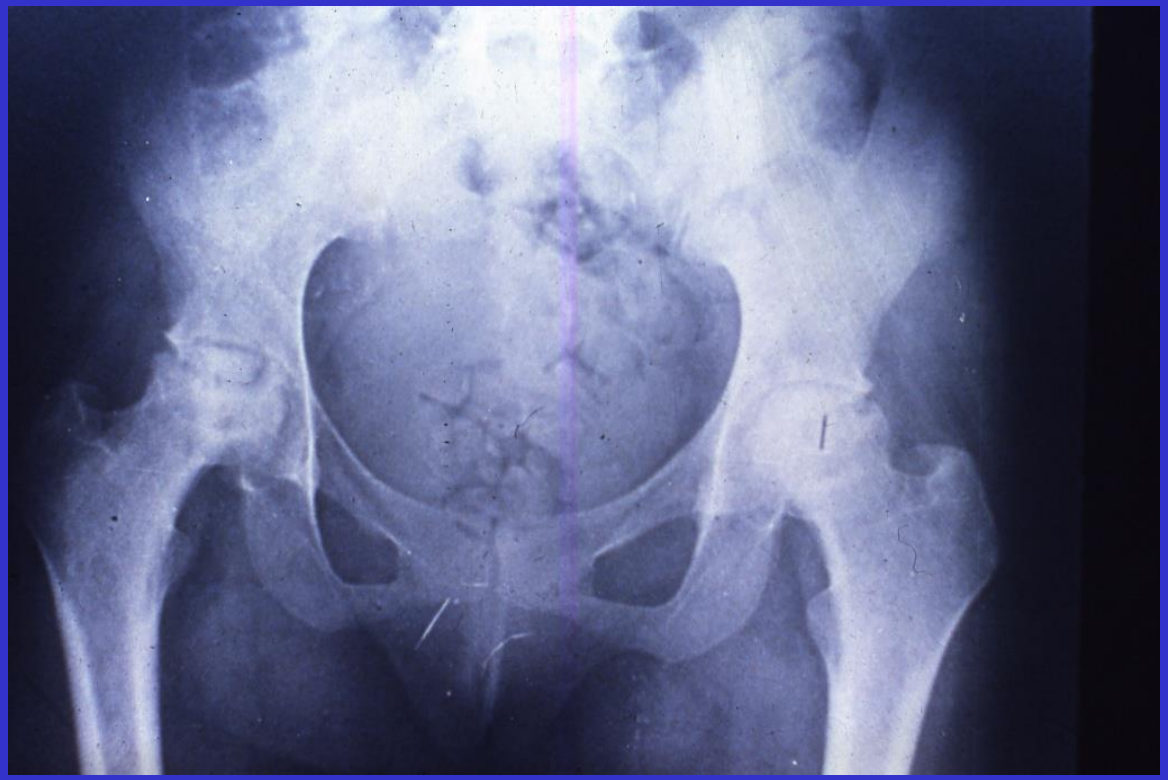
HAEMOLYTIC ANAEMIA
TISSUE INFARCTION

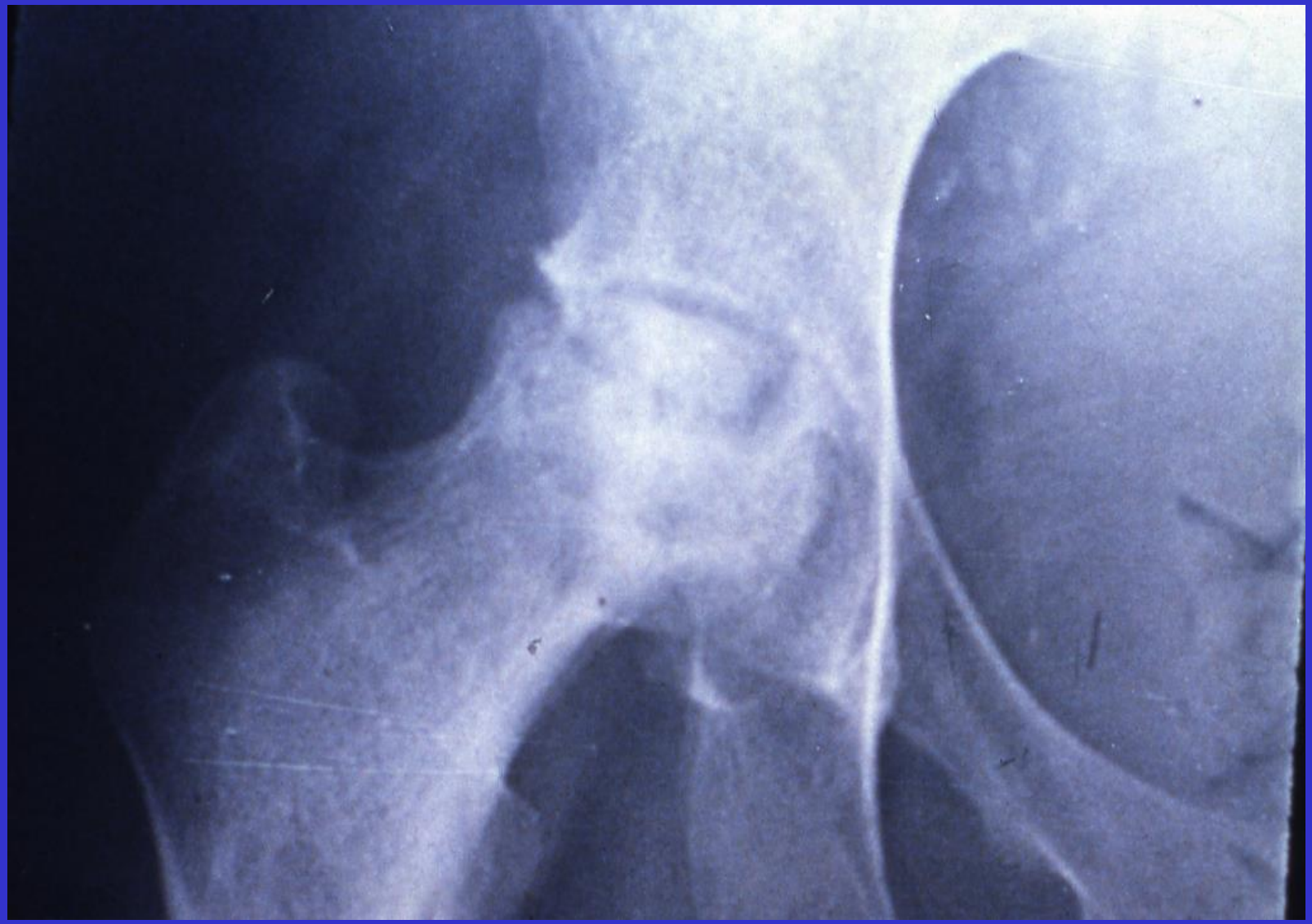
Clinical Manifestations in Sickle Anaemia

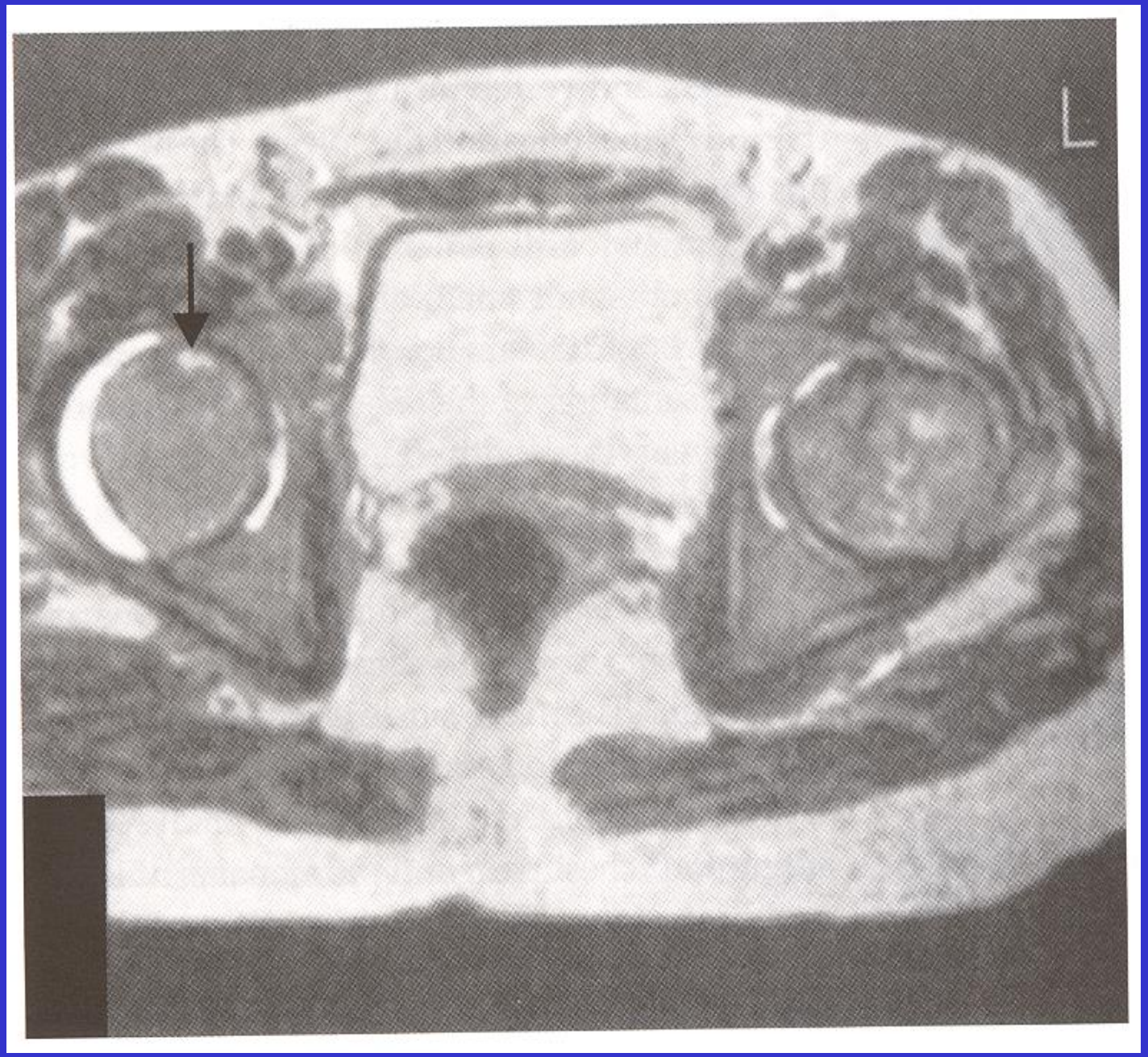
- Pallor (Anaemia)
- Jaundice & Dark Urine
- Apathy & Anorexia
- Hand-Foot Syndrome (Young Children)
- Splenic sequestration (Young Children)Hepatic Sequestration
- Bones, Joints Pain
- Abdominal Pain

Clinical Manifestations in Sickle Anaemia

- Recurrent Infections & Chest Symptoms (Acute Chest Syndrome)
- Hepato-Splenomegaly
  - → (Early Childhood)
  - → (Association with Thalassaemias)
- CNS Presentations
- Leg Ulceration
- Skeletal Deformity



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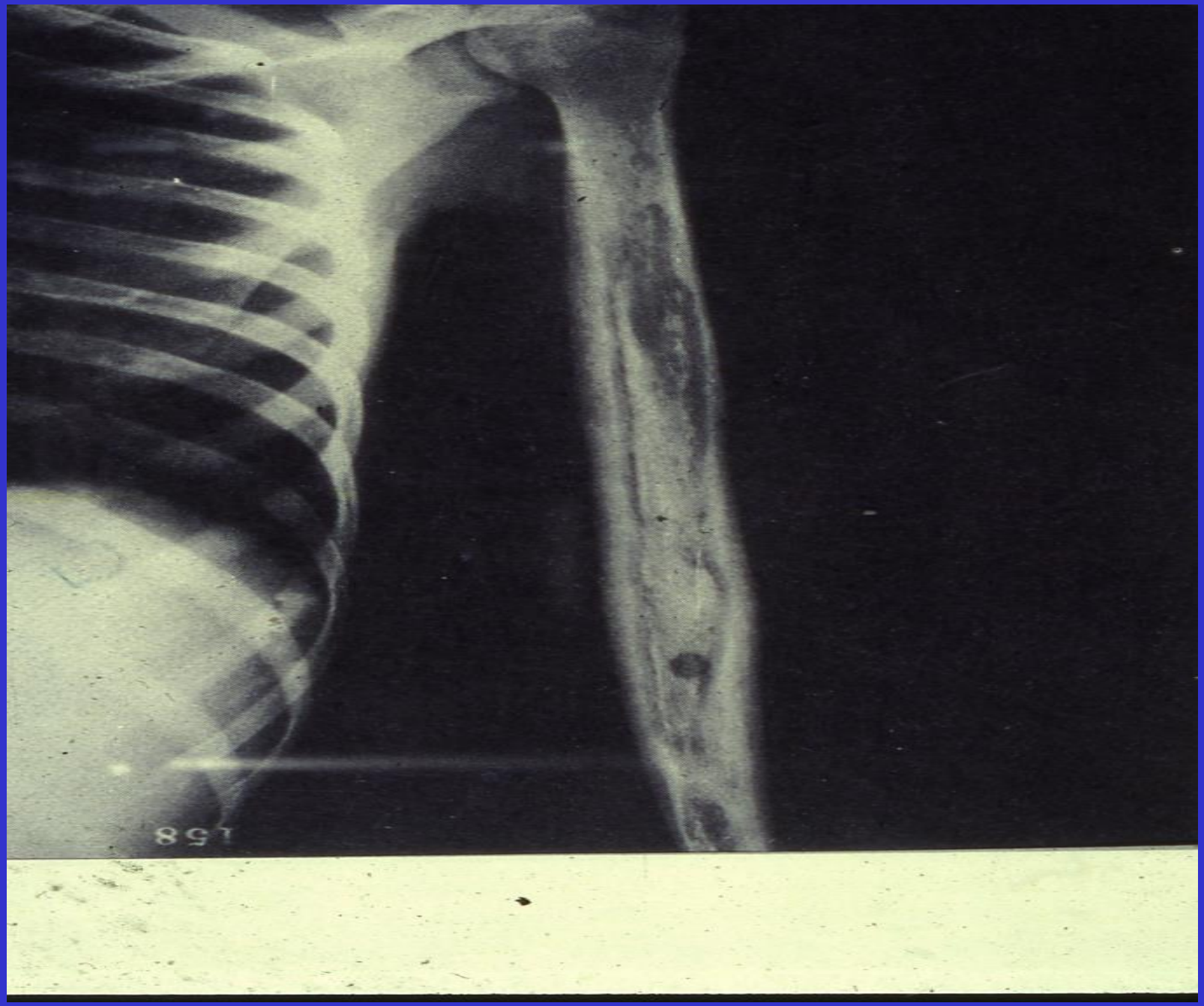
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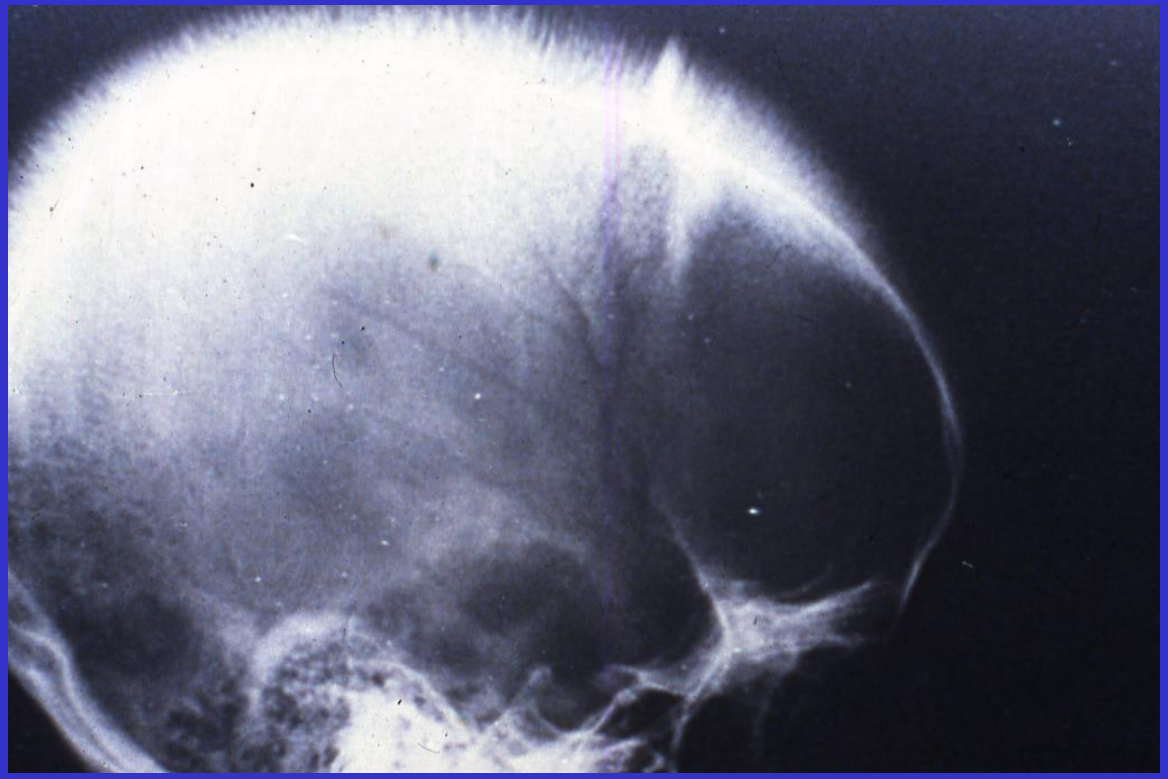
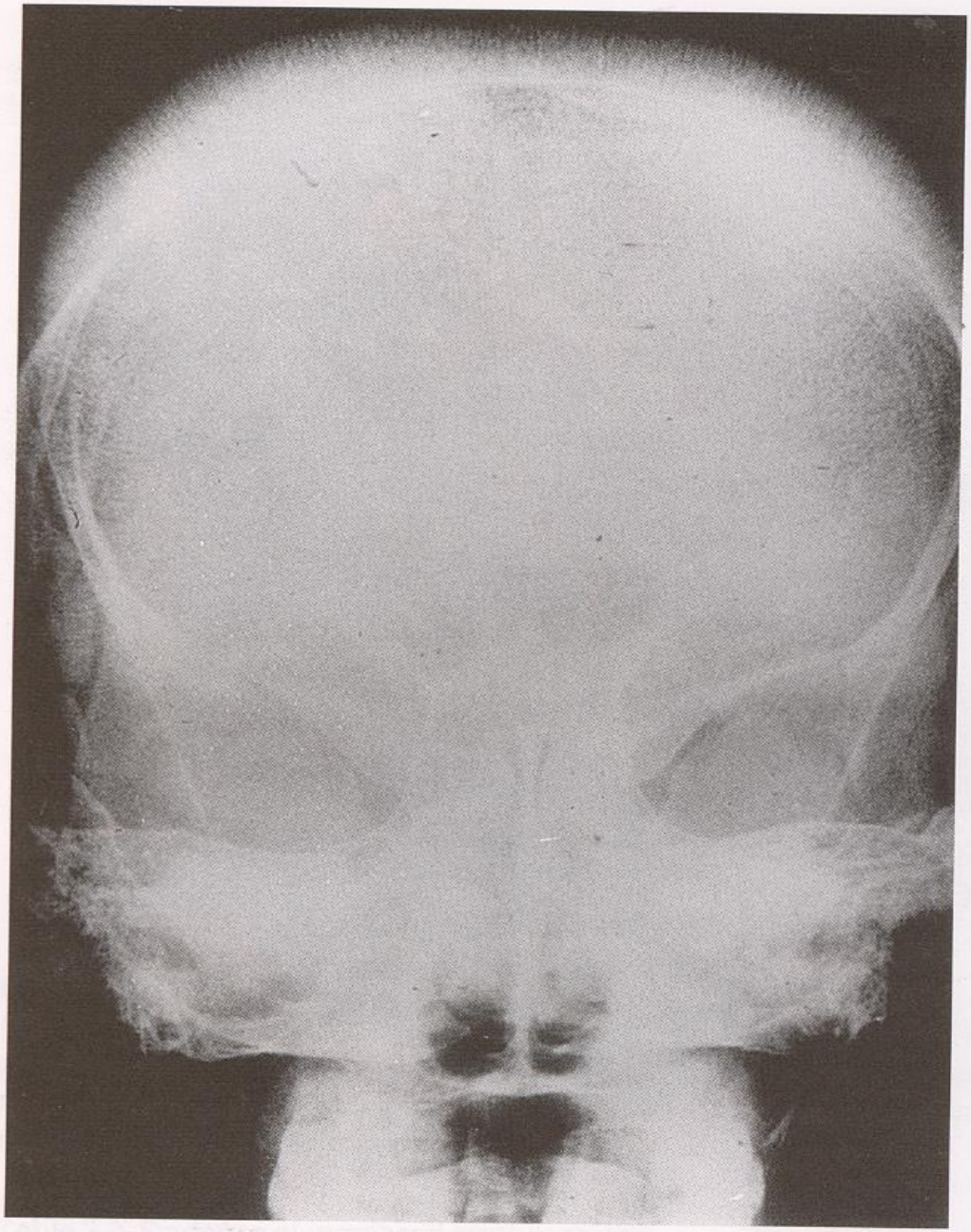
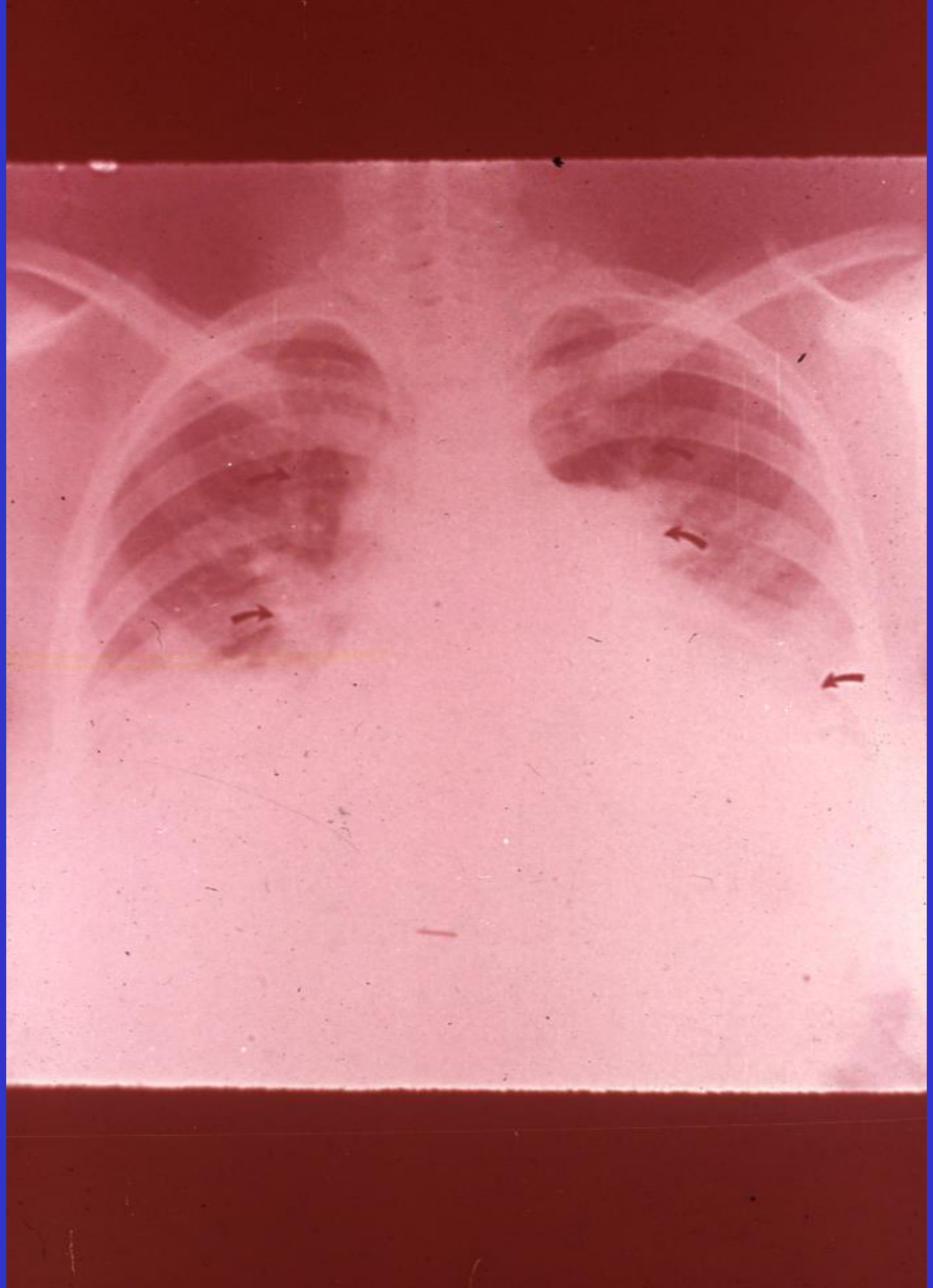
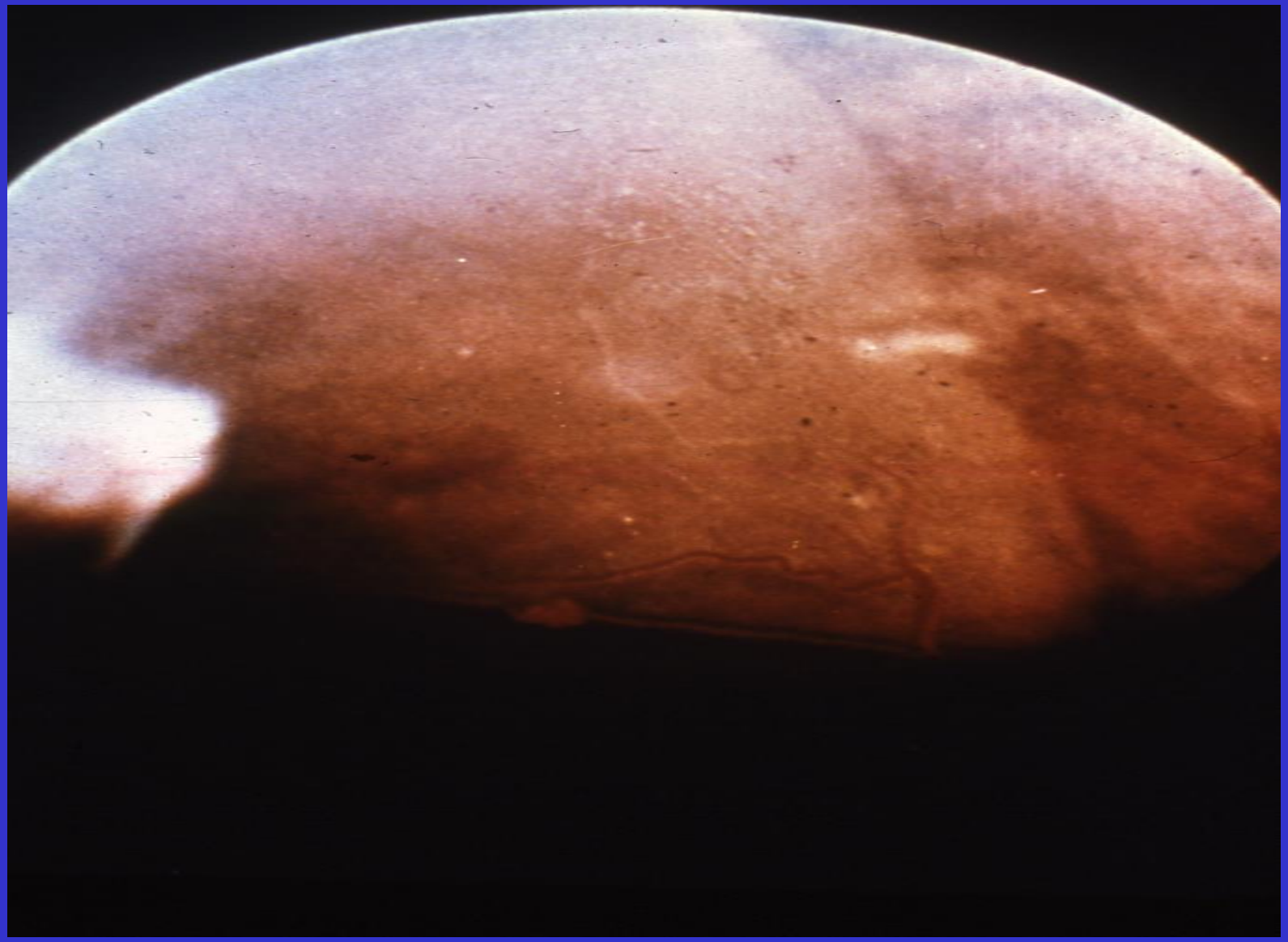
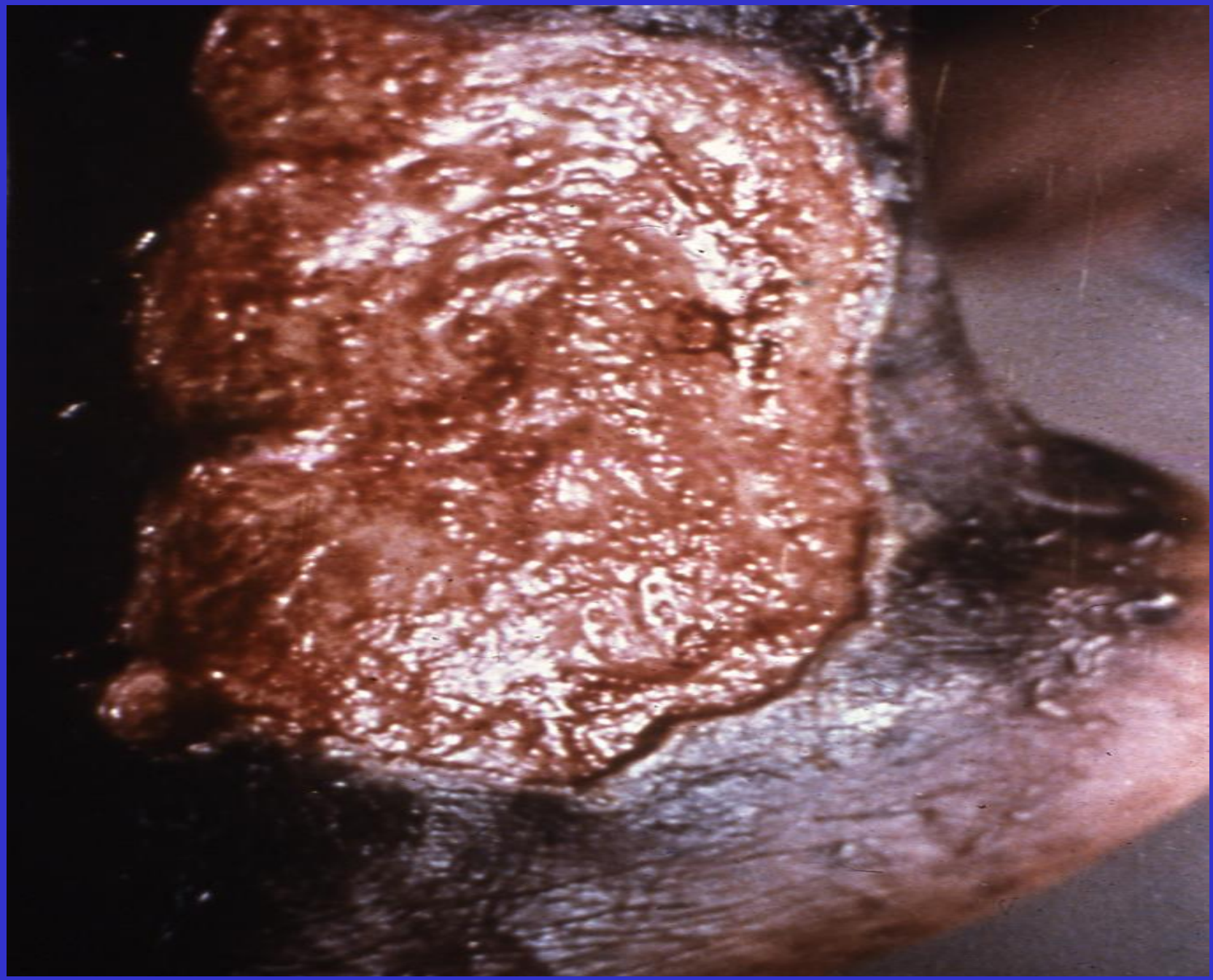
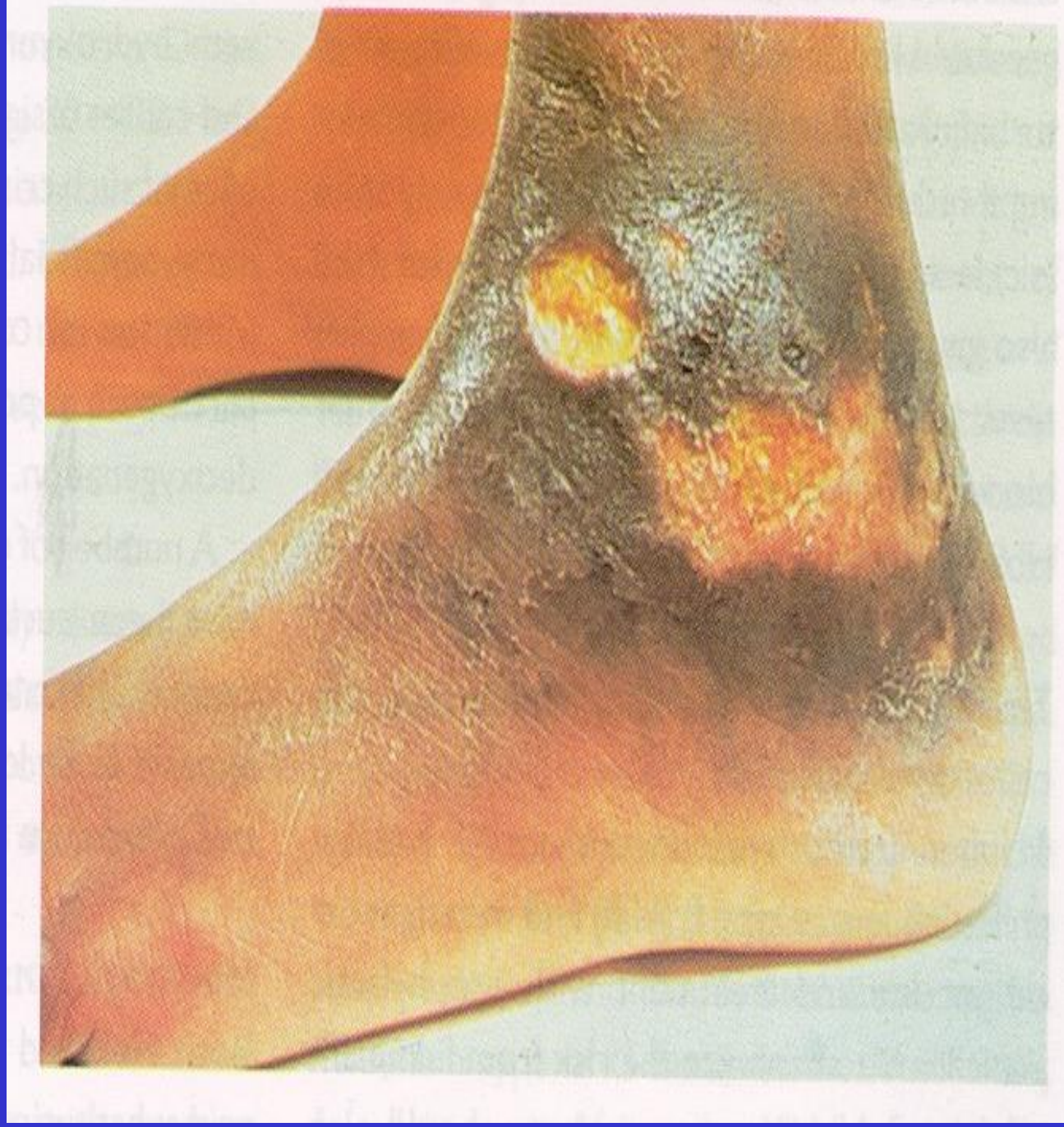






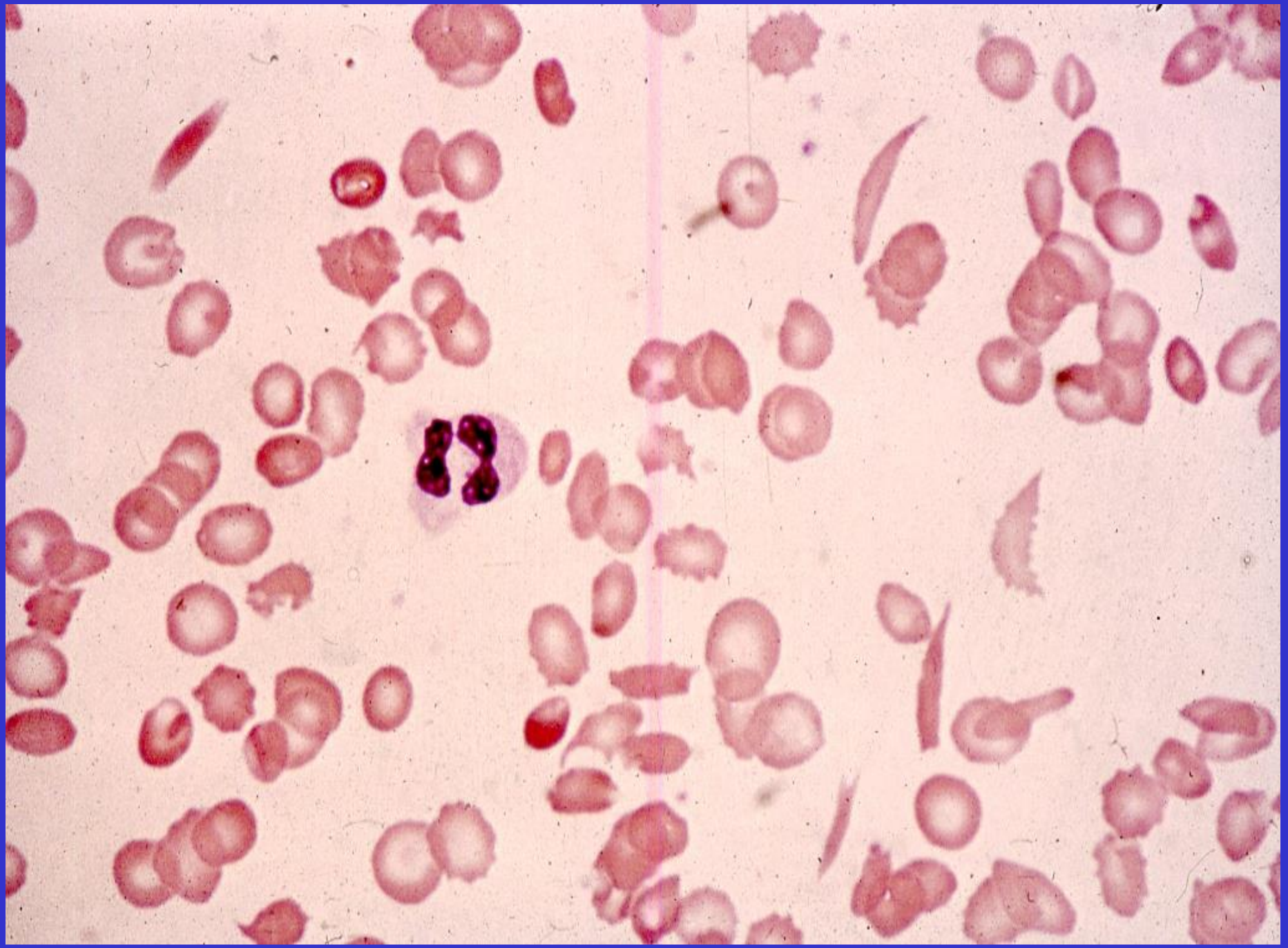


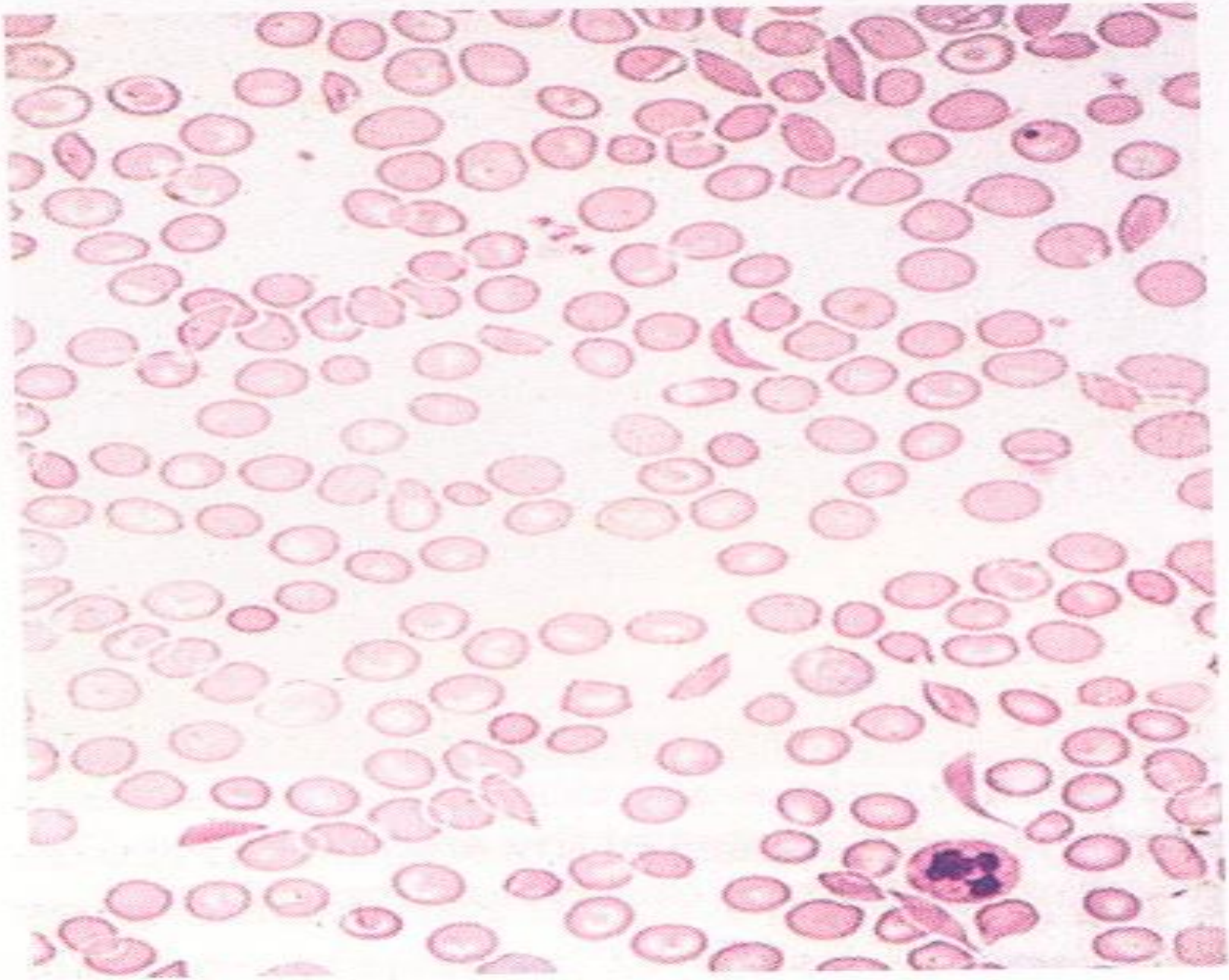


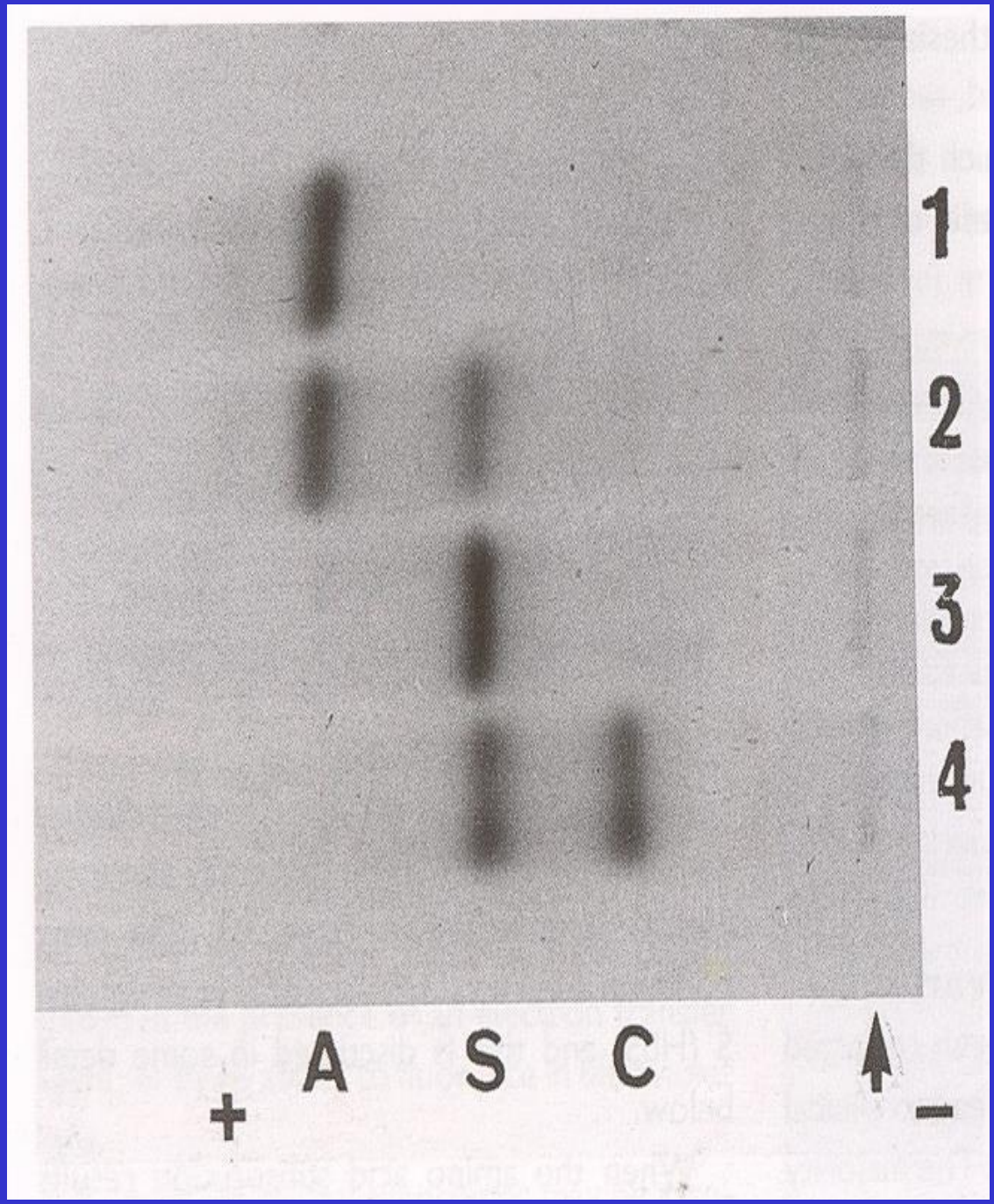


Laboratory Diagnosis

- CBC
- Blood Film
- Sickle Solubility Test
- Hb Electrophoresis
- Genetic Study



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Indications for Blood Transfusion in Sickle Cell Anaemia

* Splenic sequestration
* Hepatic sequestration
* Aplastic crisis
* Overwhelming infections
* Elective or emergency surgical operation
* Severe painful crisis associated with severe haemolysis
* Pregnancy

Indications for exchange transfusion

* Strokes
* Pulmonary infarcts with infection
* Pregnancy (Severe persistent painful crisis)
* Priapism
* Preparation for major surgery

Practical Haemaglobinopathy

HAEMOGLOBIN VARIANTS: GENE DISTRIBUTION

EFFECTS OF HAEMOGLOBIN VARIANTS

Variant	Clinical and haematological abnormalities
HbS	Recurrent painful crises (in adults) and chronic haemolytic anaemia; both related to sickling of red cells on deoxygenation*
HbC	Chronic haemolytic anaemia due to reduced red cell deformability on deoxygenation, * deoxygenated HbC is less soluble than deoxygenated HbA.
Hb Köln, Hb Hammersmith	Spontaneous or drug-induced haemolytic anaemia due to instability of the Hb and consequent intracellular precipitation.
HbM Boston, HbM Saskatoon	Cyanosis due to congenital methaemoglobinaemia as a consequence of a substitution near or in the haem pocket.
Hb Chesapeake	Hereditary polycythaemia due to increased O_2 affinity.
Hb Constant Spring, Hb Lepore, HbE	Thalassaemia-like syndrome due to decreased rate of synthesis of normal chains.
Hb Indianapolis	Thalassaemia-like syndrome due to marked instability of Hb

* Only in homozygotes

Abnormal Haemoglobin Variants

Hb C:-

* Is due to replacement of glutamic acid in position 6 of the beta chain by lysine ($\alpha_2\beta_2$ 6-GLU→ LYS).
* About 7-22% of people of West Africa ar hetrozygotes especially Nigeria and North Ghana
* Homozygotes are rare and have mild to moderate hemolytic anaemia with many thick target RBCs in the blood film and mild to moderate splenomegaly.
* The chronic hemolytic anaemia is due to reduced red cell deformability on deoxygenation.

  Deoxygenated HbC is less soluble than deoxygenated HbA.
* Double heterozygotes with sickle Hb S/C give moderate to sever anaemia with symptoms of sickle cell disease.

Hb D Punjab

($\alpha_2\beta_2$-121 GLU → GLN)

Prevalent in Indian and Pakistani in every 100 persons about 1 trait (1% of the population).

Trait are usually health.

Homozygous D/D have mild to moderate anaemia.

Combined double heterozygotes Hb S/D can give rise to moderate to a severe anaemia and symptoms of sickle cell disease.

Hb E:

* ($\alpha_2\beta_2$ 26 GLU → LYS) is one of the most common beta-chain variants.
* It is very prevalent in South East Asia (50%) of the population are heterozygotes.
* Patients who are homozygous generally have mild haemolytic anaemia, microcytic hypochromic red cells and mild enlargement of the spleen.
* Carriers are symptomless unless they have combined other mutations such as the one for alpha thalassemia, or beta-thalassemia trait.

Hb O Arab

($\alpha_2\beta_2$-121 GLU → LYS)

Heterozygotes are not symptomatic.

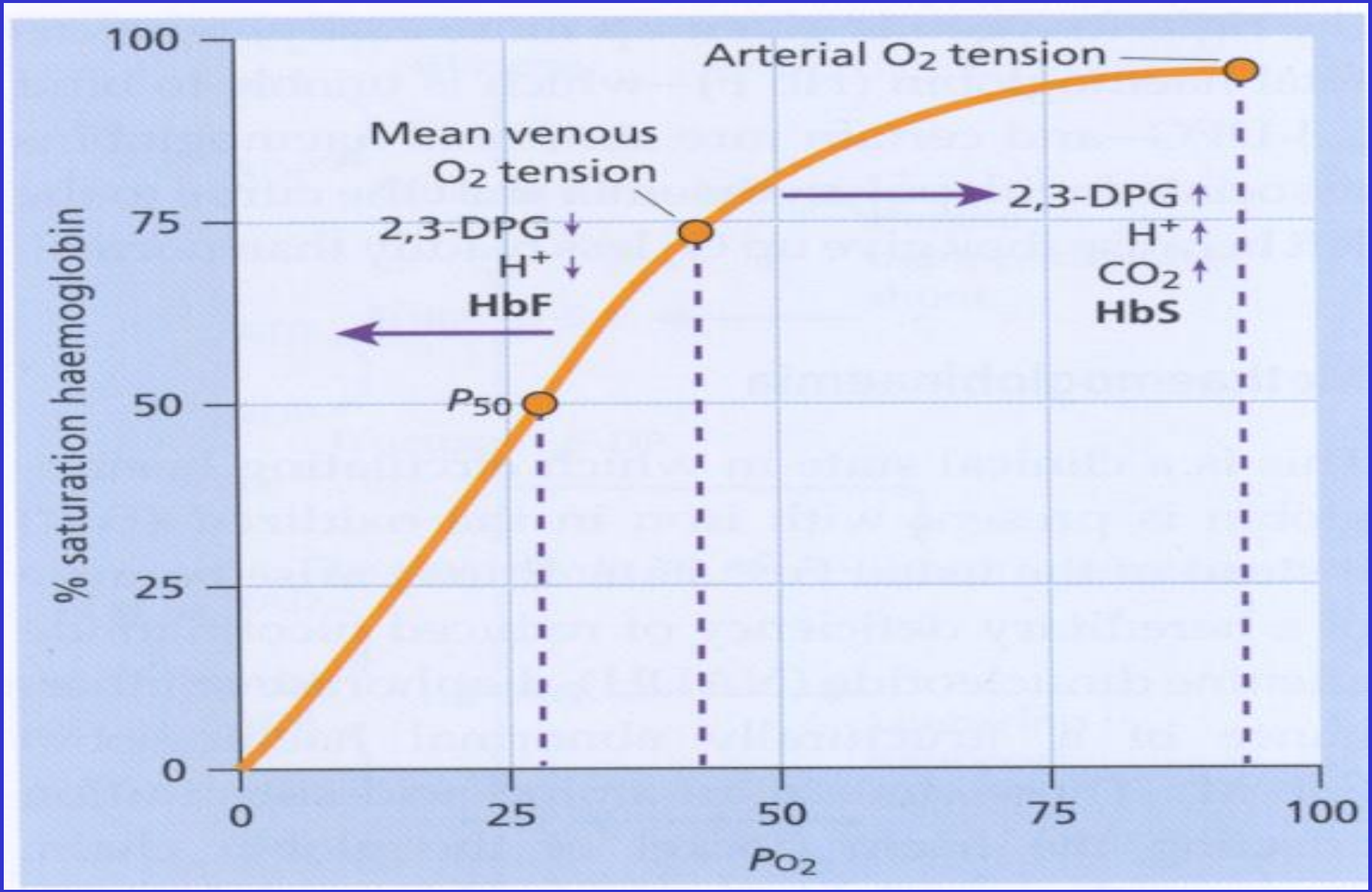
Double heterozygous with sickle S/O are clinically severe.

Hb O- Arab enhance the polymerization of HbS.

High Oxygen affinity haemoglobins

Hb Chesapeake:

* (α_2-92 ARG → LEU β_2).
* Carriers are without clinical symptoms.
* Homozygous of erythrocytosis (polychemia) due to increased O_2 affinity.
* The patients have no splenomegaly. (except for patient's with concomitant β-thalassemia).
* They have normal WBC, and normal platelets.
* High Hb, High RBCs count and high haematocrit. (HCT).



The haemoglobin oxygen (O_2) dissociation cruve. 2,3-DPG, 2,3-diphosphoglycerate.

Unstable Haemoglobins

Hb koln ($\alpha_2\beta_2$-98 VAL → MET)

Hb Hammersmith ($\alpha_2\beta_2$ 42 PHE → SER)

Hb Hasharon (α_2-47 ASP → HIS β_2).

- **These abnormal haemoglobin cause haemolysis in the newborn (congenital non-spherocytic haemolytic anaemia).**
- **Heinz body hemolytic anaemia with sensitivity to oxidant drugs, such as sulfonamides.**
- **Reticulocytosis out of proportion to the level of Hb.**
- **Increased formation of methemoglobin.**
- **Spontaneous or drug induced haemolytic anaemia due to instability of the haemoglobin and consequent intracellular precipitation.**
- **Thalassaemia – like peripheral blood picture.**

Clinically: The patient have anemia, jaundice, splenomegaly / hepatomegaly and gall stones.

Low oxygen affinity haemoglobins

More than 50 variants with reduced oxygen affinity have been identified.

Hb kansas ($\alpha_2\beta_2$102 ASN → THR)

Hb Aukland ($\alpha_2\beta_2$ 25 GLY → ASP)

Rare as homozygotes.

Patients have anaemia and congenital cynosis due to reduced oxygen affinity.

Congenital methaemoglobinaemia

Hb M Boston (α_2 58 HIS → TYR - β_2)

Hb M Saskatoon (α_2.β_2-63 HIS → TYR)

Hb M Hyde park ($\alpha_2\beta_2$ 92 HIS → TYR)

Hb M IWATE ($\alpha_2$87 HIS → TYR-β_2)

Cynosis in homozygotes due to congenital methaemoglobinaemia as a consequences of substitution of amonoacids near or in haem pocket.

Hb iIndianapolis

(α_2-$\beta_2$112 CYS – ARG)

Is a rare and slightly unstable beta-globin variant.

Carriers are clinically normal with only mild reticulocytosis.

Homozygons have haemolytic anaemia and renal failure in severe cases.

Thalassaemia-like syndrome due to marked instability of the Hb.

KKUH

Heamatology Unit

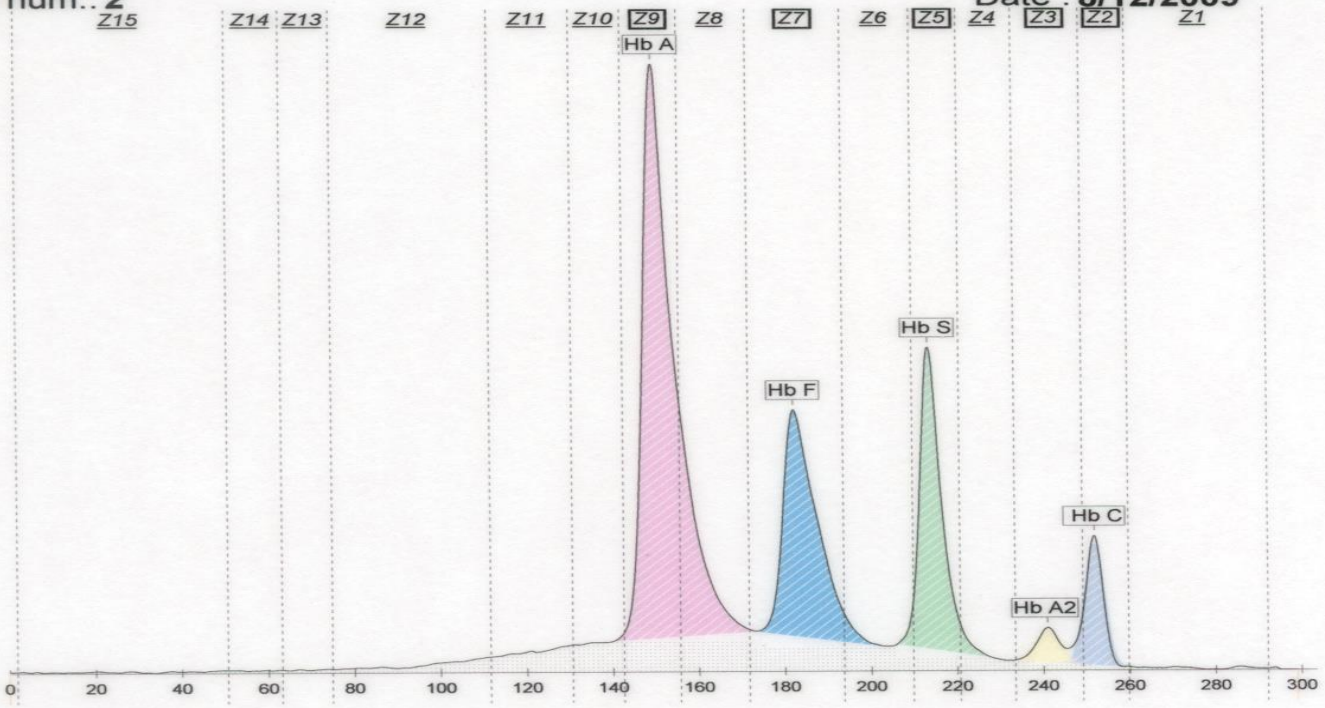
Hb Electrophoresis

Hospital No.: **QC Hb AFSC CONTROL-**

ID : **Hb AFSC CONTROL-2**

Sample num.: **2**

Date : **8/12/2009**



Hb Electrophoresis

Fractions	%	Ref. %
Hb A	**51.3**	46.7 - 56.9
Hb F	**21.4**	17.4 - 22.4
Hb S	**18.3**	17.3 - 22.3
Hb A2	**2.3**	2.1 - 3.3
Hb C	**6.7**	4.6 - 7.0

KKUH

Heamatology Unit

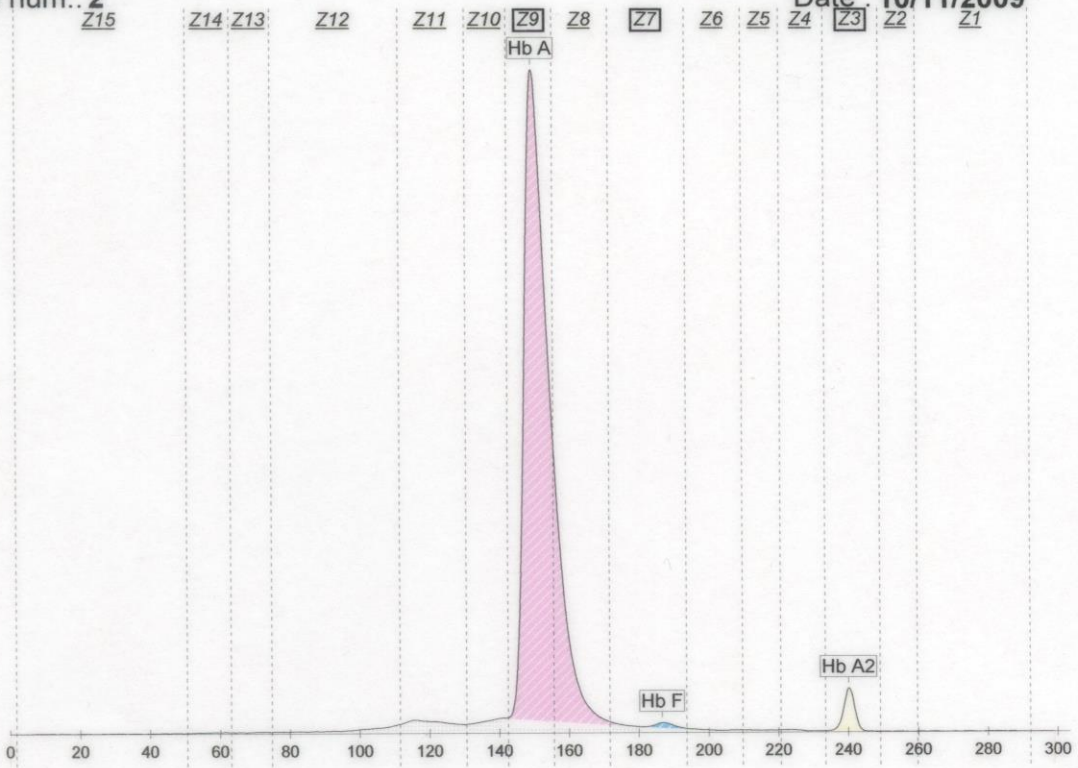
Hb Electrophoresis

Hospital No.: **933376**

ID : **061773**

Sample num.: **2**

Date : **10/11/2009**



Hb Electrophoresis

Fractions	%	Ref. %	
Hb A	**96.7**	96.8 - 97.8	
Hb F	**0.5**	=< 2.0	<
Hb A2	**2.8**	1.5 - 3.5	

KKUH

Heamatology Unit

Hb Electrophoresis

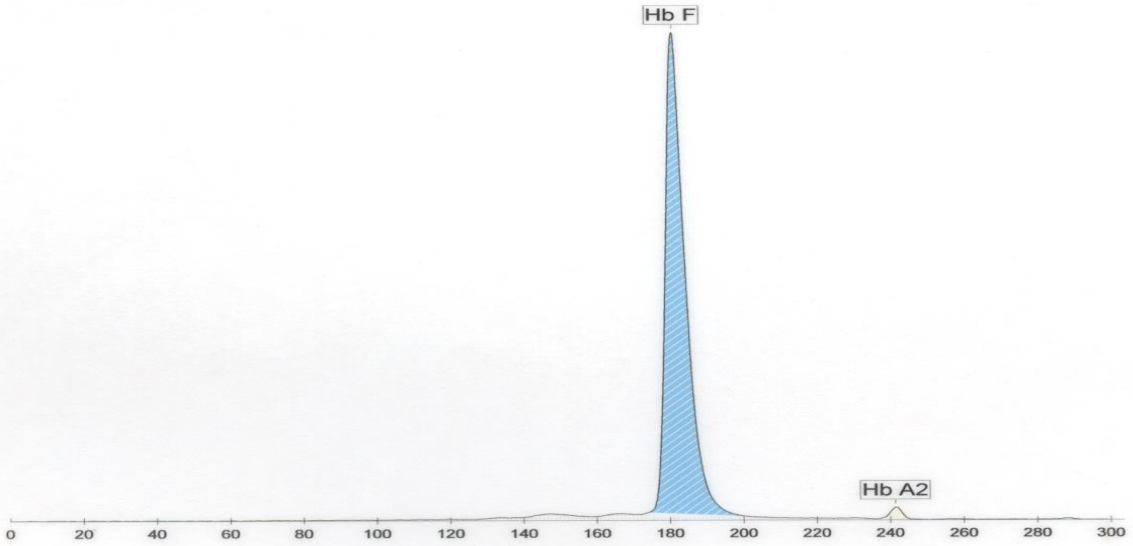
INSTRUMENT ID : KKUH : 24509

Hospital No.: **921107**

Sample No **54**

ID : **063761**

Date : **09/05/2010**



Fractions	%	Ref. %
Hb F	98.5	
Hb A2	1.5	

Comment :

28/3/2010

CBC Hb 98

MCV 73

NRBC 34

KKUH

Heamatology Unit

Hb Electrophoresis

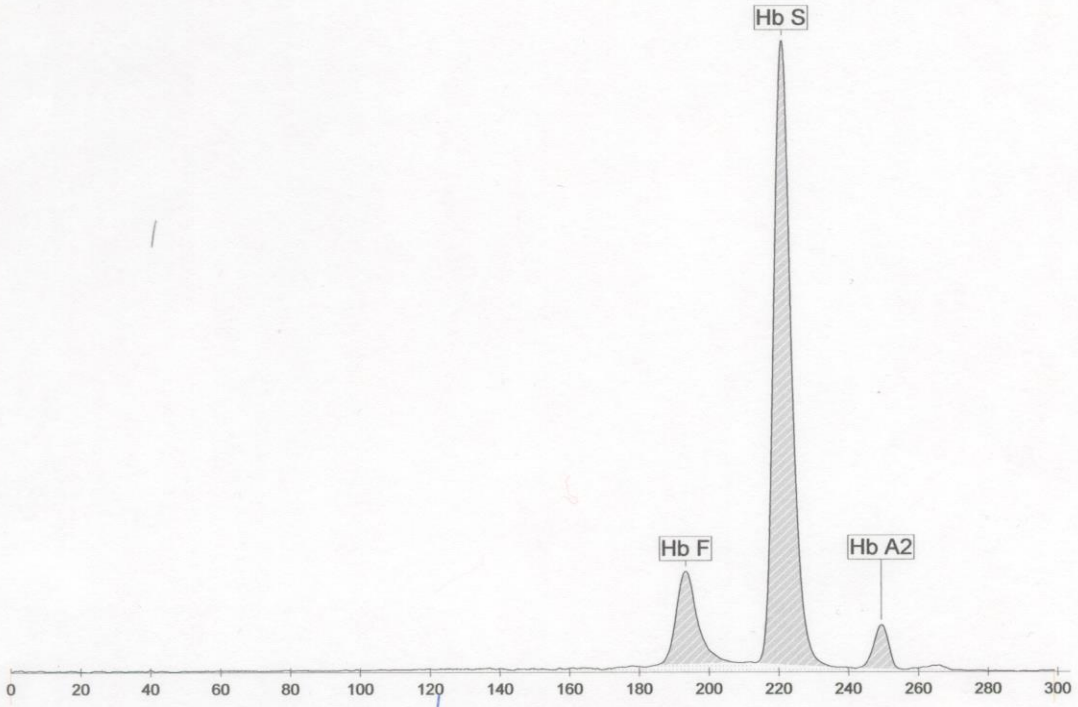
INSTRUMENT ID : KKUH : 24509

Hospital No.: **233095**

Sample No **20**

ID : **063478**

Date : **17/04/2010**



Fractions	%	Ref. %
Hb F	**14.7**	
Hb S	**80.5**	
Hb A2	**4.8**	

KKUH

Heamatology Unit

Hb Electrophoresis

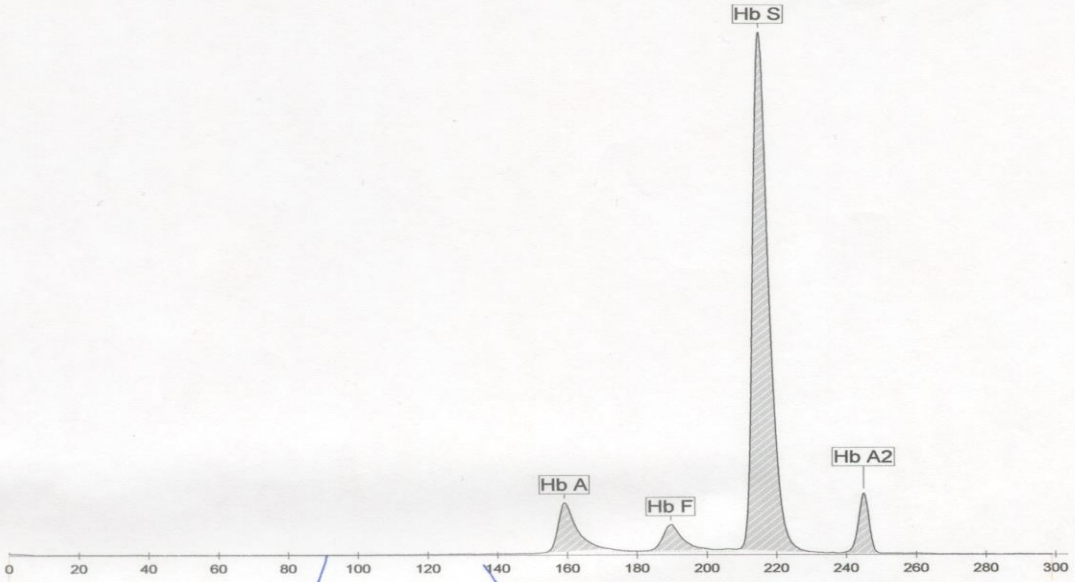
INSTRUMENT ID : KKUH : 24509

Hospital No.: **913628**

Sample No **34**

ID : **063511**

Date : **19/04/2010**



Fractions	%	Ref. %
Hb A	**8.7**	
Hb F	**4.9**	
Hb S	**80.1**	
Hb A2	**6.3**	

Comment :

Homozygous sickle cell thal

KKUH

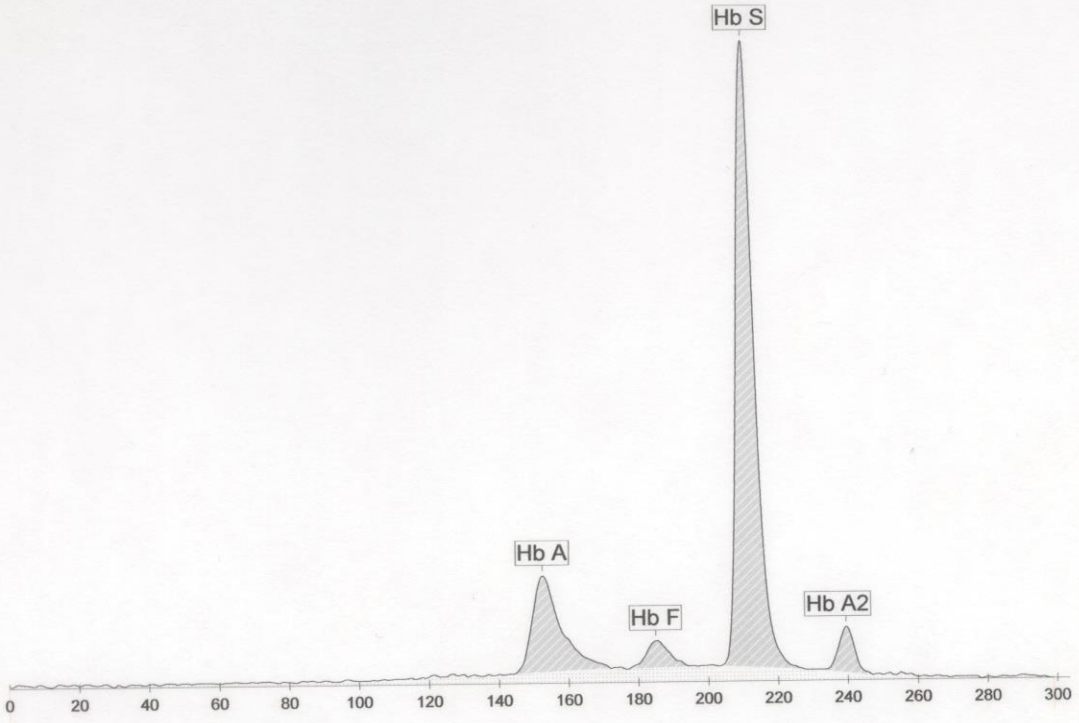
Heamatology Unit

Hb Electrophoresis

INSTRUMENT ID : KKUH : 24509

Hospital No.: **010755** ID : **064209**

Sample No **19** Date : **28/06/2010**



Fractions	%	Ref. %
Hb A	**18.0**	
Hb F	**4.0**	
Hb S	**73.3**	
Hb A2	**4.7**	

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Heamatology Unit

Hb Electrophoresis

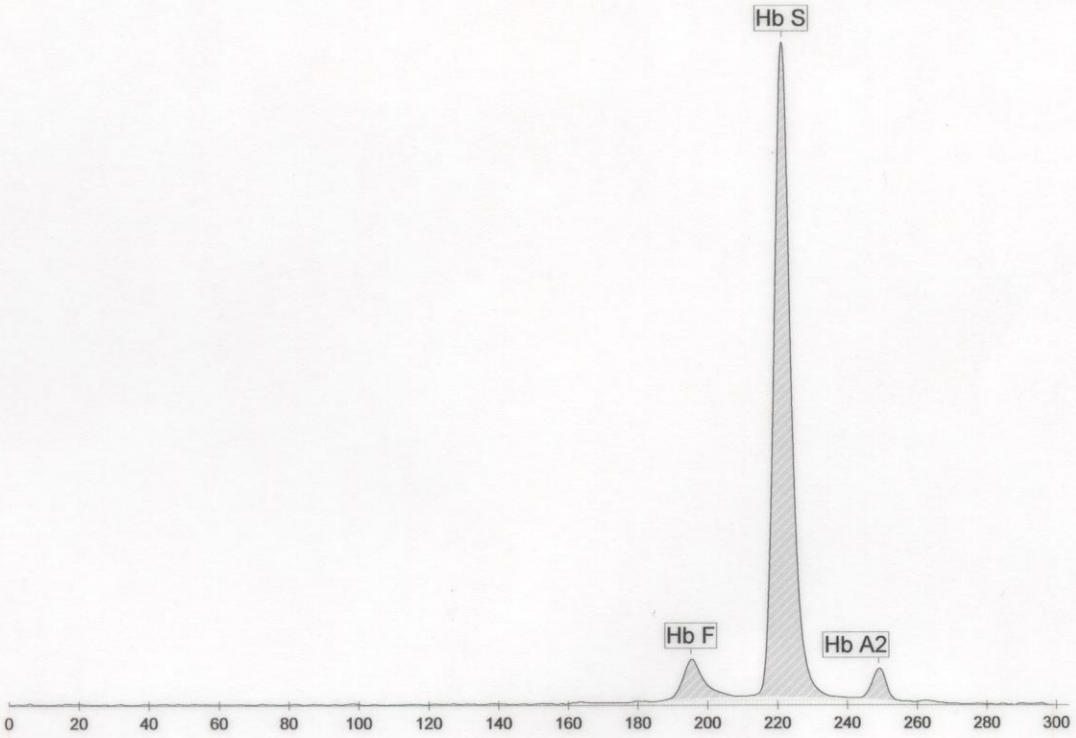
INSTRUMENT ID : KKUH : 24509

Hospital No.: **594729**

Sample No **37**

ID : **064199**

Date : **27/06/2010**



Fractions	%	Ref. %
Hb F	**6.5**	
Hb S	**89.9**	
Hb A2	**3.6**	

KKUH

Heamatology Unit

Hb Electrophoresis

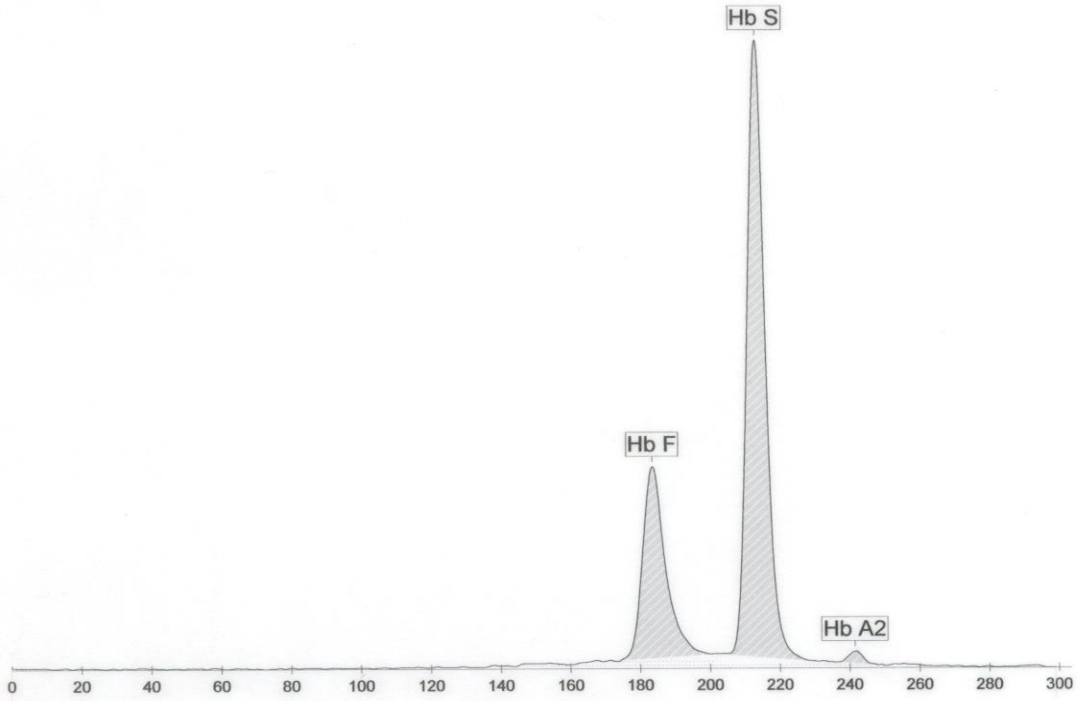
INSTRUMENT ID : KKUH : 24509

Hospital No.: **610043**

Sample No **52**

ID : **064229**

Date : **29/06/2010**



Fractions	%	Ref. %
Hb F	28.1	
Hb S	70.8	
Hb A2	1.1	

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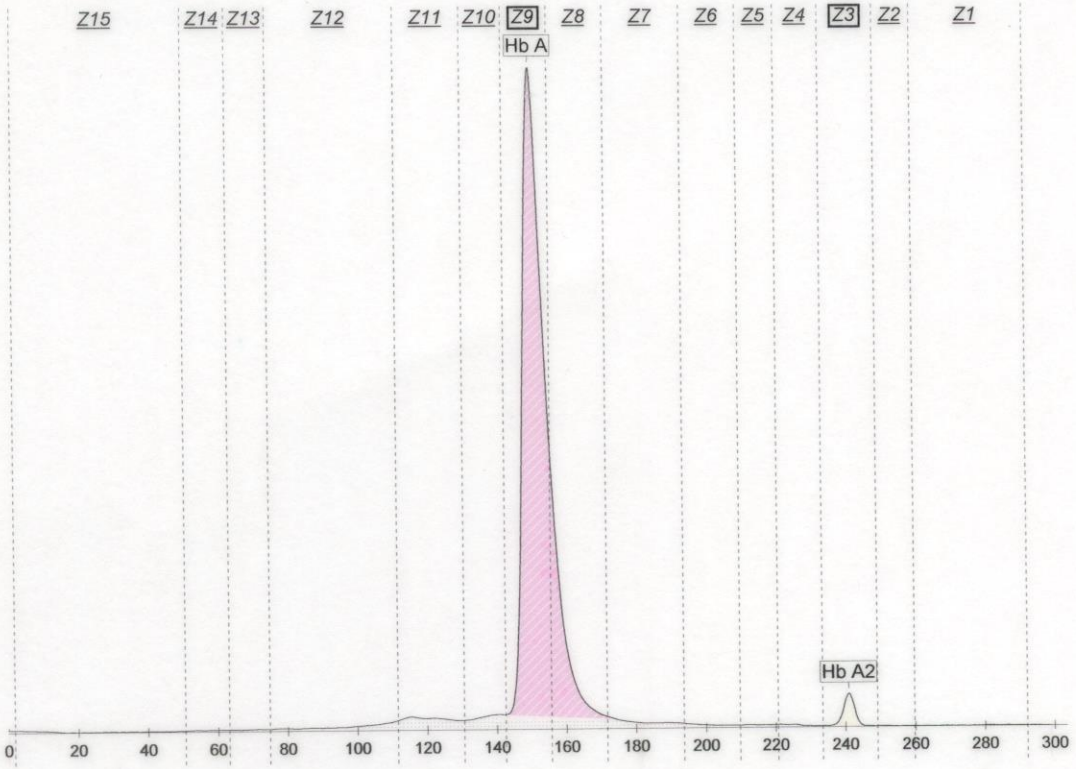
Heamatology Unit

Hb Electrophoresis

INSTRUMENT ID : KKUH : 24509

Hospital No.: **Rack: SEBIA Pos.: 2** ID : **ABDULLAH**

Sample No **20** Date : **19/05/2010**



Fractions	%	Ref. %
Hb A	**97.7**	95.0 - 99.0
Hb A2	**2.3**	1.5 - 3.5

KKUH

Heamatology Unit

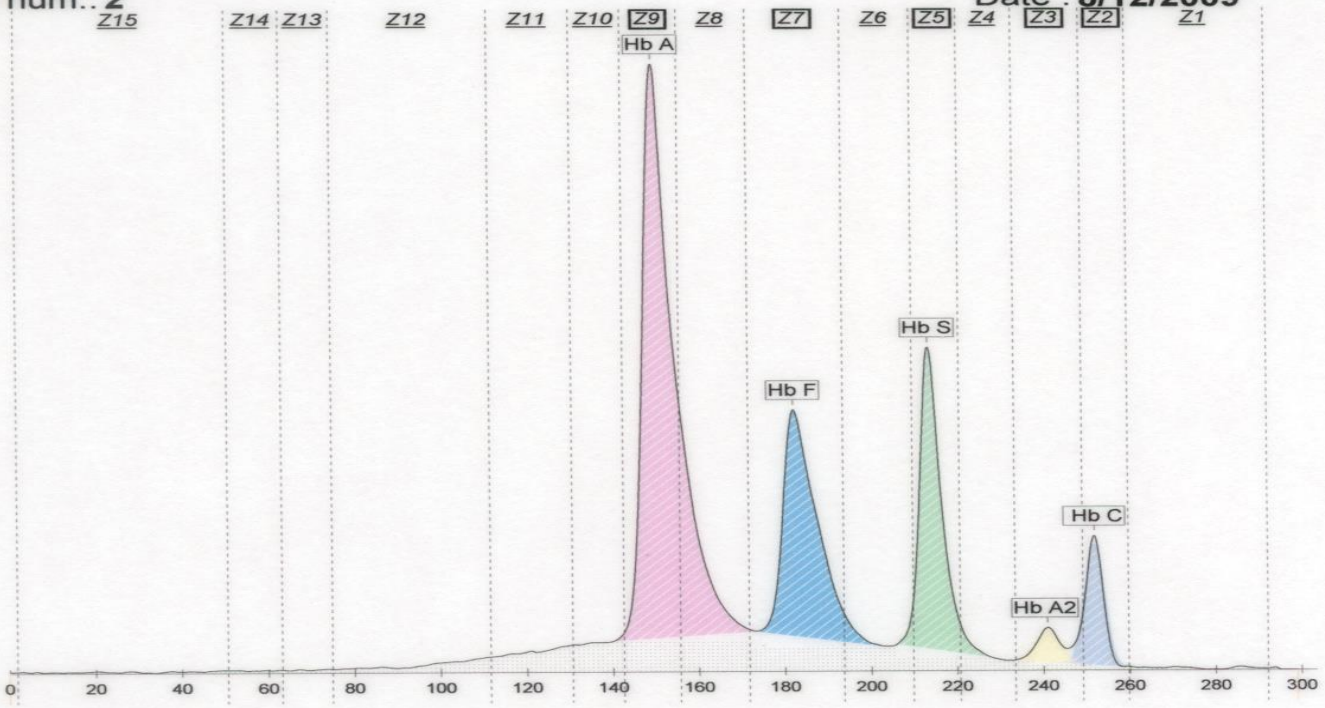
Hb Electrophoresis

Hospital No.: **QC Hb AFSC CONTROL-**

ID : **Hb AFSC CONTROL-2**

Sample num.: **2**

Date : **8/12/2009**



Hb Electrophoresis

Fractions	%	Ref. %
Hb A	**51.3**	46.7 - 56.9
Hb F	**21.4**	17.4 - 22.4
Hb S	**18.3**	17.3 - 22.3
Hb A2	**2.3**	2.1 - 3.3
Hb C	**6.7**	4.6 - 7.0

KKUH

Heamatology Unit

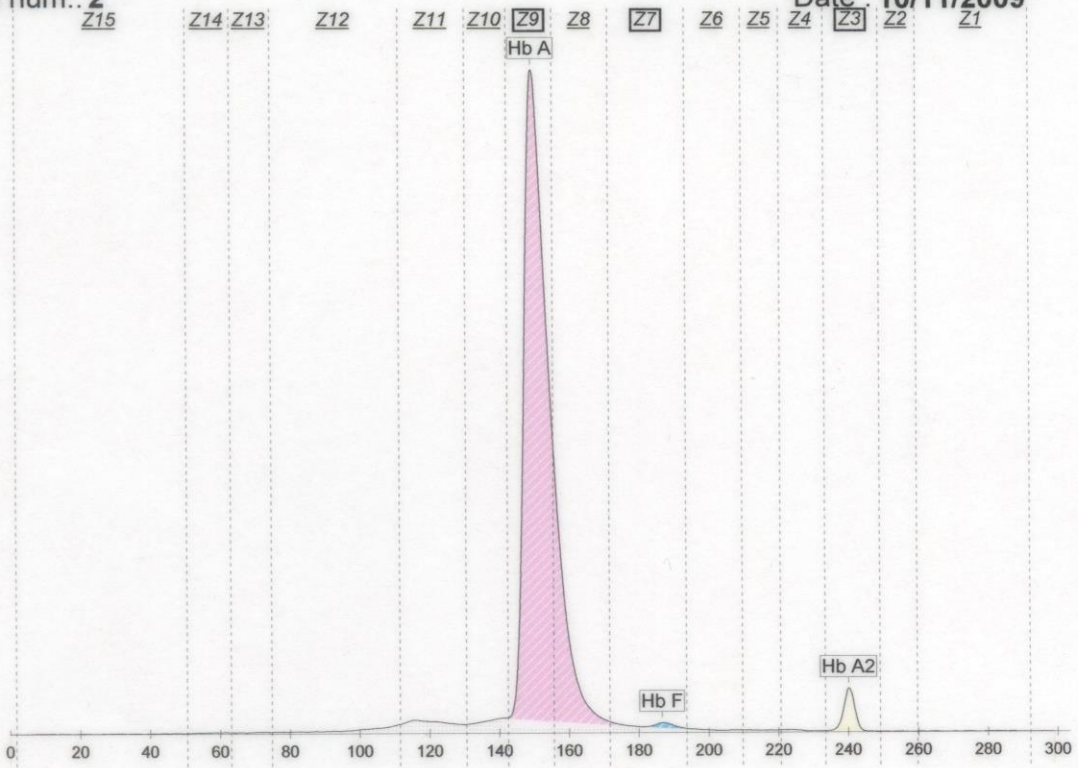
Hb Electrophoresis

Hospital No.: **933376**

ID : **061773**

Sample num.: **2**

Date : **10/11/2009**



Hb Electrophoresis

Fractions	%	Ref. %	
Hb A	**96.7**	96.8 - 97.8	
Hb F	**0.5**	=< 2.0	<
Hb A2	**2.8**	1.5 - 3.5	

KKUH

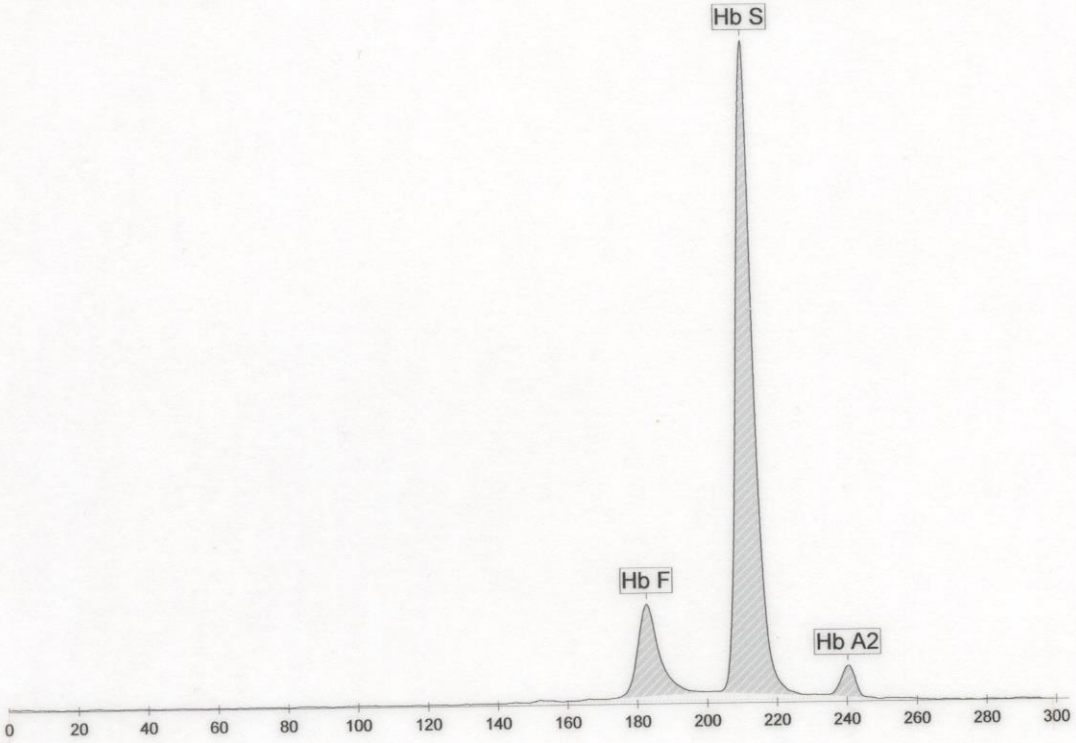
Heamatology Unit

Hb Electrophoresis

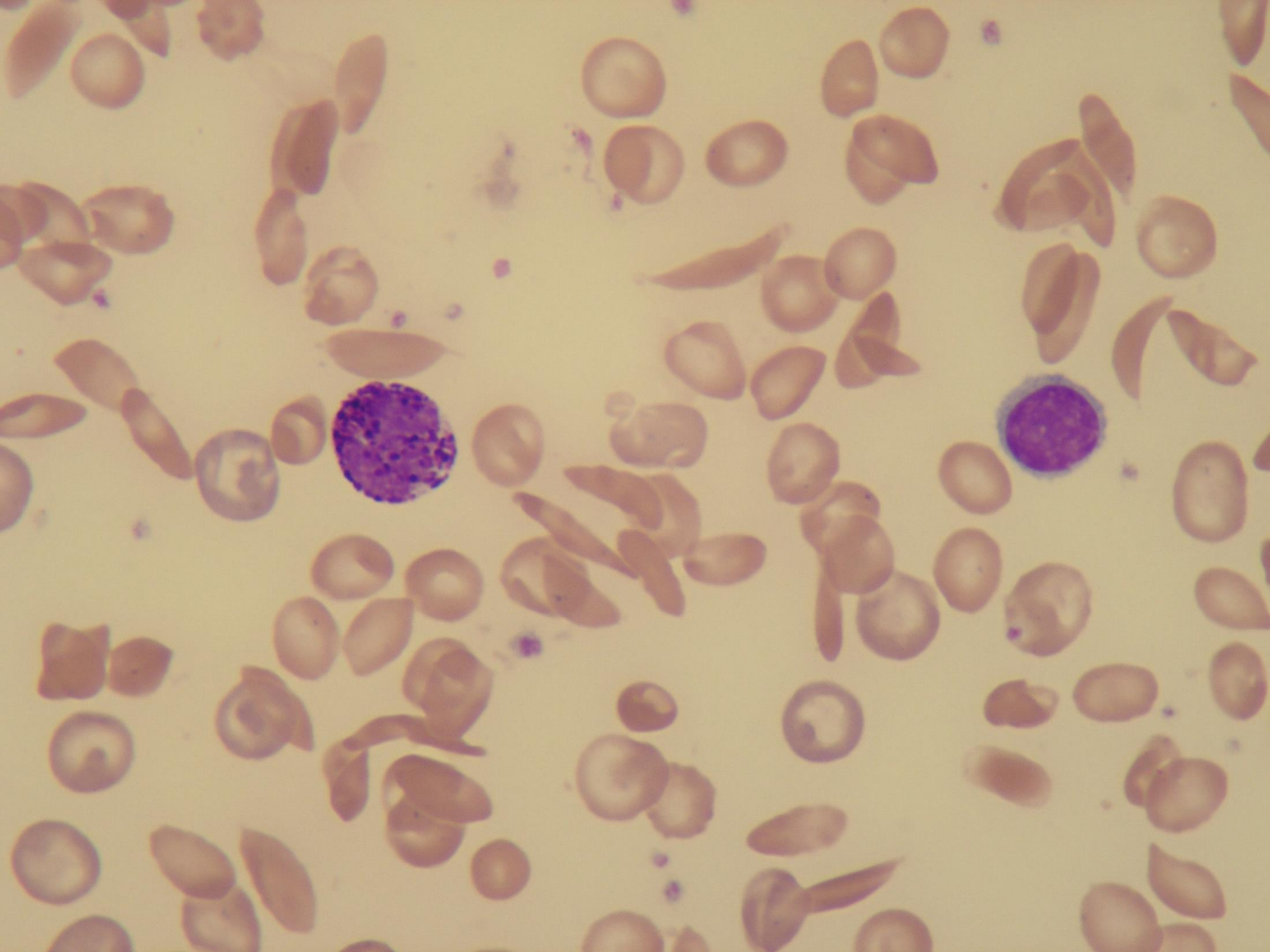
INSTRUMENT ID : KKUH : 24509

Hospital No.: **873506** ID : **064230**

Sample No **53** Date : **29/06/2010**



Fractions	%	Ref. %
Hb F	**14.5**	
Hb S	**82.2**	
Hb A2	**3.3**	



KKUH

Heamatology Unit

Hb Electrophoresis

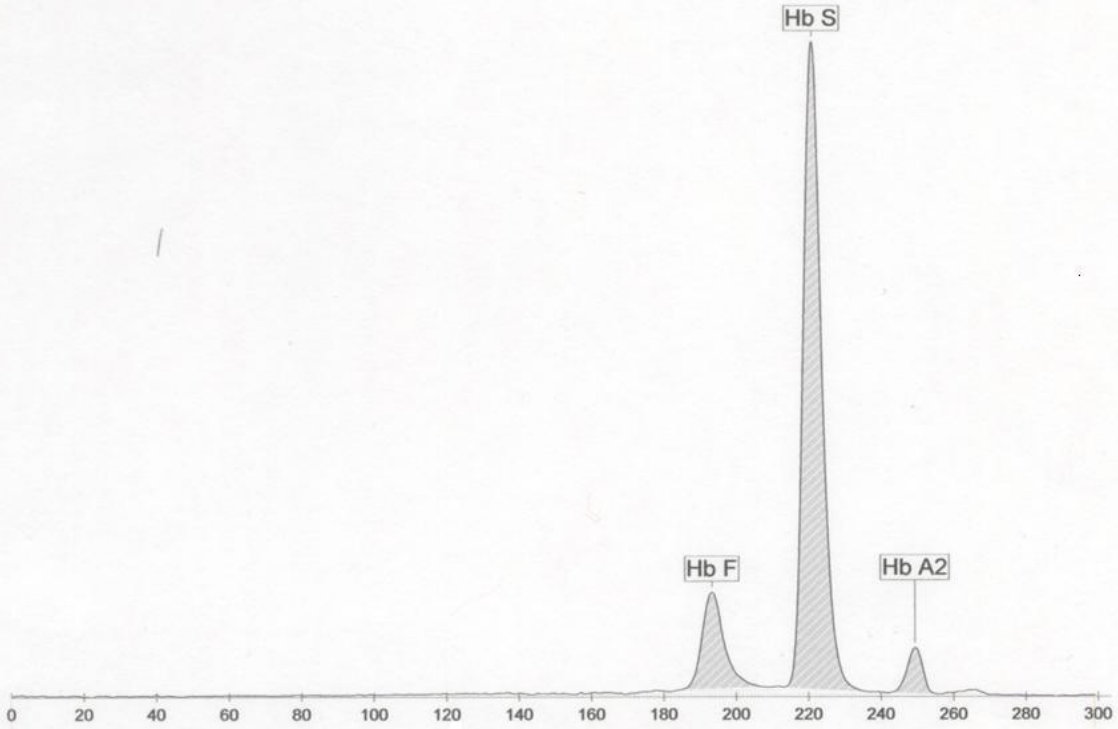
INSTRUMENT ID : KKUH : 24509

Hospital No.: **233095**

Sample No **20**

ID : **063478**

Date : **17/04/2010**



Fractions	%	Ref. %
Hb F	**14.7**	
Hb S	**80.5**	
Hb A2	**4.8**	

KKUH

Heamatology Unit

Hb Electrophoresis

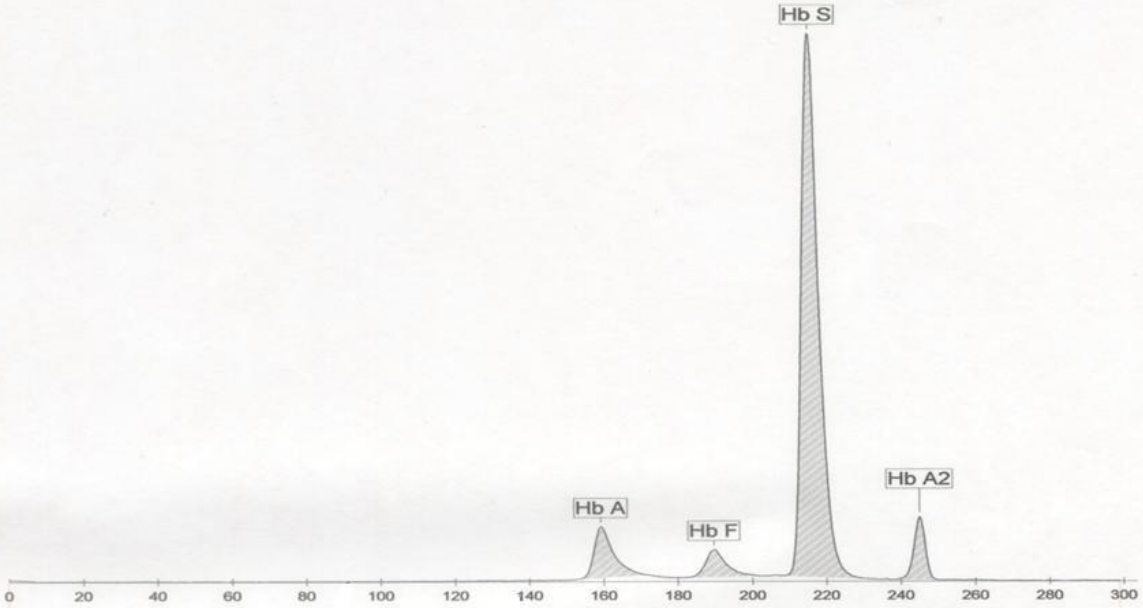
INSTRUMENT ID : KKUH : 24509

Hospital No.: **913628**

Sample No **34**

ID : **063511**

Date : **19/04/2010**



Fractions	%	Ref. %
Hb A	**8.7**	
Hb F	**4.9**	
Hb S	**80.1**	
Hb A2	**6.3**	

Comment :

KKUH

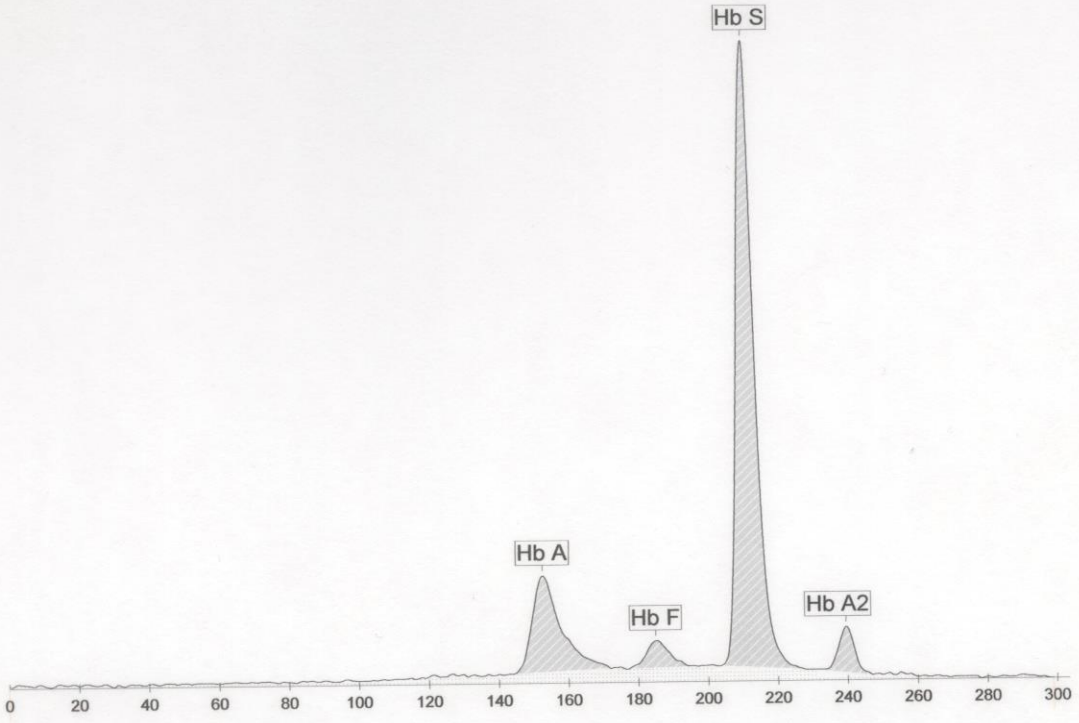
Heamatology Unit

Hb Electrophoresis

INSTRUMENT ID : KKUH : 24509

Hospital No.: **010755** ID : **064209**

Sample No **19** Date : **28/06/2010**



Fractions	%	Ref. %
Hb A	**18.0**	
Hb F	**4.0**	
Hb S	**73.3**	
Hb A2	**4.7**	

KKUH

Heamatology Unit

Hb Electrophoresis

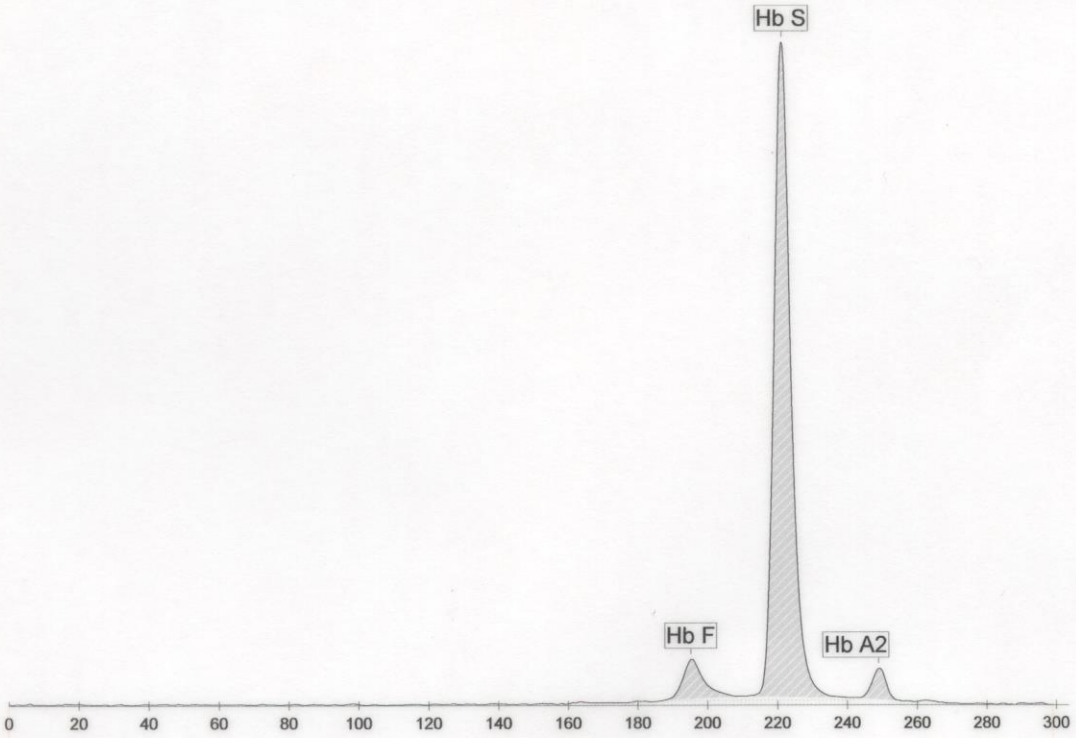
INSTRUMENT ID : KKUH : 24509

Hospital No.: **594729**

Sample No **37**

ID : **064199**

Date : **27/06/2010**



Fractions	%	Ref. %
Hb F	**6.5**	
Hb S	**89.9**	
Hb A2	**3.6**	

KKUH

Heamatology Unit

Hb Electrophoresis

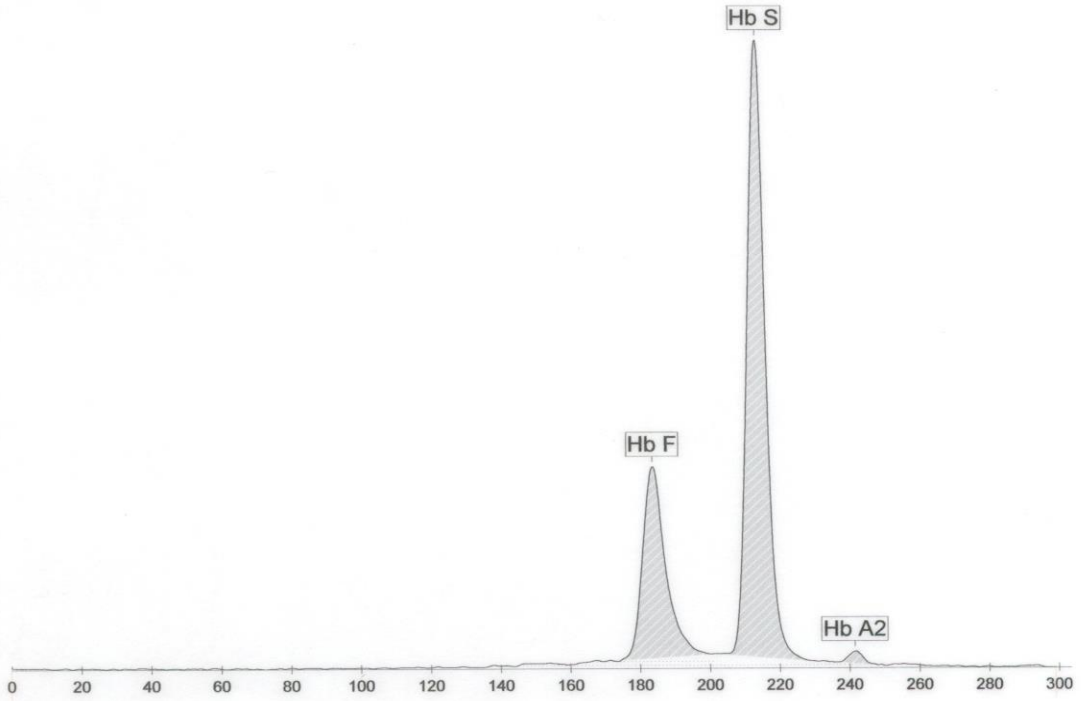
INSTRUMENT ID : KKUH : 24509

Hospital No.: **610043**

Sample No **52**

ID : **064229**

Date : **29/06/2010**



Fractions	%	Ref. %
Hb F	28.1	
Hb S	70.8	
Hb A2	1.1	

KKUH

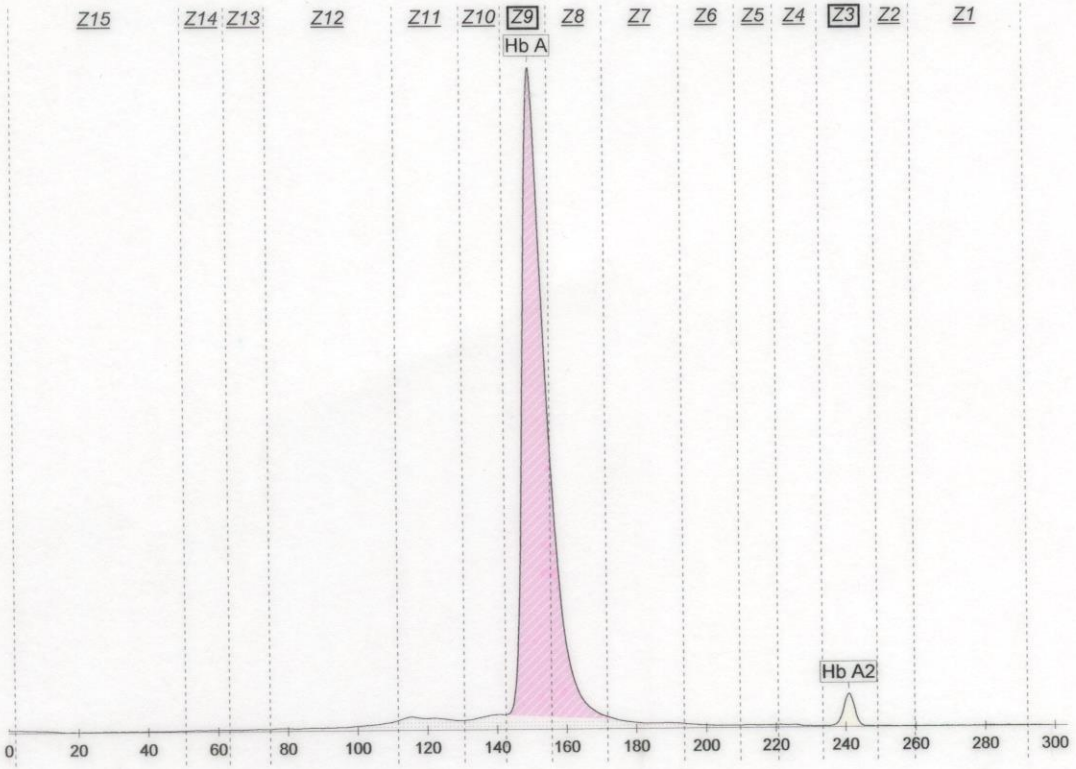
Heamatology Unit

Hb Electrophoresis

INSTRUMENT ID : KKUH : 24509

Hospital No.: **Rack: SEBIA Pos.: 2** ID : **ABDULLAH**

Sample No **20** Date : **19/05/2010**



Fractions	%	Ref. %
Hb A	**97.7**	95.0 - 99.0
Hb A2	**2.3**	1.5 - 3.5

KKUH

Heamatology Unit

Hb Electrophoresis

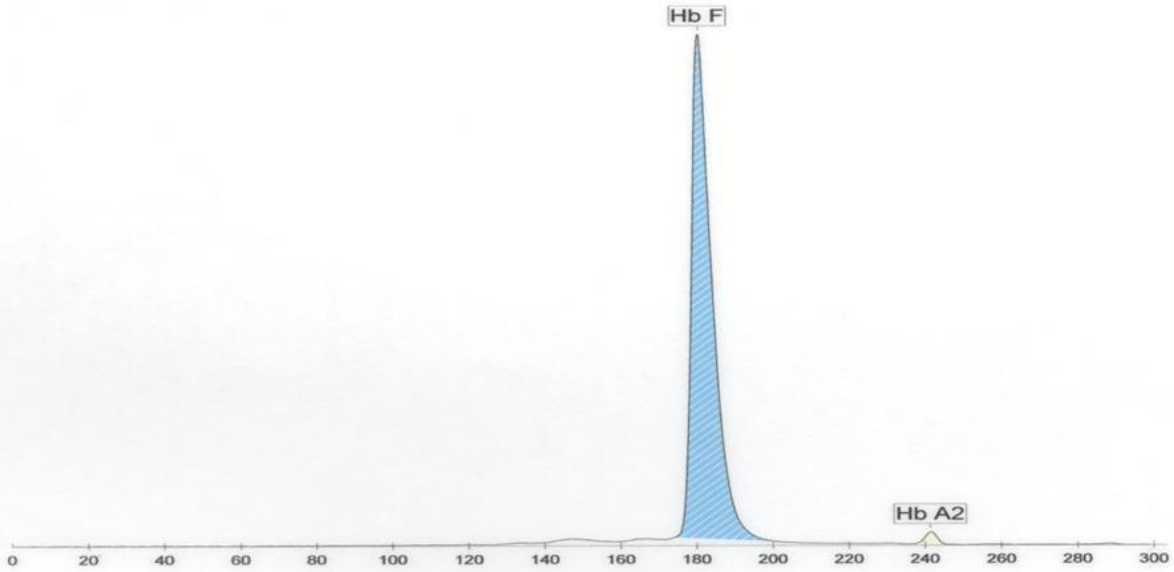
INSTRUMENT ID : KKUH : 24509

Hospital No.: **921107**

Sample No **54**

ID : **063761**

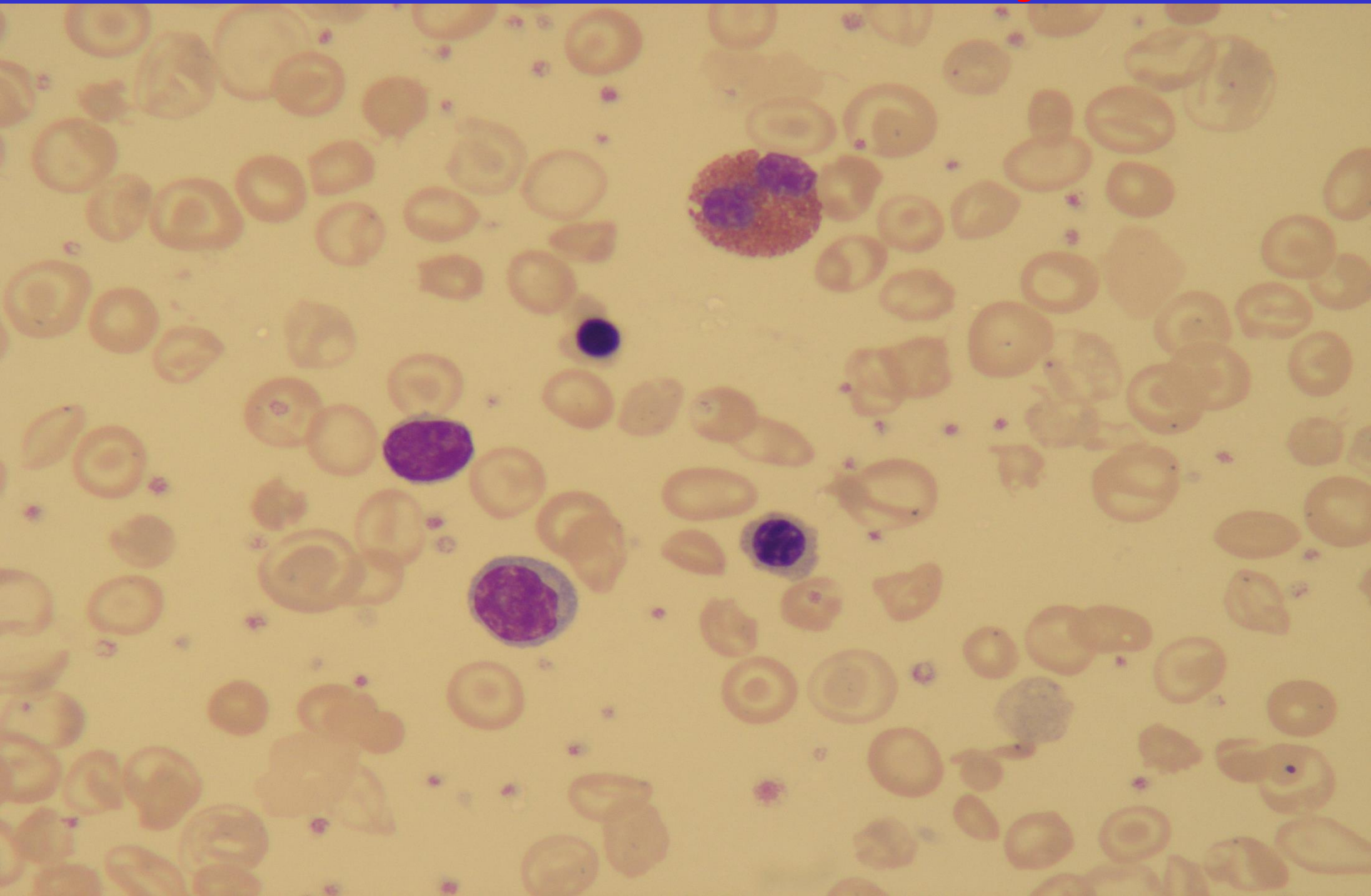
Date : **09/05/2010**



Fractions	%	Ref. %
Hb F	**98.5**	
Hb A2	**1.5**	

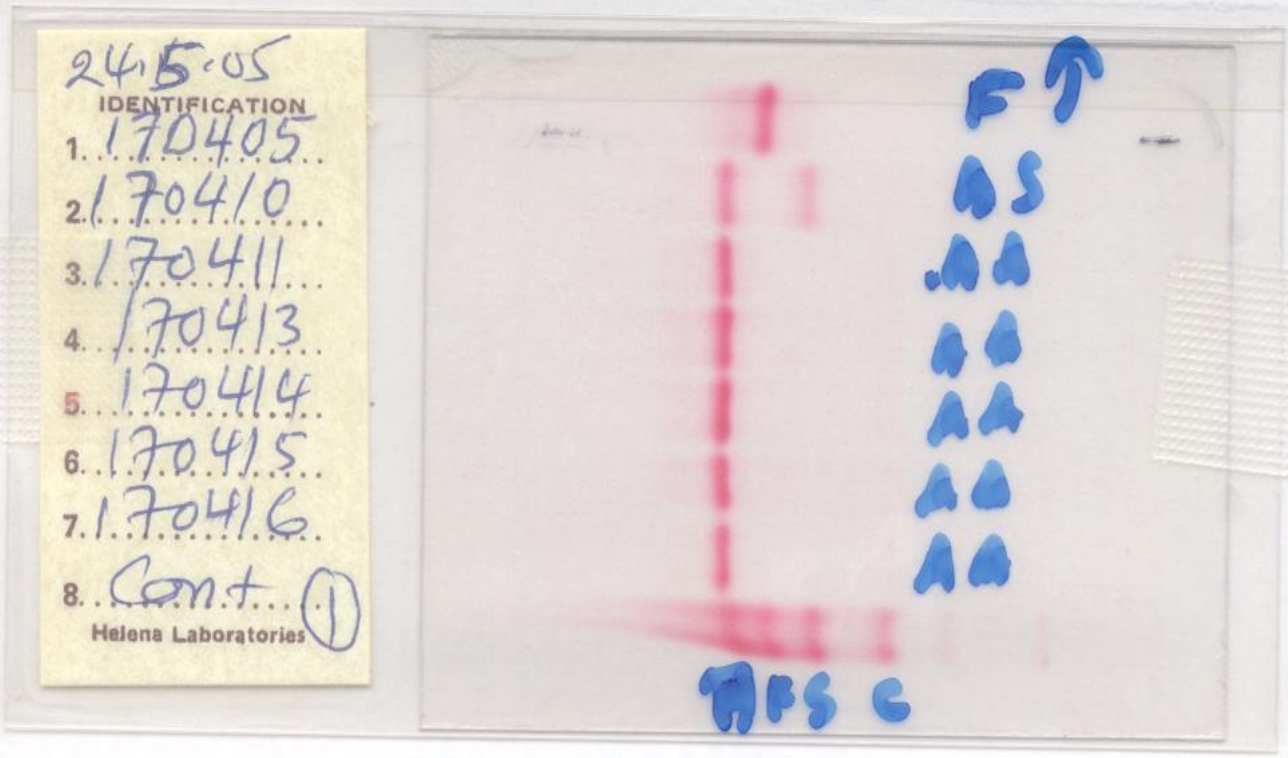
Comment :

Beta Thalassaemia Major



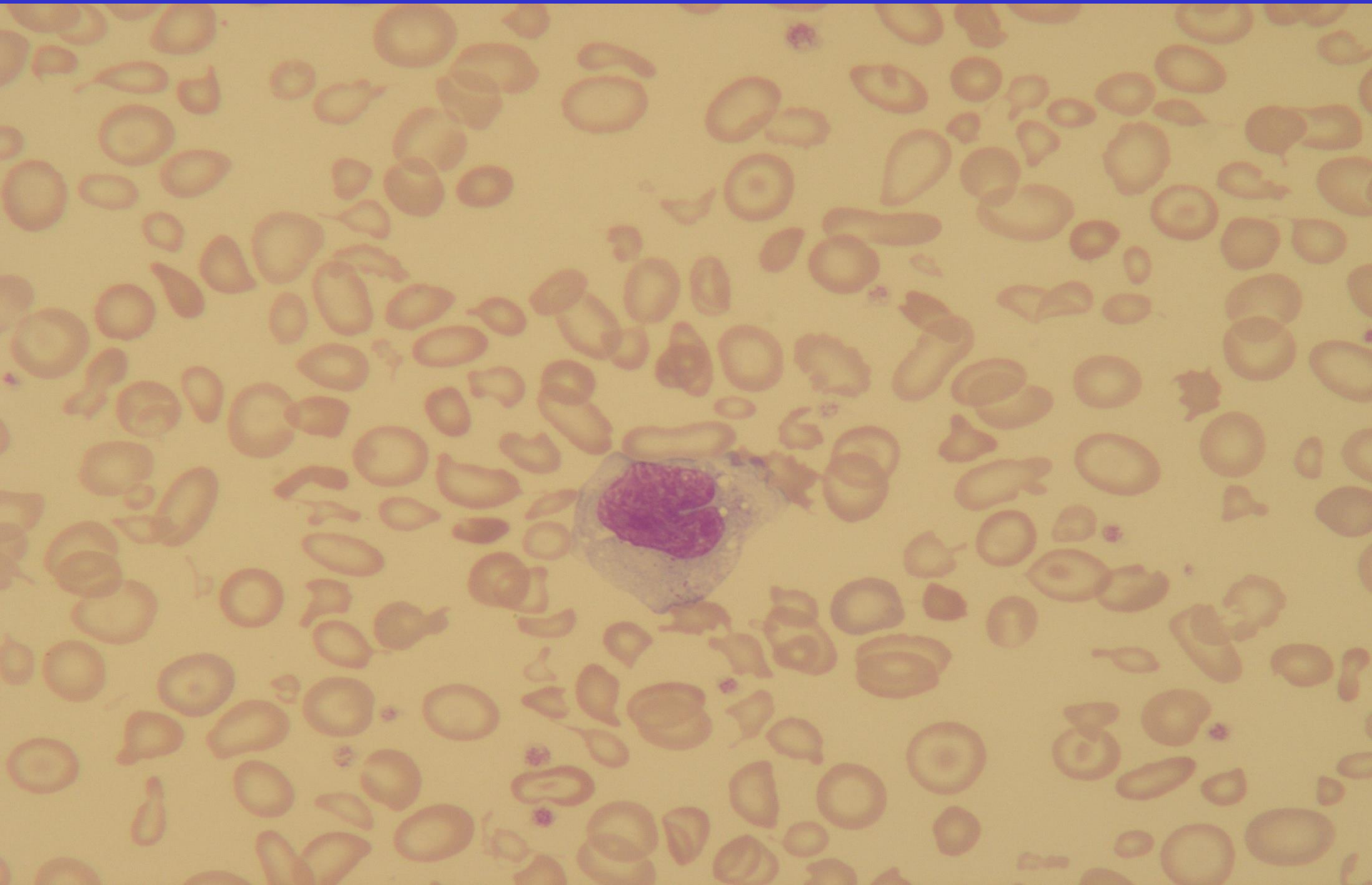
(+)

(−)

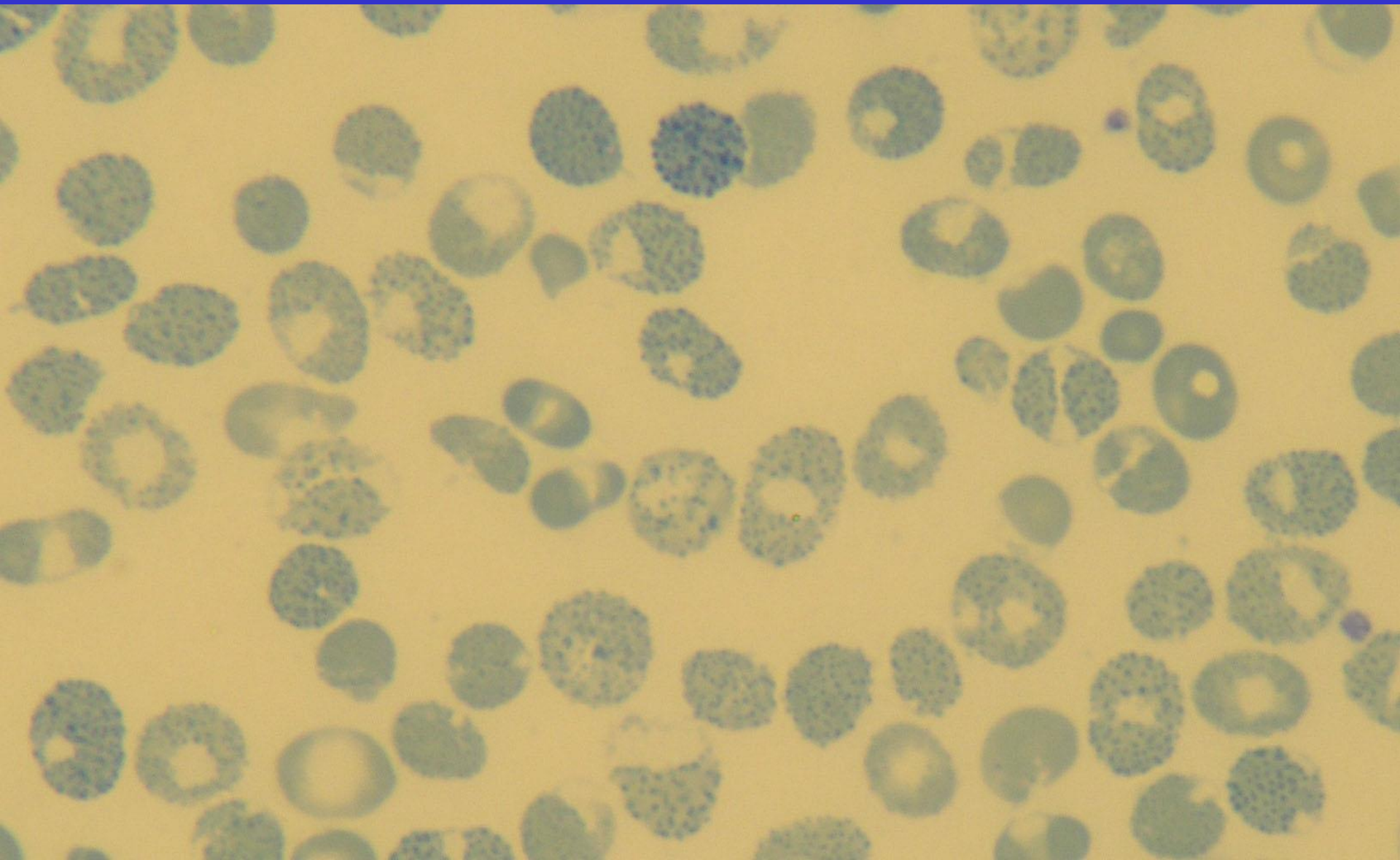

24.5.05
IDENTIFICATION
1. 170405
2. 170410
3. 170411
4. 170413
5. 170414
6. 170415
7. 170416
8. Cont. (1)
Helena Laboratories
F
AFS C


(+)
(−)
9.5.05
IDENTIFICATION
1. 170368
2. 170369
3. 684245
4. Cont...
5.
6.
7. (2)
8.
Helena Laboratories
SS
AA
SS
AH
AA
origin
H
S
C

Alpha Thalassaemia



Haemoglobin H Disease



(+) ⟵ (−)

10.2.2001

IDENTIFICATION

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2. 1270
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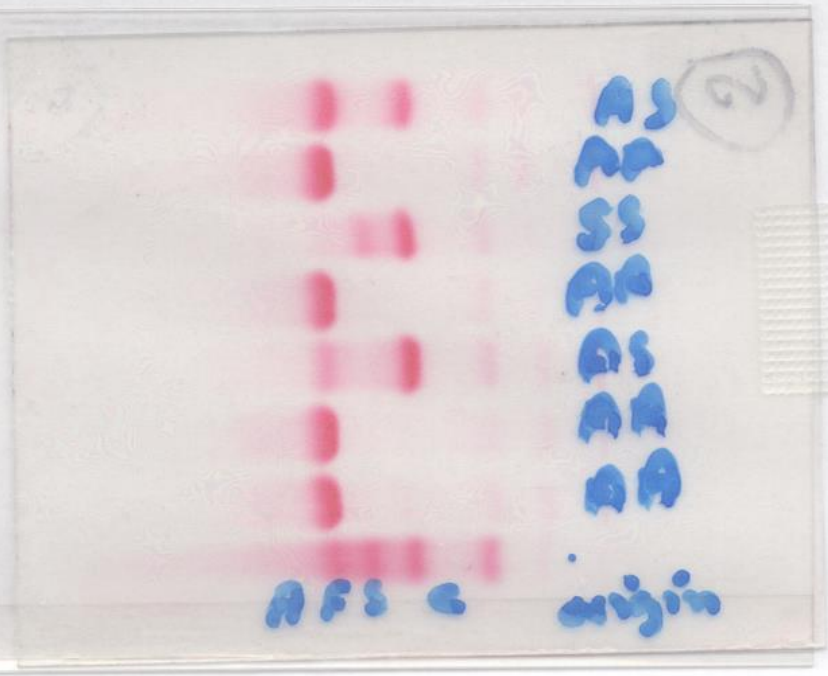
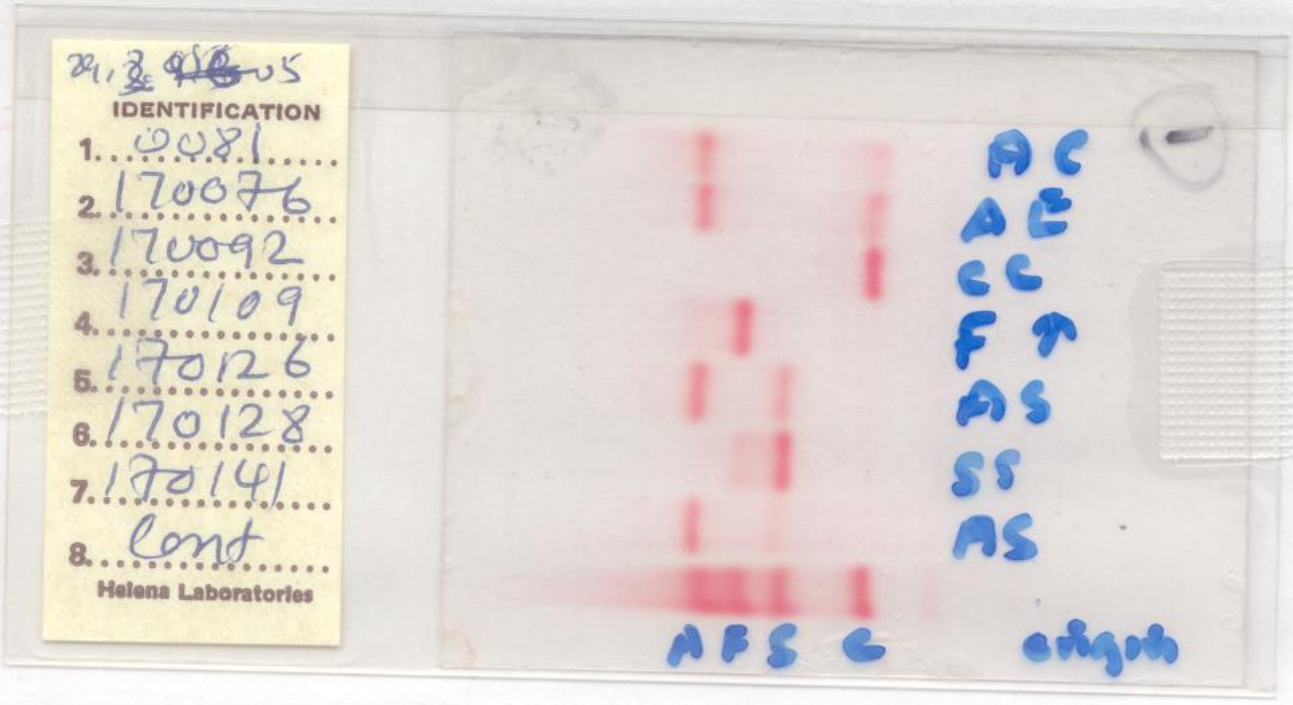
4. 1283
5. 1289
6. 6013
7. 6014
8. Cont.

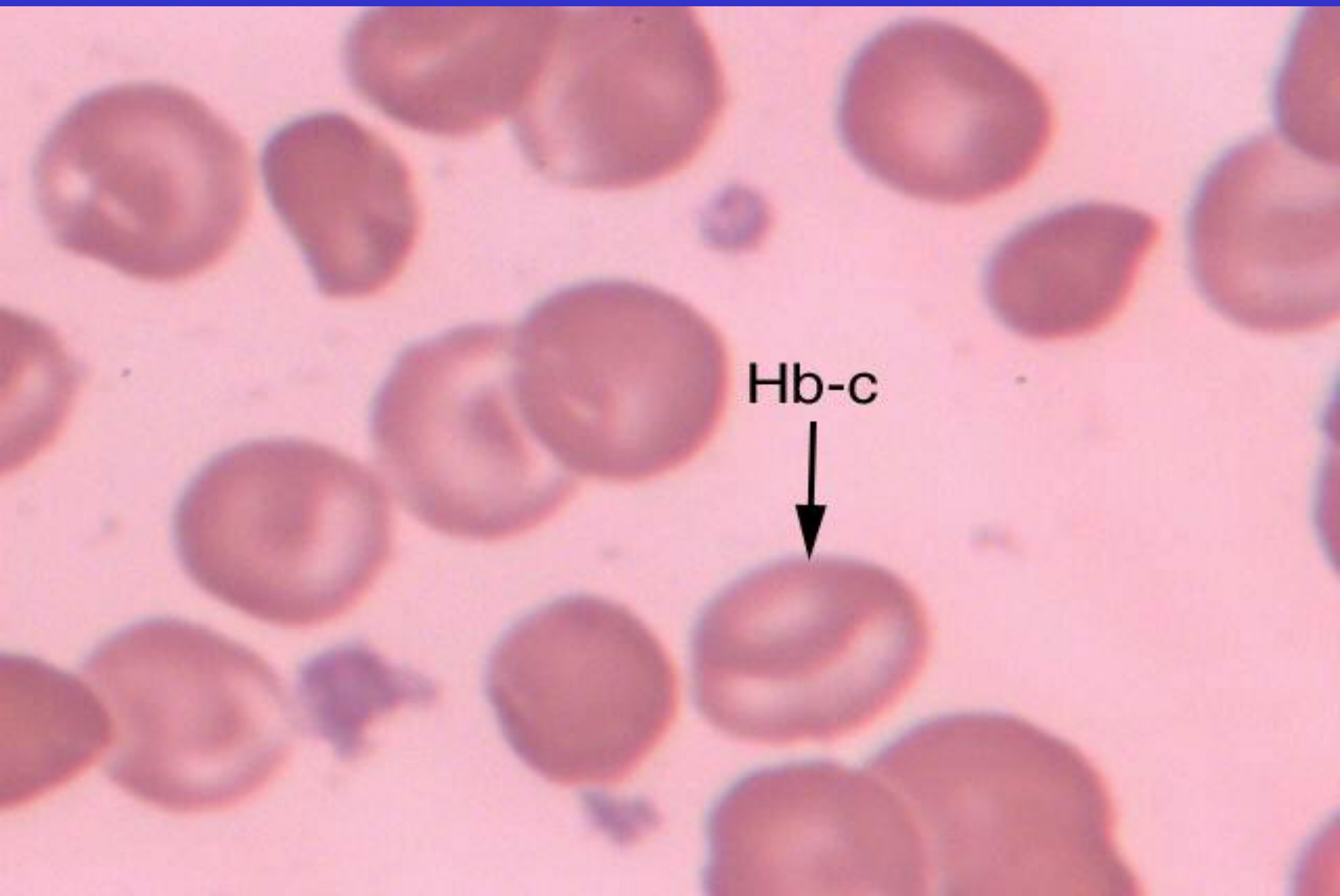
Helena Laboratories (2)



(+) ⟵ (-)



HAEMOGLOBIN C DISEASE



Thank you