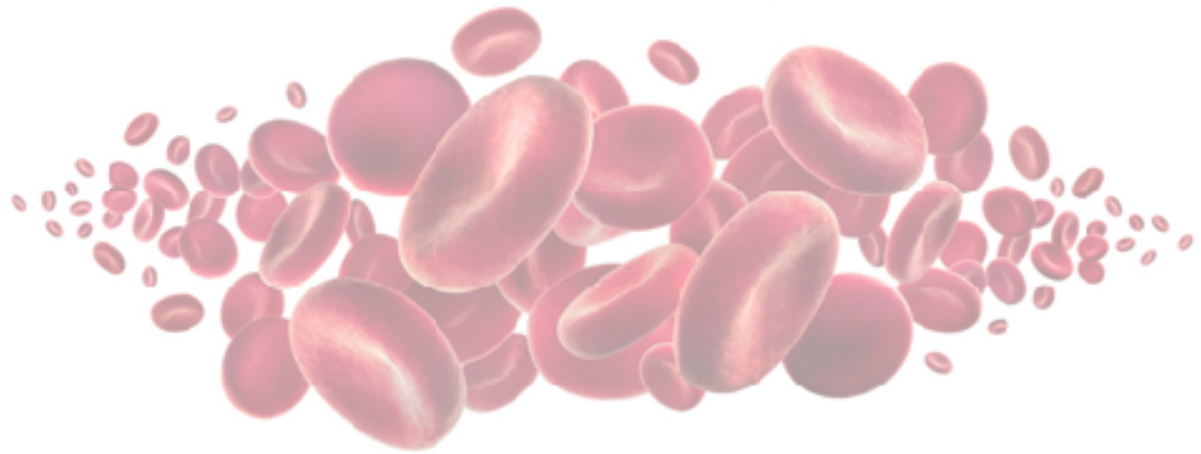

Common Issue in Hematology

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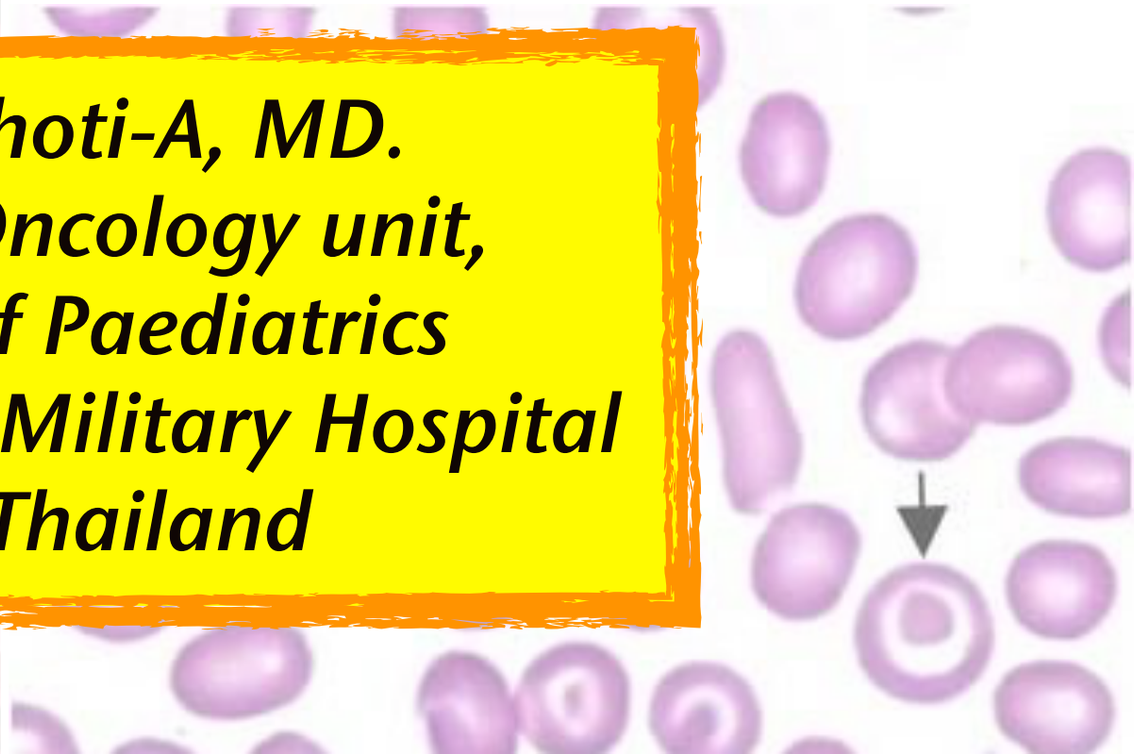
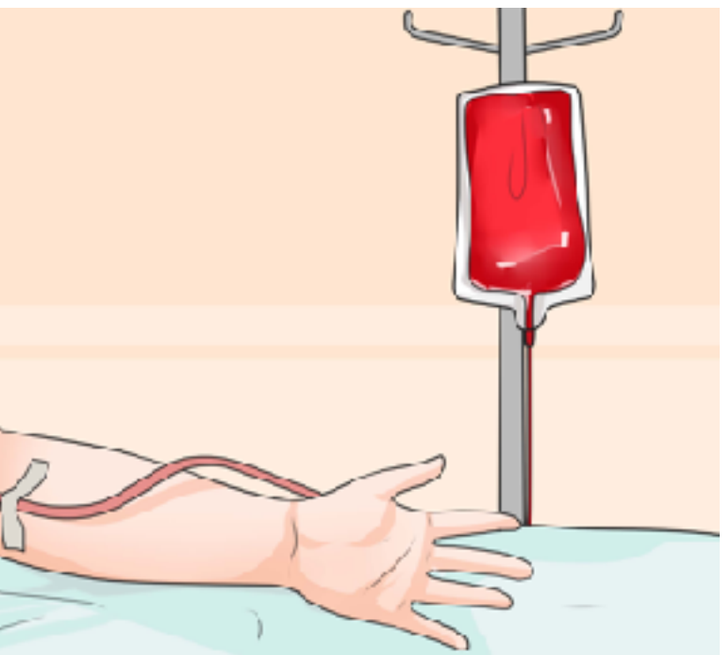
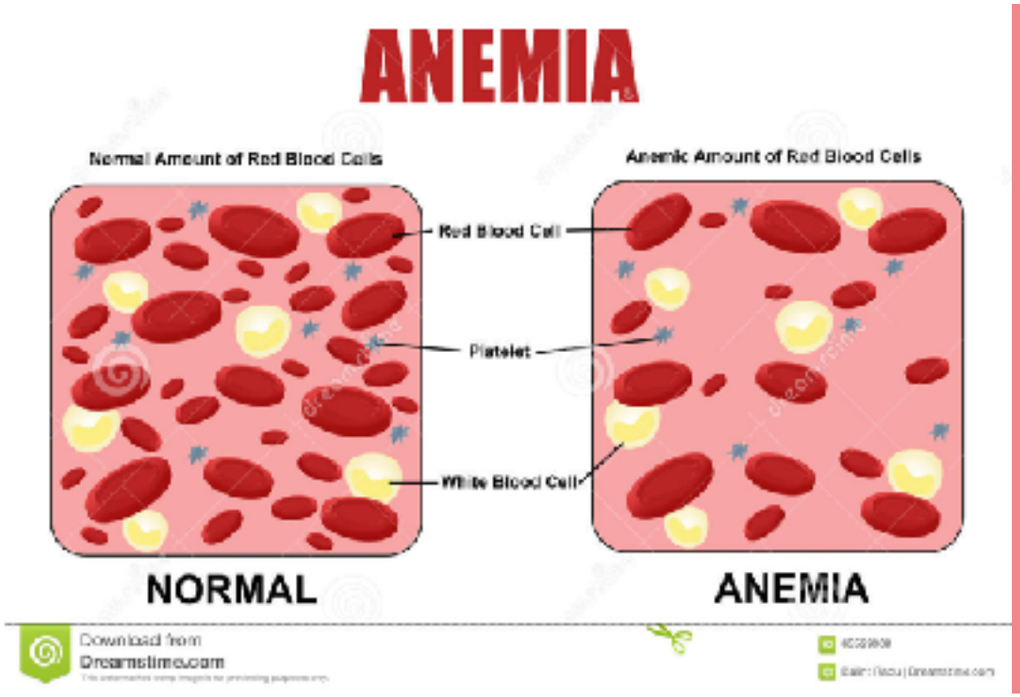
Bangkok, Thailand

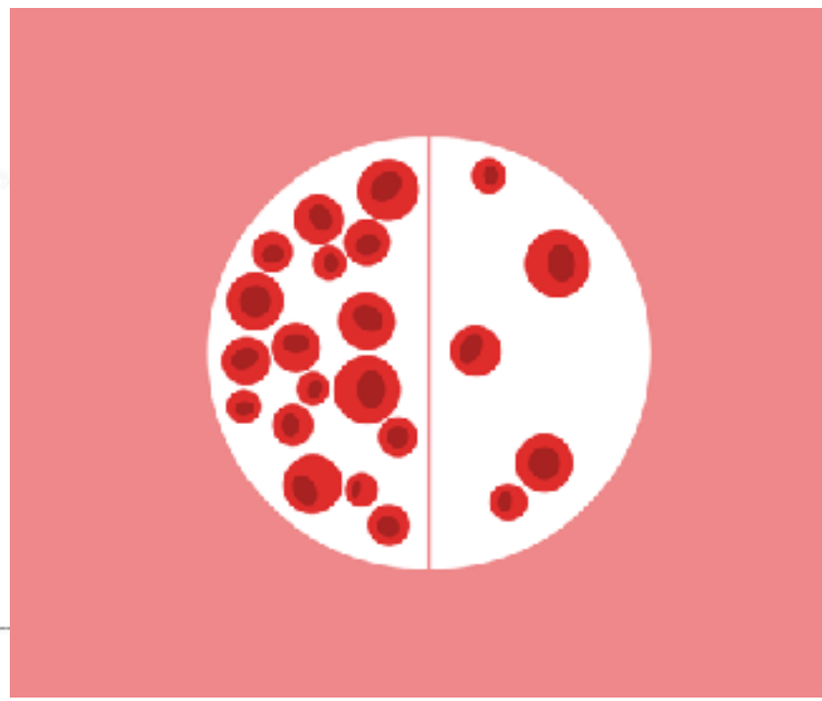


Learning objectives

- **Approach to anemia**
- **What you should know about “70 baht” CBC**
- **Hemoglobin typing**
- **Thalassemia**
- **Rational use of blood component - The concept**







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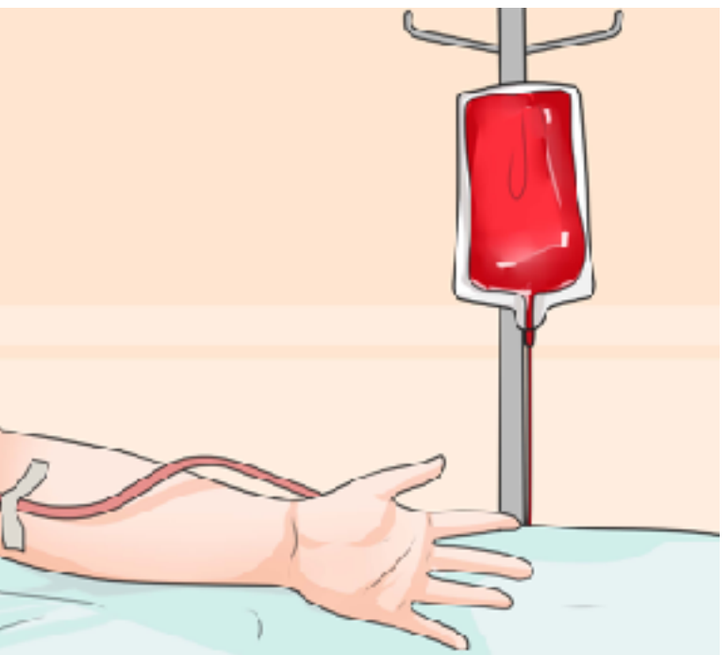
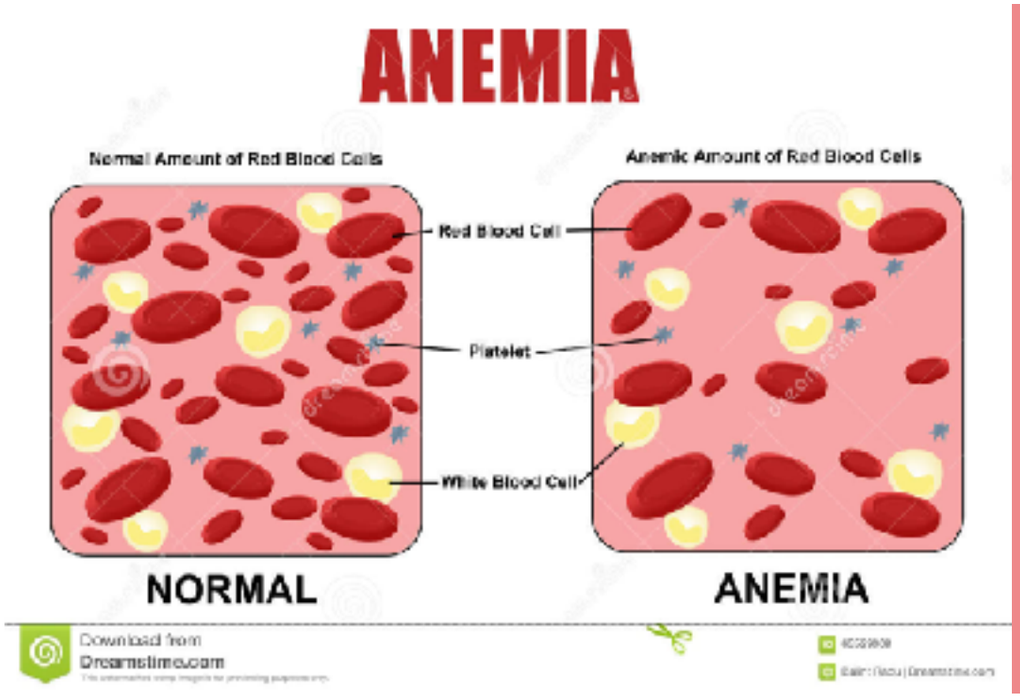
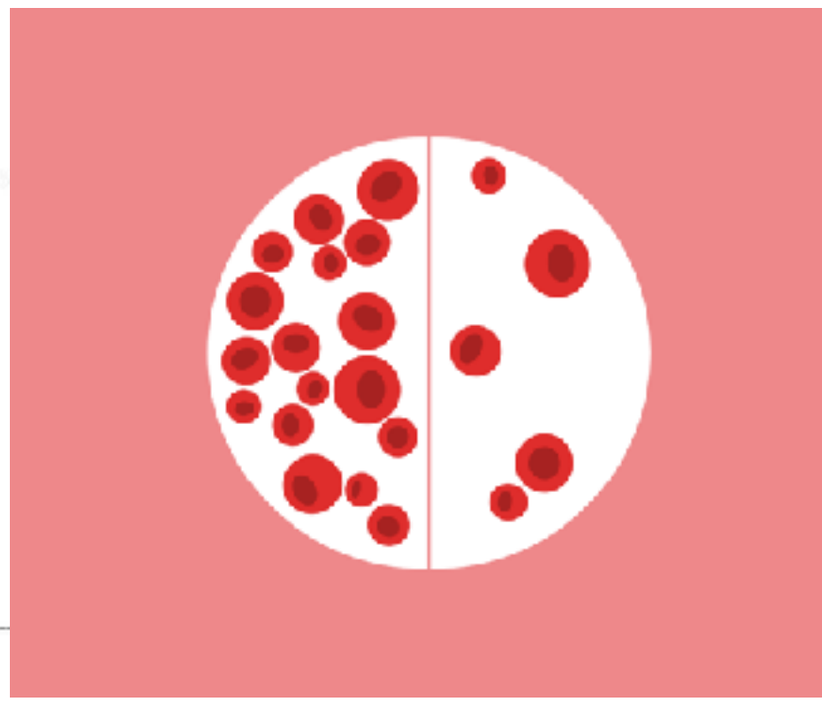
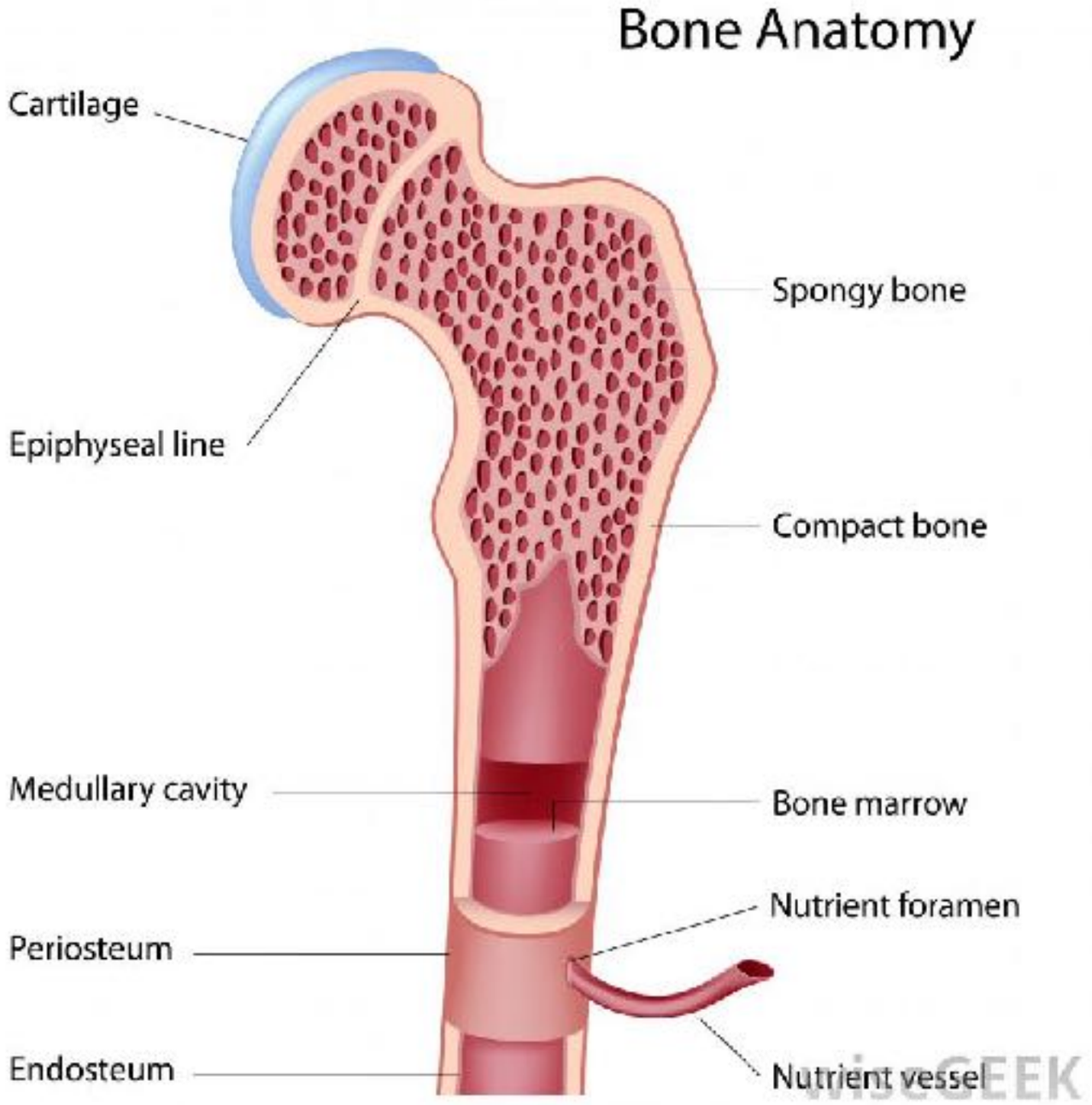
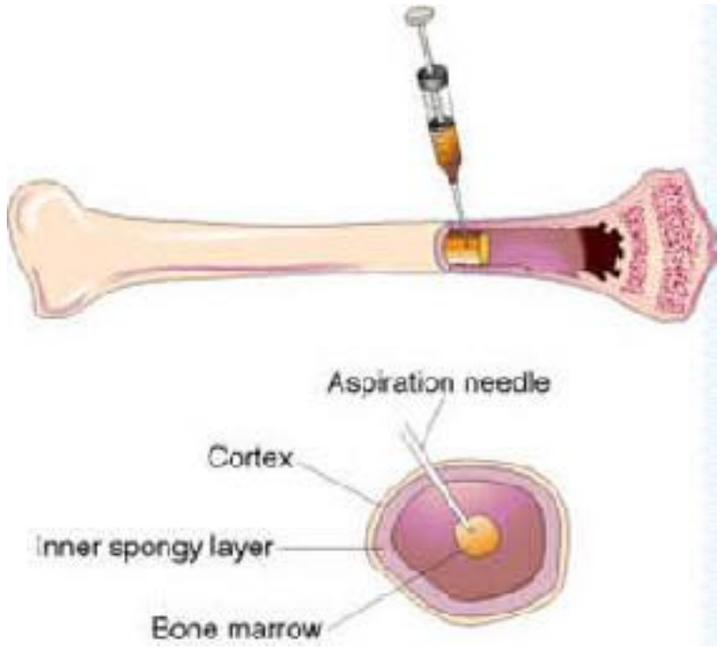




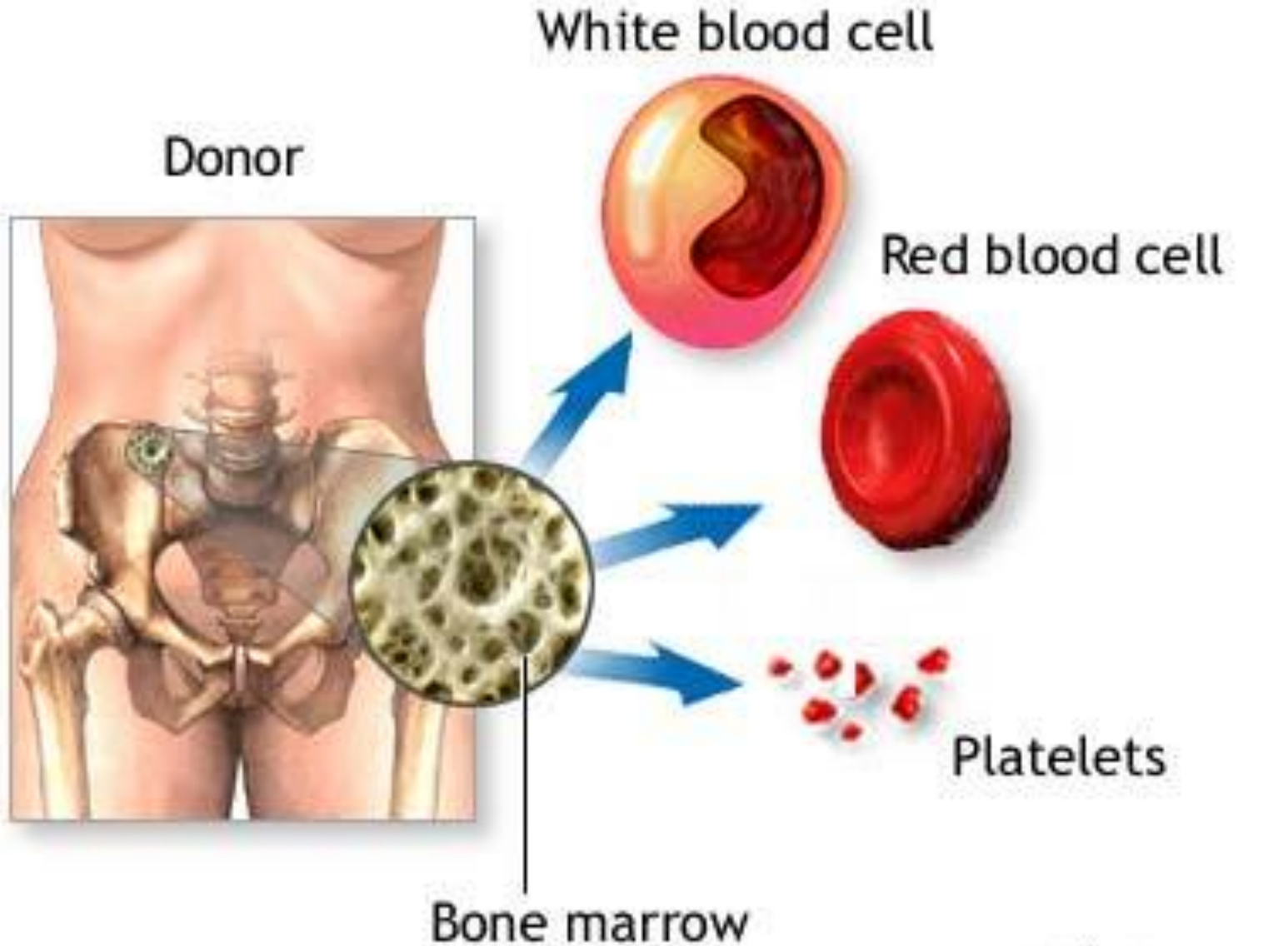


Bone Marrow


Bone Anatomy
Cartilage
Spongy bone
Epiphyseal line
Compact bone
Medullary cavity
Bone marrow
Nutrient foramen
Periosteum
Endosteum
Nutrient vessel
wiseGEEK


Aspiration needle
Cortex
Inner spongy layer
Bone marrow

Bone Marrow


White blood cell
Donor
Red blood cell
Platelets
Bone marrow
A.D.A.M.

Erythropoiesis

Bone marrow

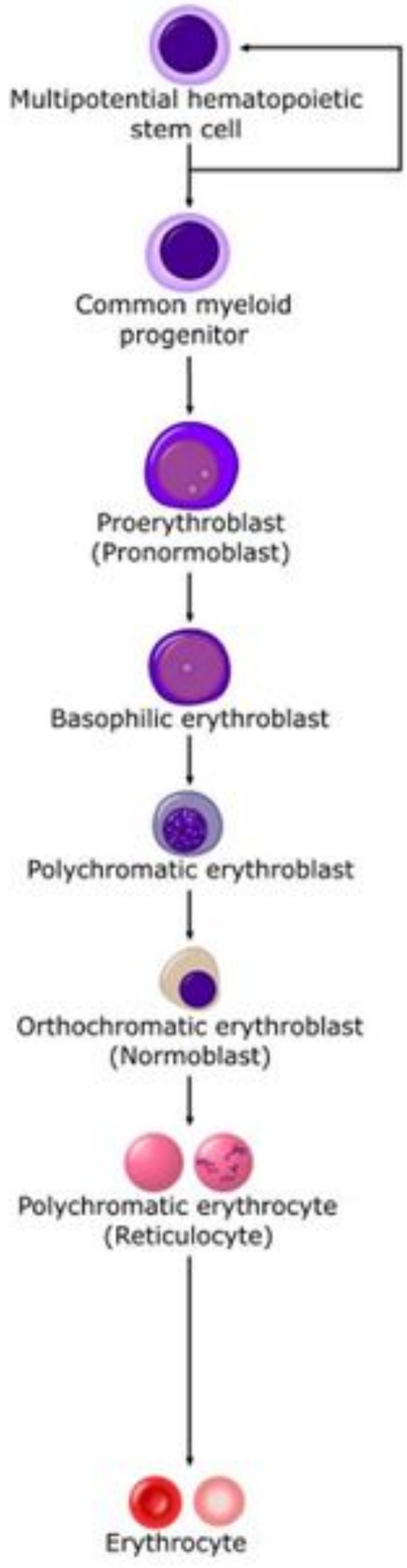
Pluripotent stem cells

Chemical regulation

- ***Cytokines***
- ***Erythroid specific growth factor***
- ***Erythropoietin (EPO)***

Life span

- ***Reticulocyte – 4 days***
- ***RBC – 120 days***



Definition

		Hb (g/dL)	Hct (mg%)
Child	6 mo - 6 yrs	11	33
	6 yrs - 14 yrs	12	36
Adult	Male	13	39
	Female	12	36
	Pregnant 1st,3rd trimester	11	33
	Pregnant 2nd trimester	10.5	33

Ref : Corbett&Buchsel, 2005

Evaluation of the patient

HISTORY

- **Is the bone marrow suppressed?**
- **Is the patient nutritionally deficient? Pica?**

Decreased Production

Evaluation of the patient

HISTORY

- **Is the bone marrow suppressed?**
- **Is the patient nutritionally deficient? Pica?**
- **Is there evidence for increased RBC destruction?**

Increased Degradation

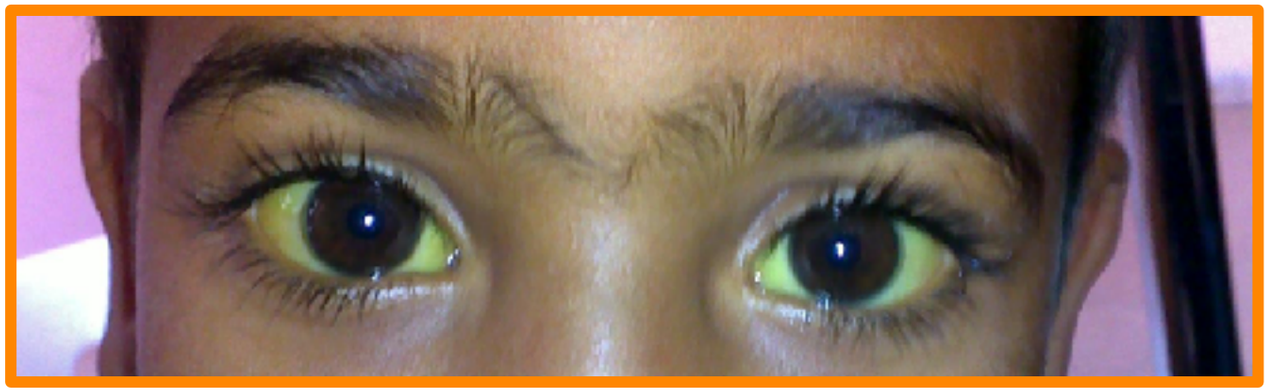
Evaluation of the patient

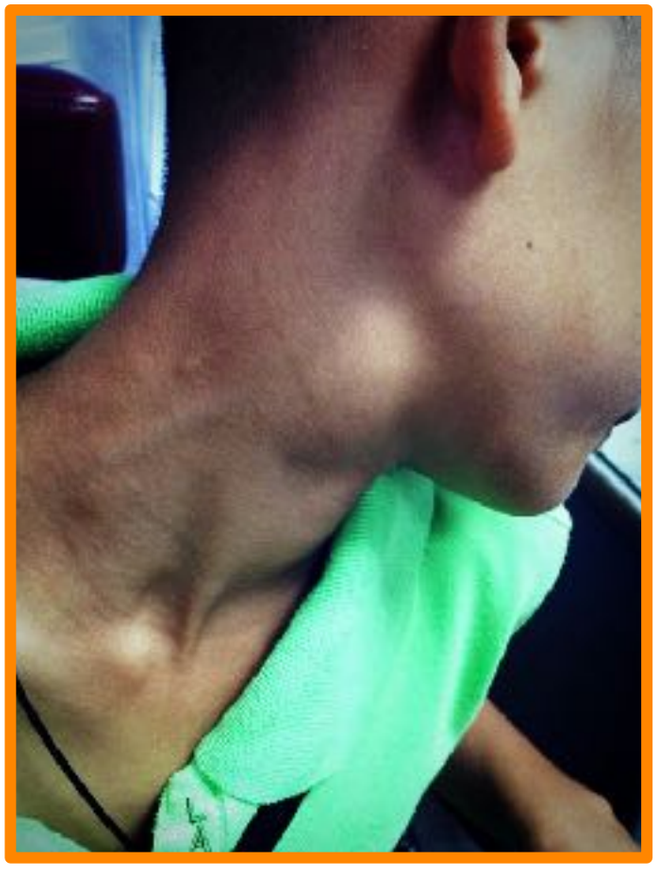
HISTORY

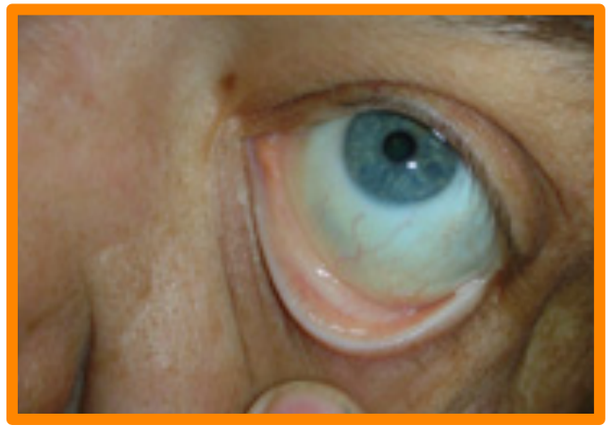
- **Is the bone marrow suppressed?**
- **Is the patient nutritionally deficient? Pica?**
- **Is there evidence for increased RBC destruction?**
- **Is the patient bleeding?**
  - **Actively? In past?**

Blood Loss

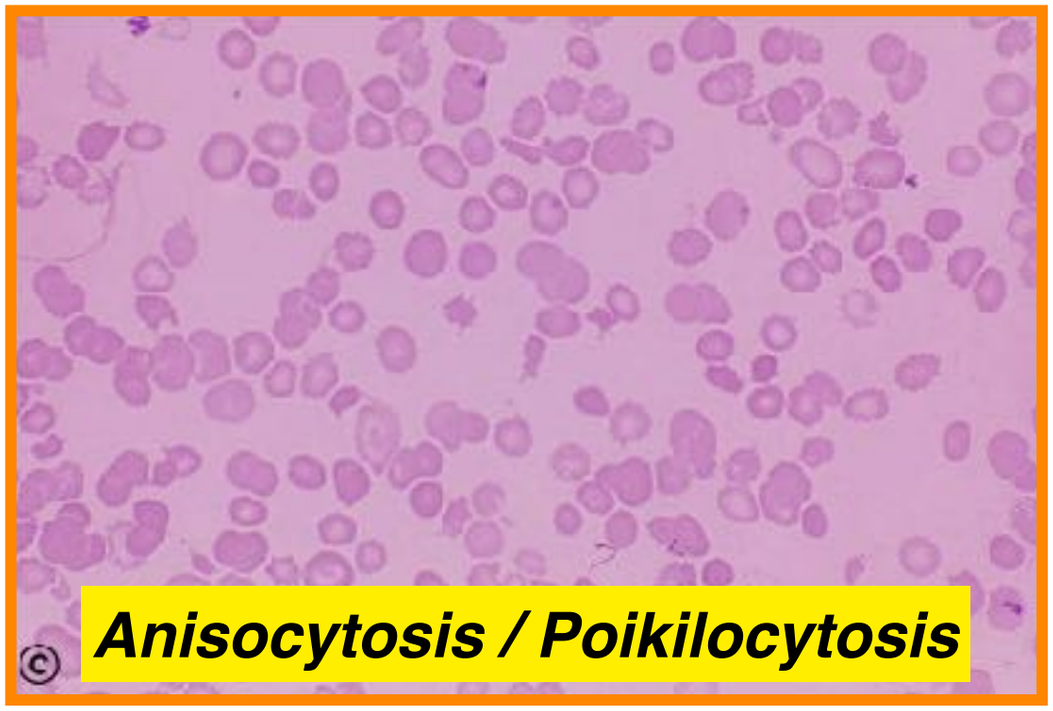
Physical Examination

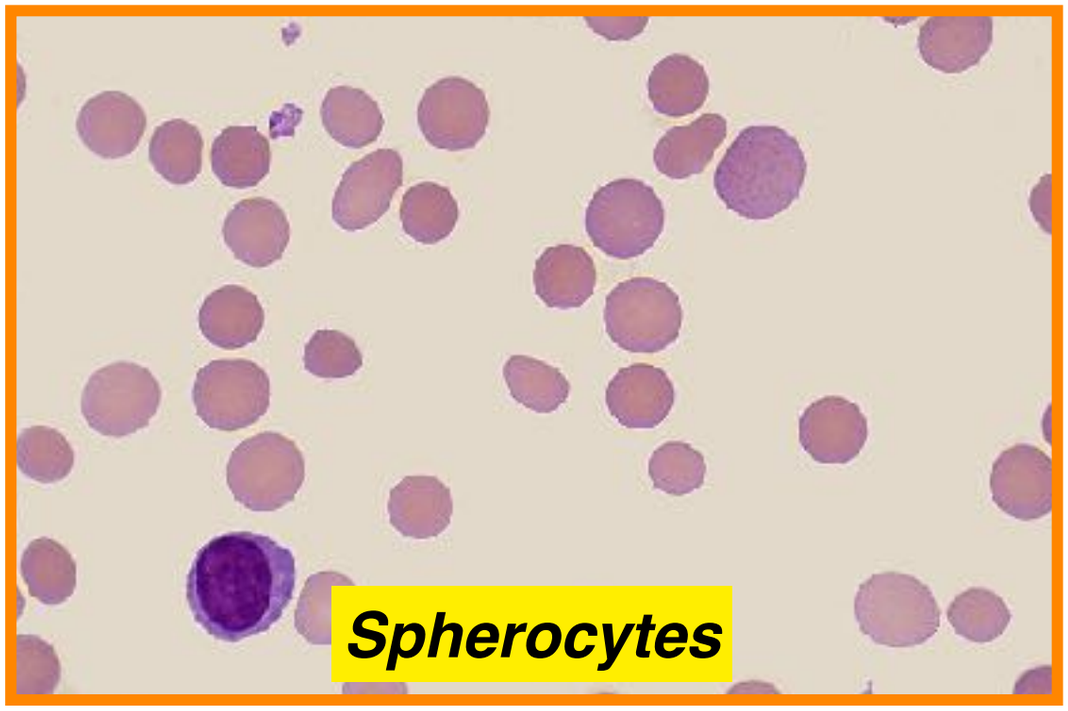


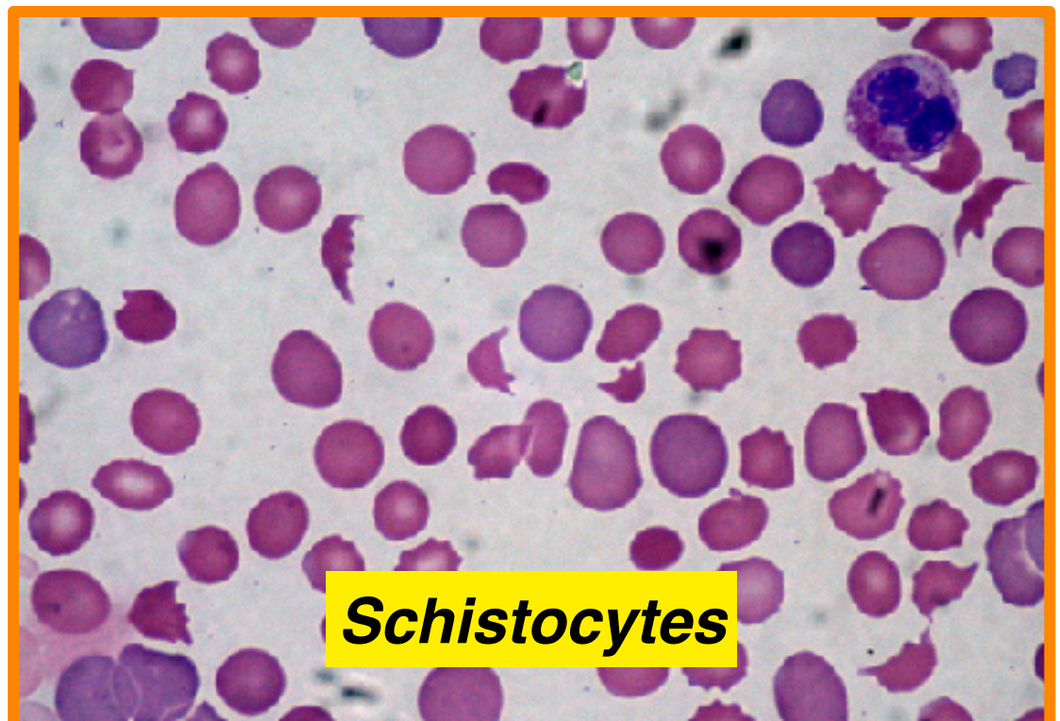




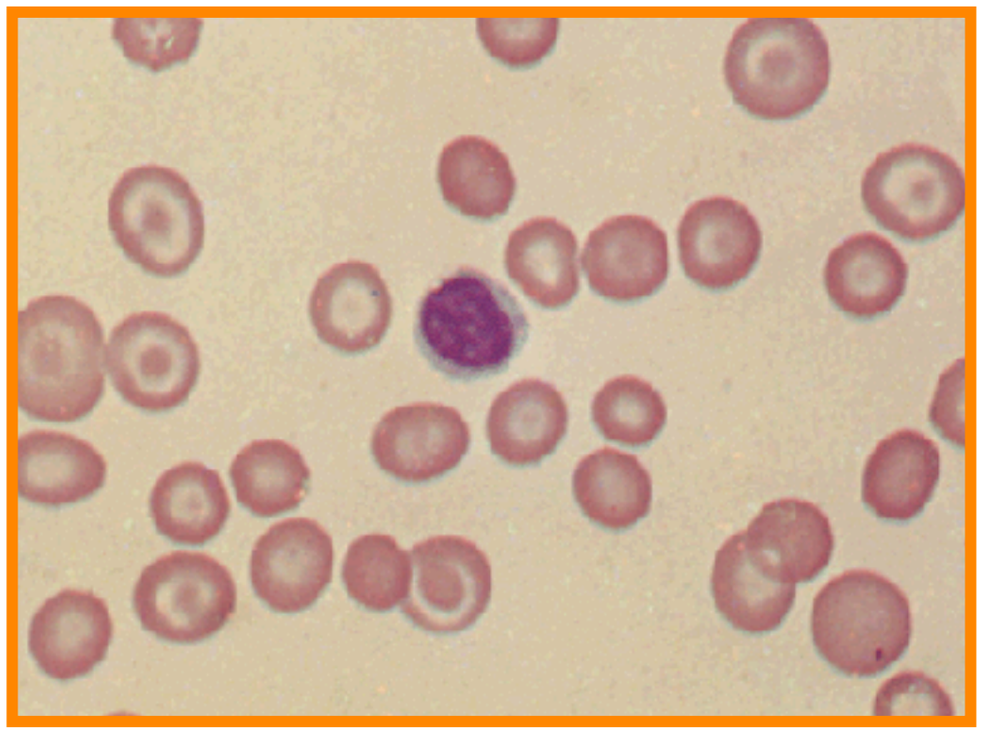
Peripheral blood smear





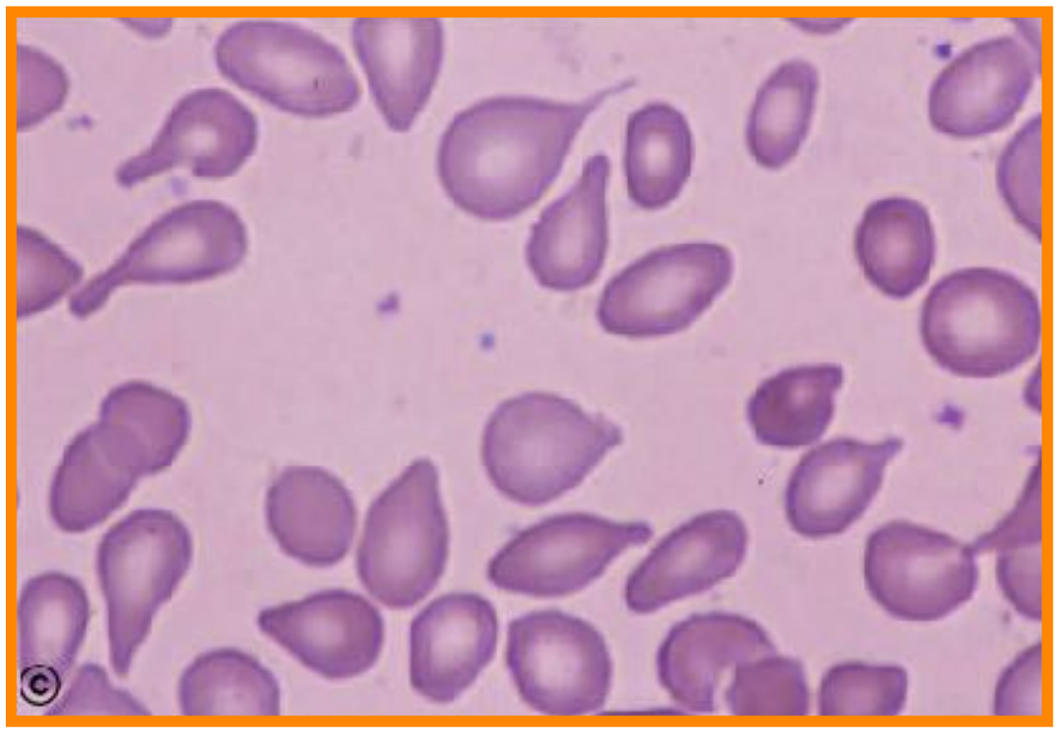


Peripheral blood smear



Target cell

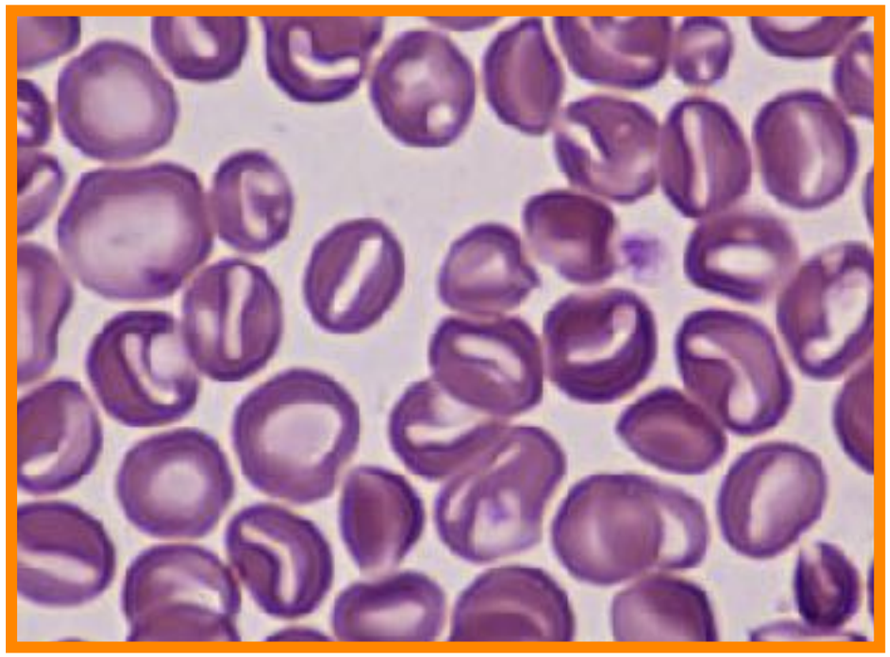
1. *Liver Disease*
2. *Thalassemia*
3. *Hb E*
4. *Post splenectomy*



Tear drop cell

1. *Myelofibosis*
2. *Infiltration of BM*
3. *Tumors of BM*
4. *Thalassemia*

Peripheral blood smear



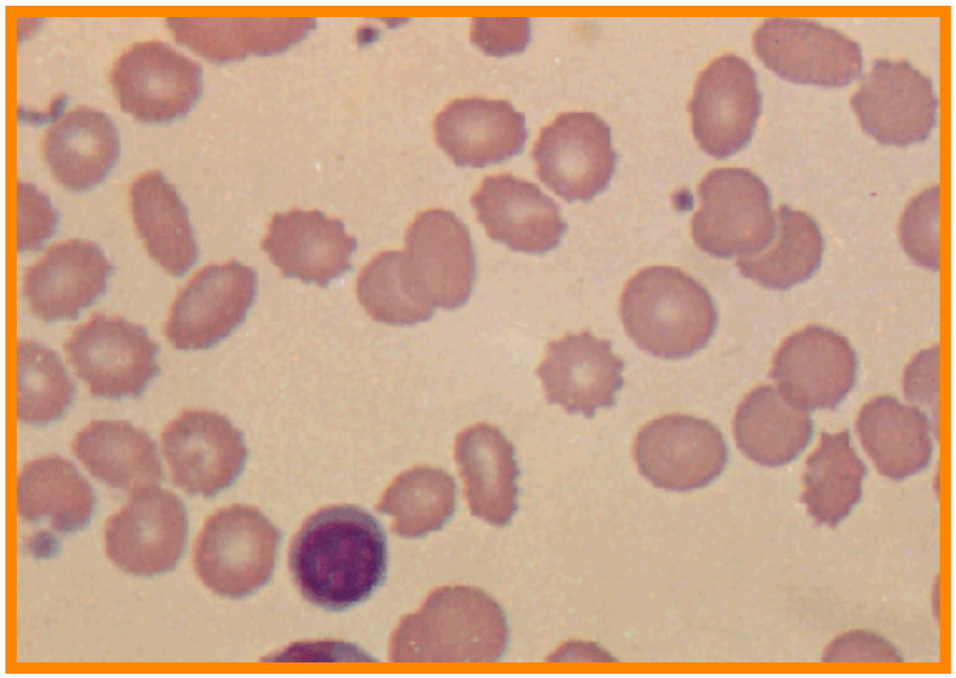
Stromatocytes

1. Liver Disease

2. Acute Alcoholism

3. H Stomatocyosis

4. Malignancies



Echinocytes

1. Uremia

2. Peptic ulcer

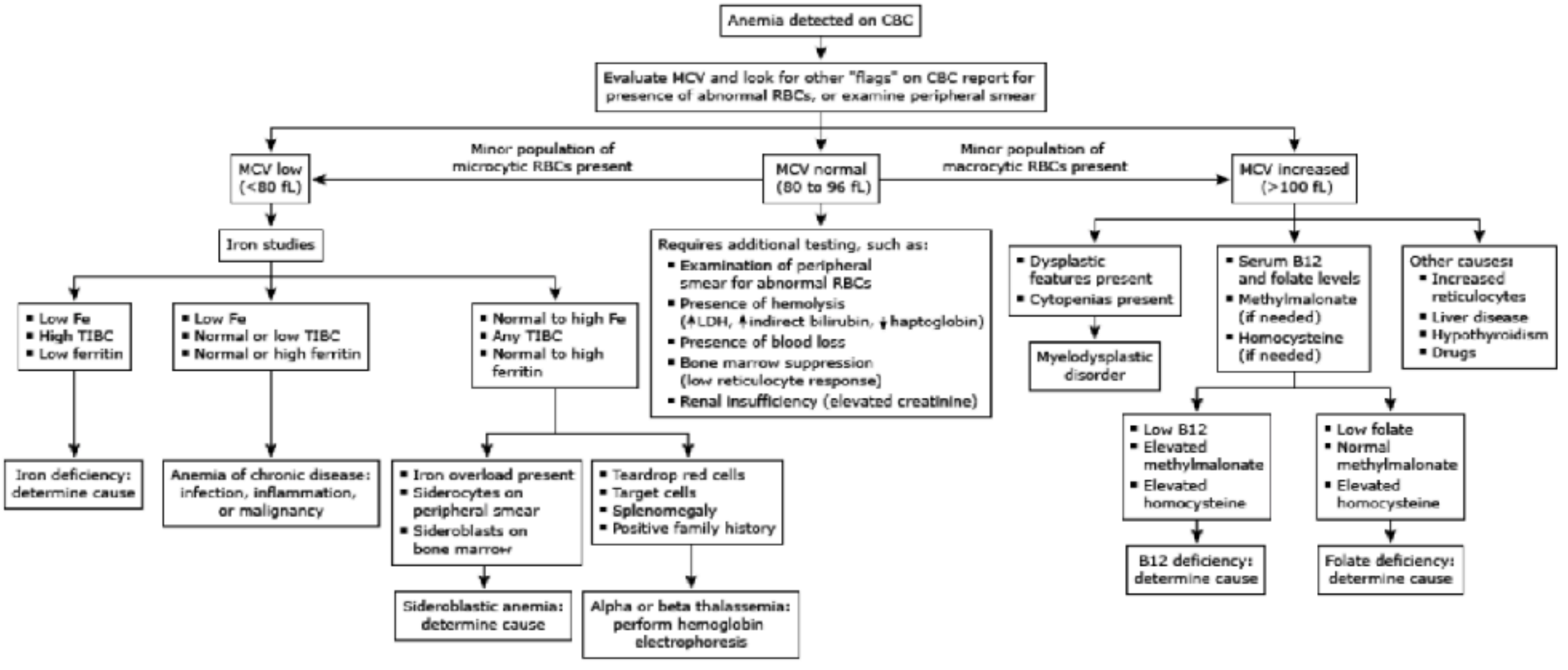
3. Gastric Ca

4. PK-D

Called Burr Cells

Approach to anemia

Evaluation of anemia in the adult according to the mean corpuscular volume



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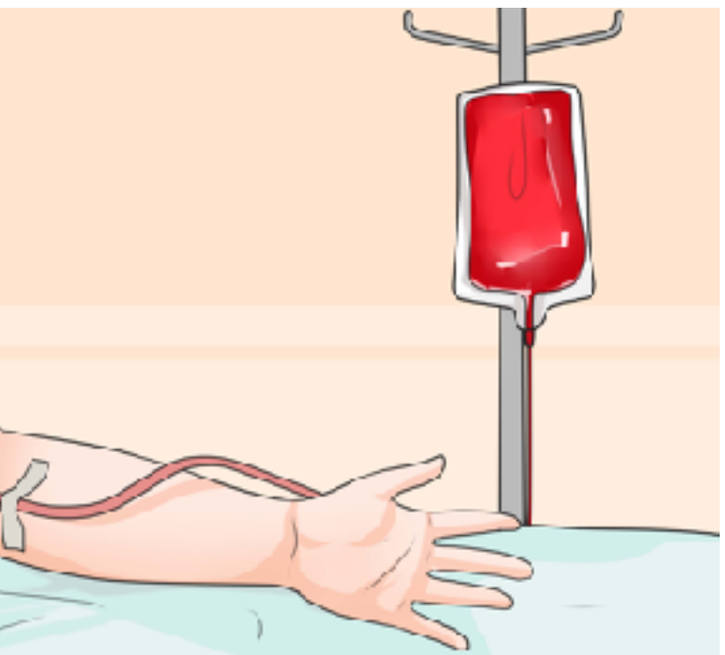

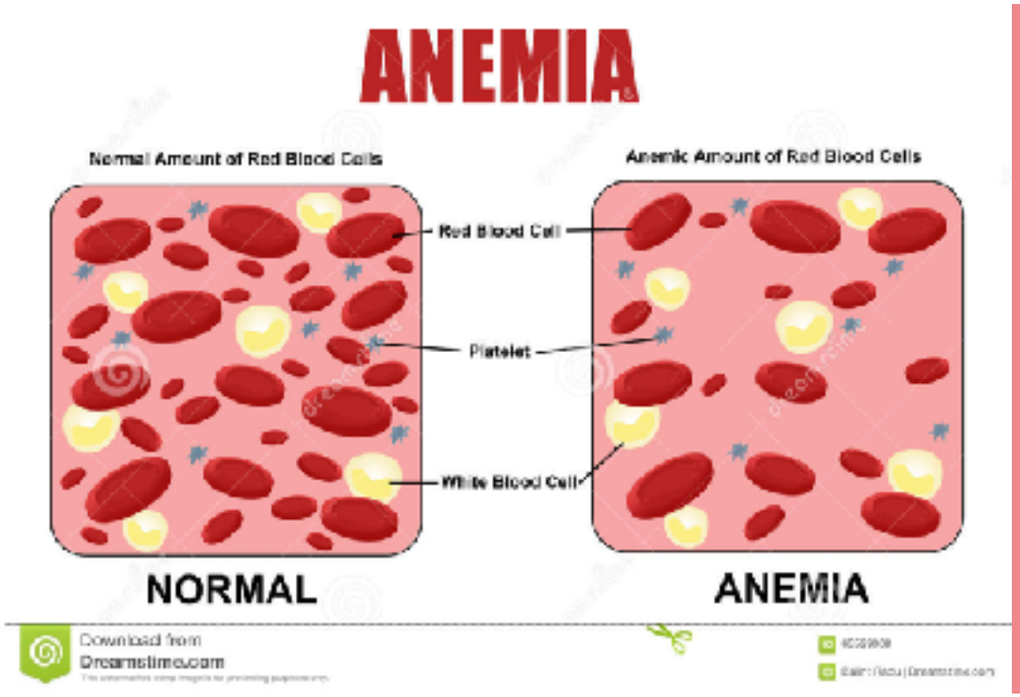

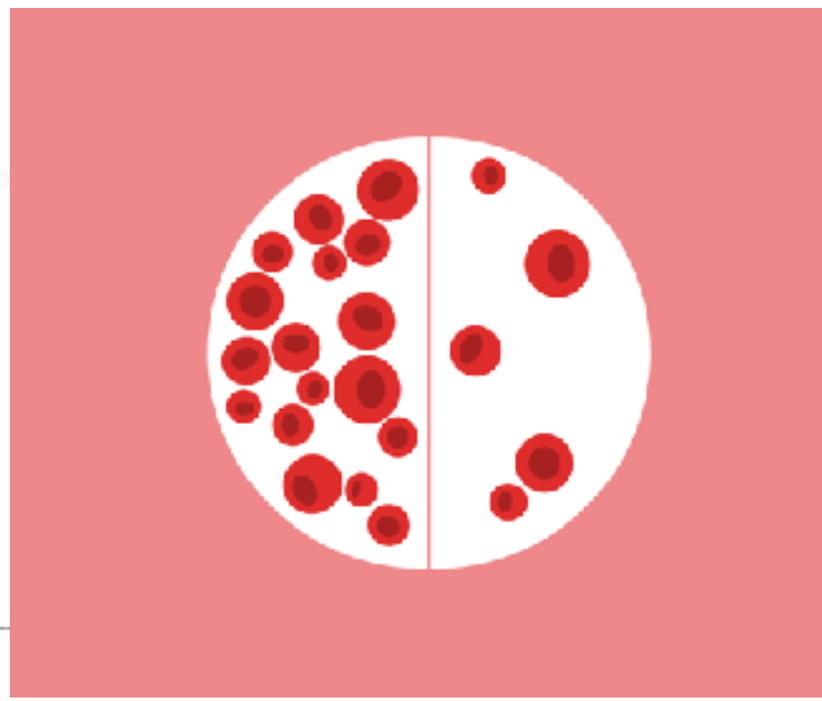

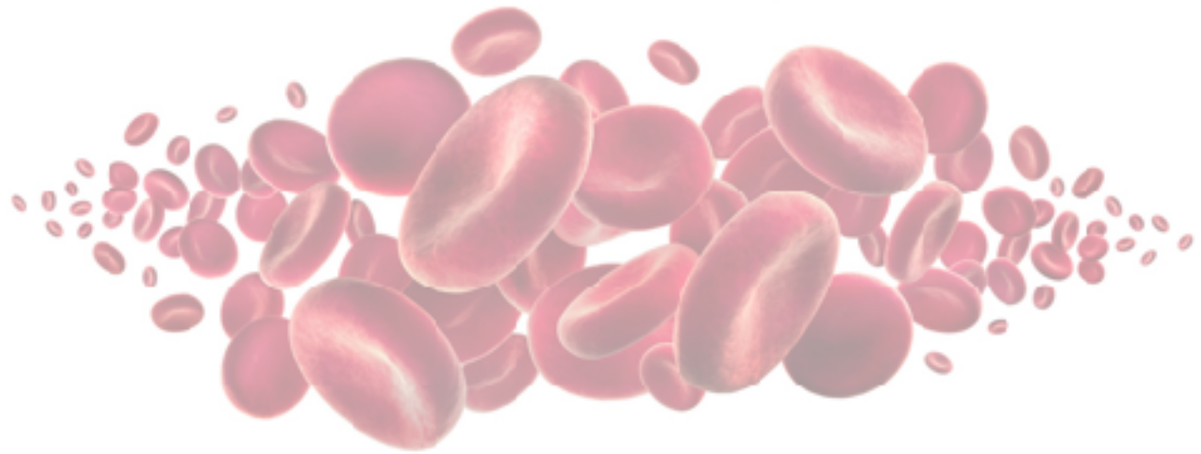

What you should know about “CBC”

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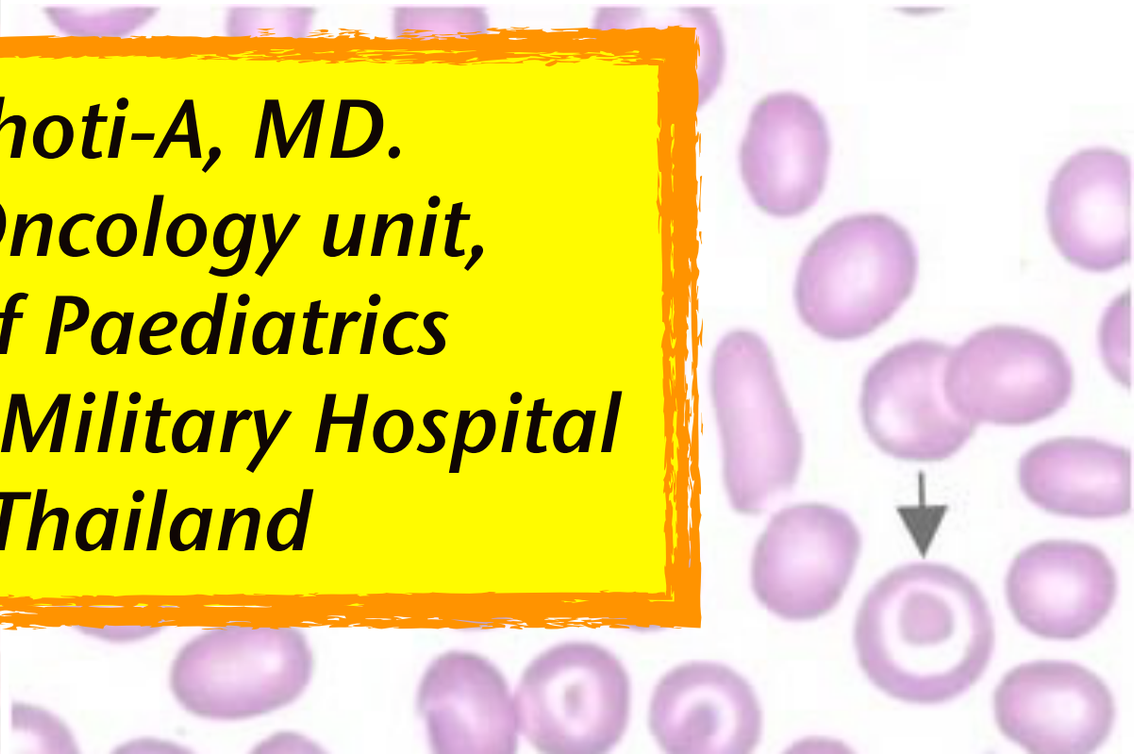
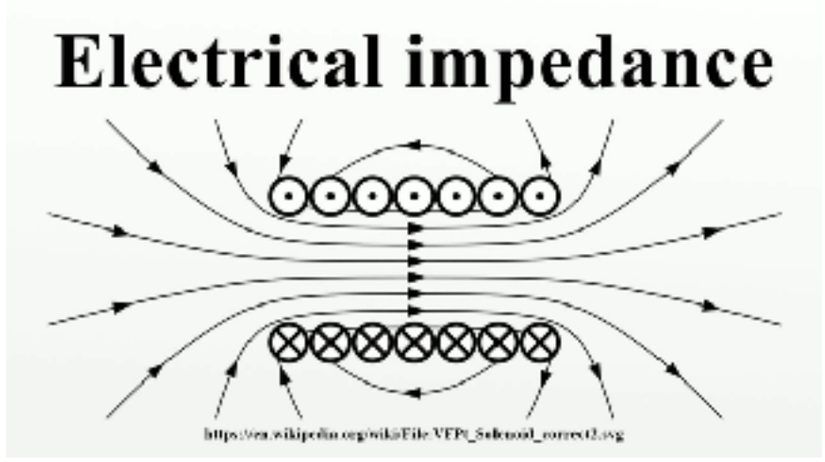


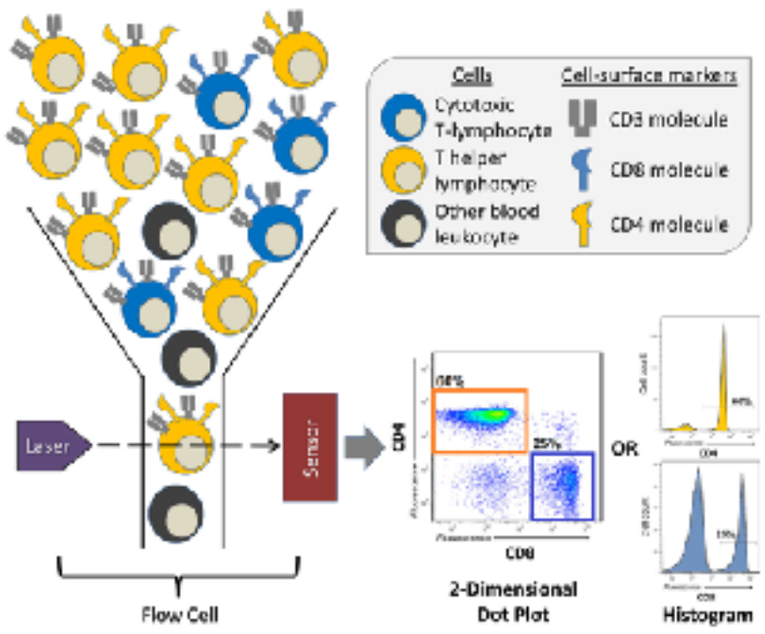
Principle of automated CBC

Electrical impedance and conductivity

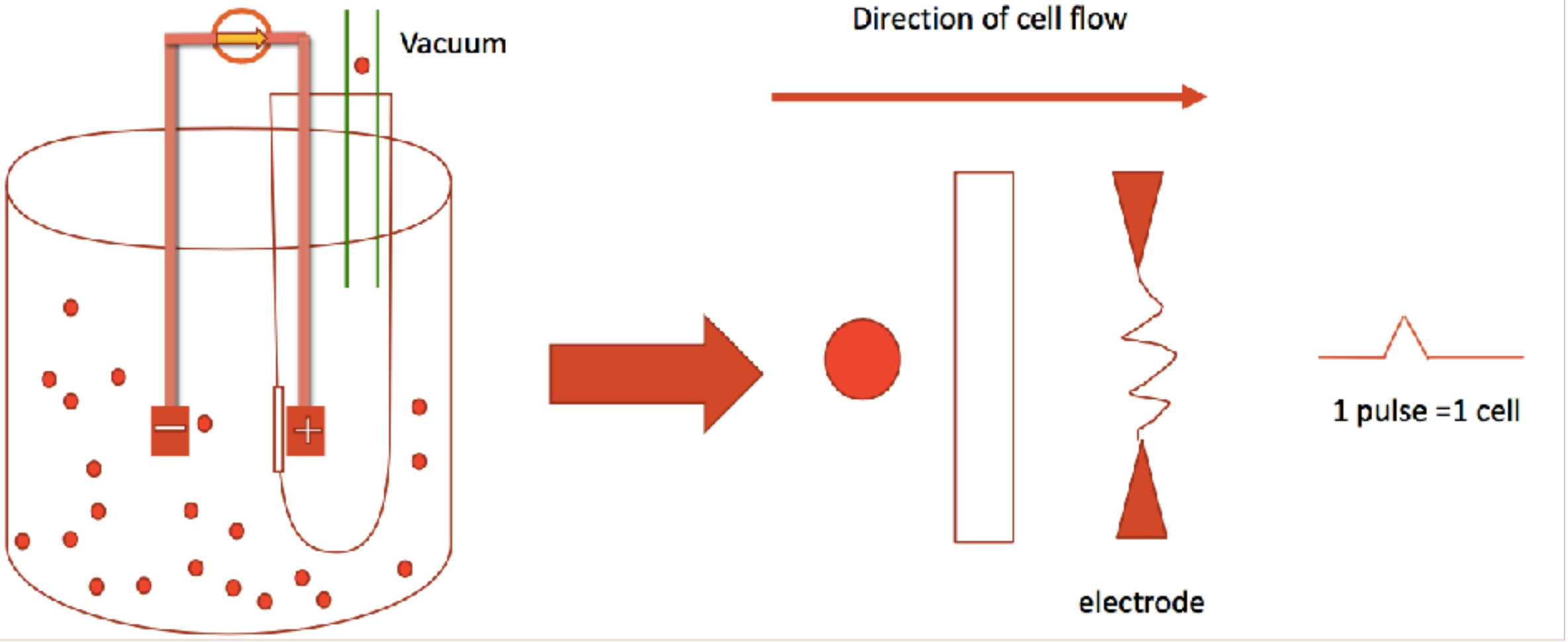
Flow cytometry

- Fluorescence
- Light scatter at various angle
- Light absorption of cell stained in flow





Electrical impedance and conductivity



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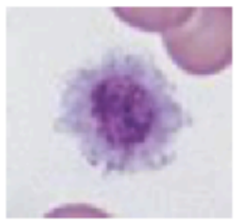
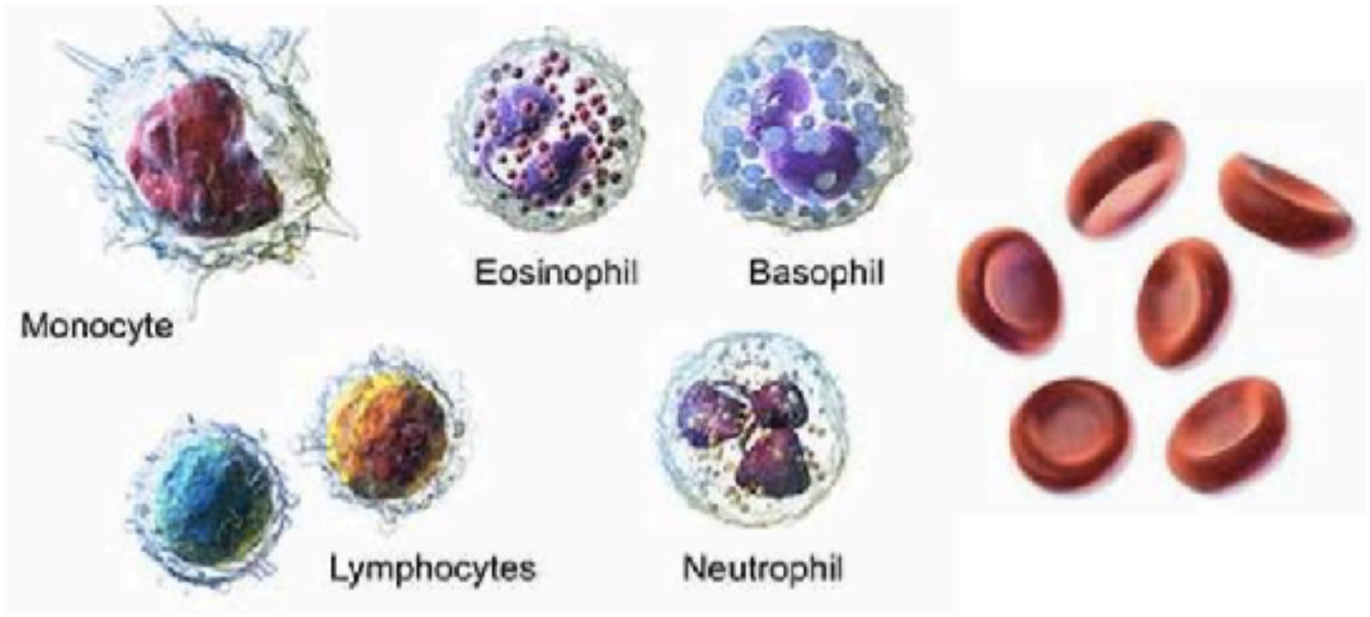


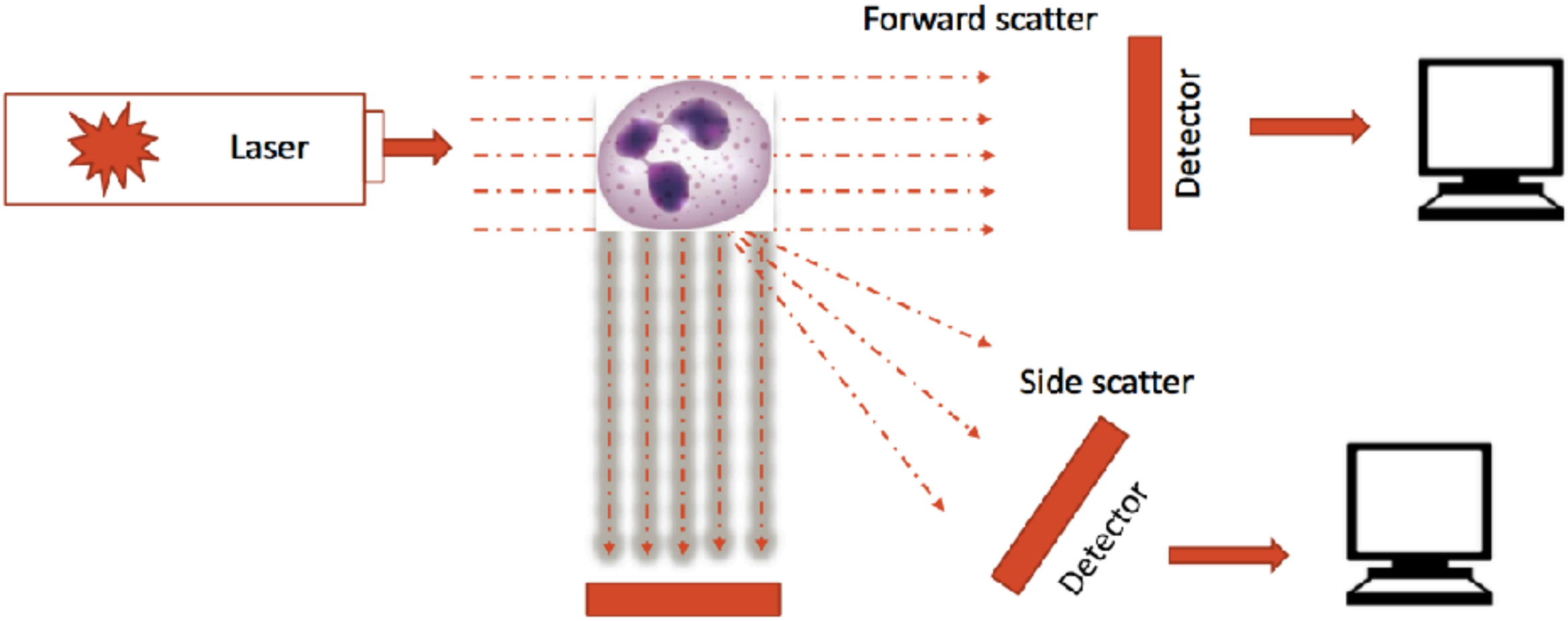
2-20 fL



36-360 fL

Separated by Volume

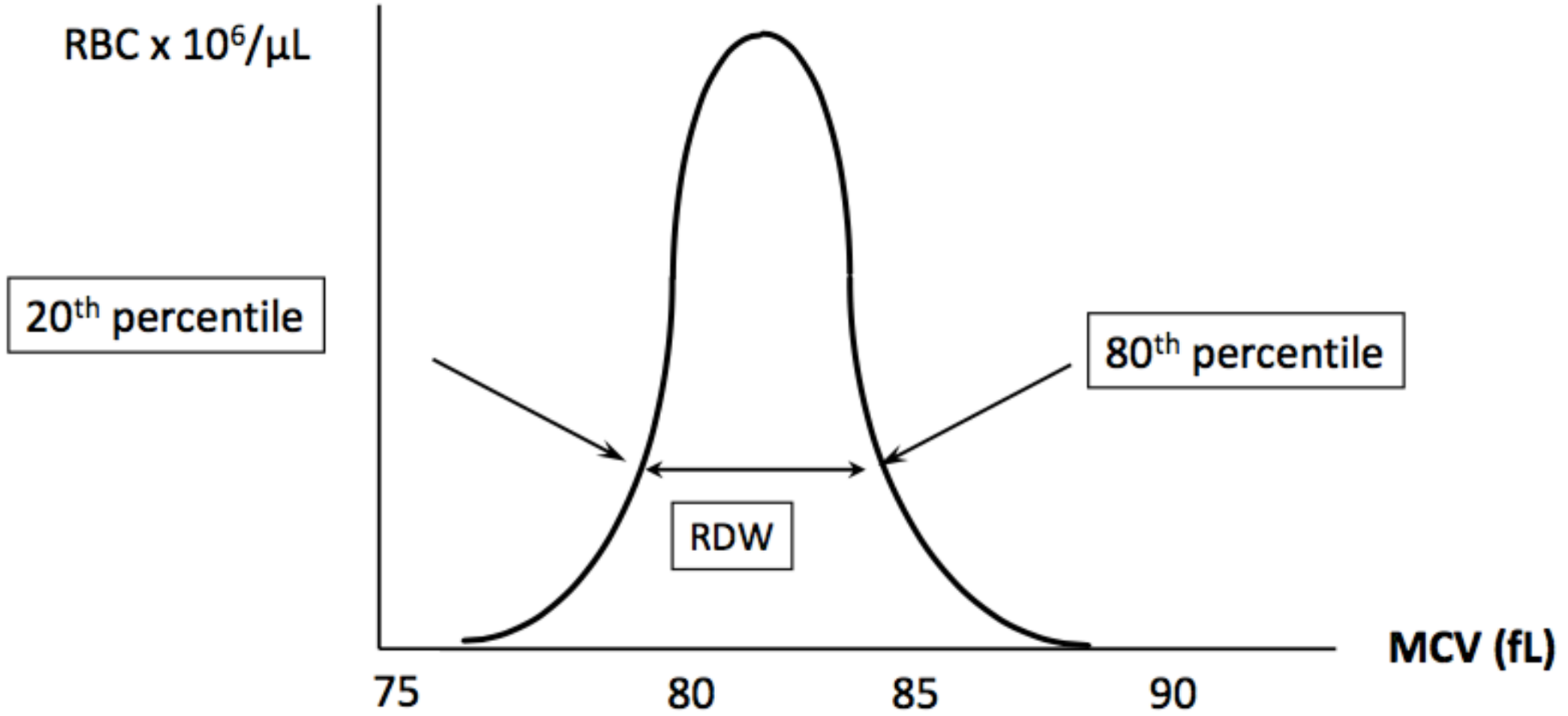
Flow cytometry



RBC Parameters

	Parameter	Unit of reporting	Method of determination
Direct measurement	RBC count	x $10^6/\mu L$	
	Hb	g/dL	
	MCV	fL	
Indirect measurement	Hct	%	$\frac{RBC \times MCV}{10}$
	MCH	pg	$\frac{Hb \times 10}{RBC}$
	MCHC	g/dL	$\frac{Hb}{Hct \times 100}$
	RDW	%	

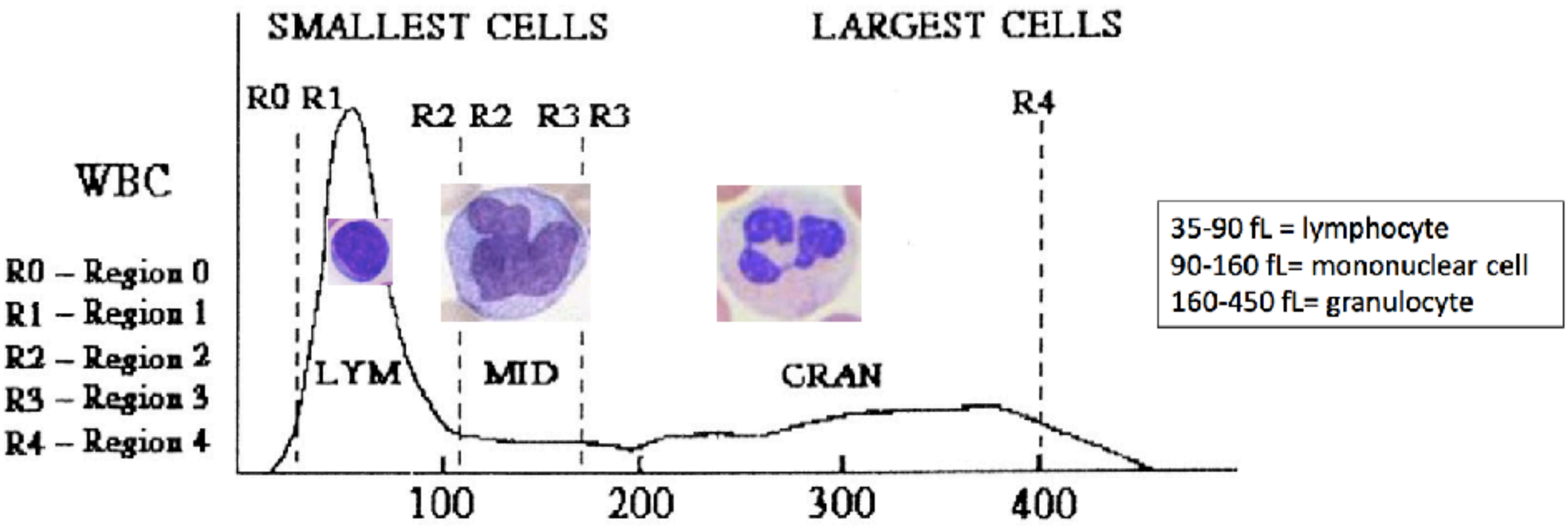
RBC Distribution Width



$$RDW = \frac{80^{th}\ \text{percentile} - 20^{th}\ \text{percentile}}{80^{th}\ \text{percentile} + 20^{th}\ \text{percentile}} \times \text{constant}$$

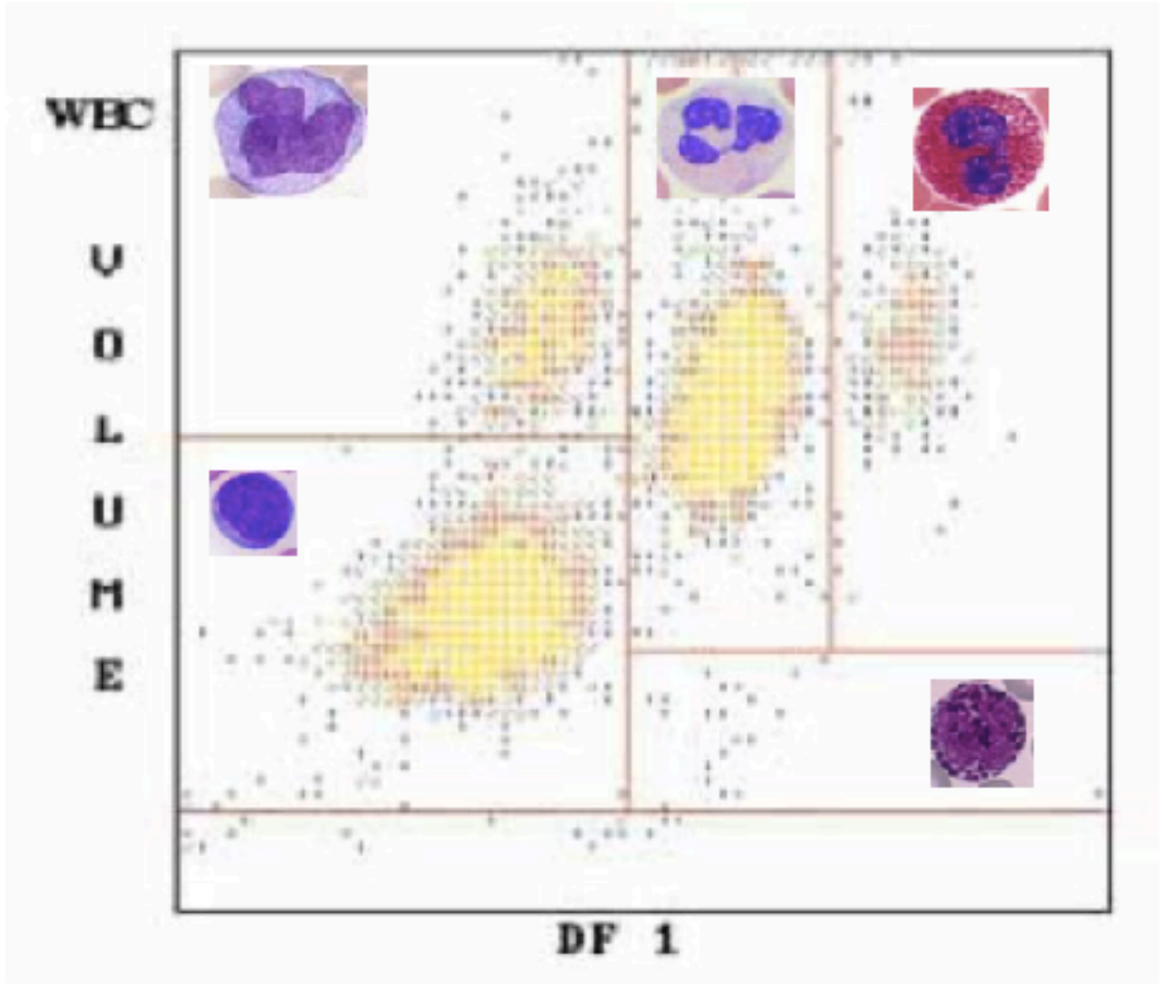
WBC Count

Three part differential



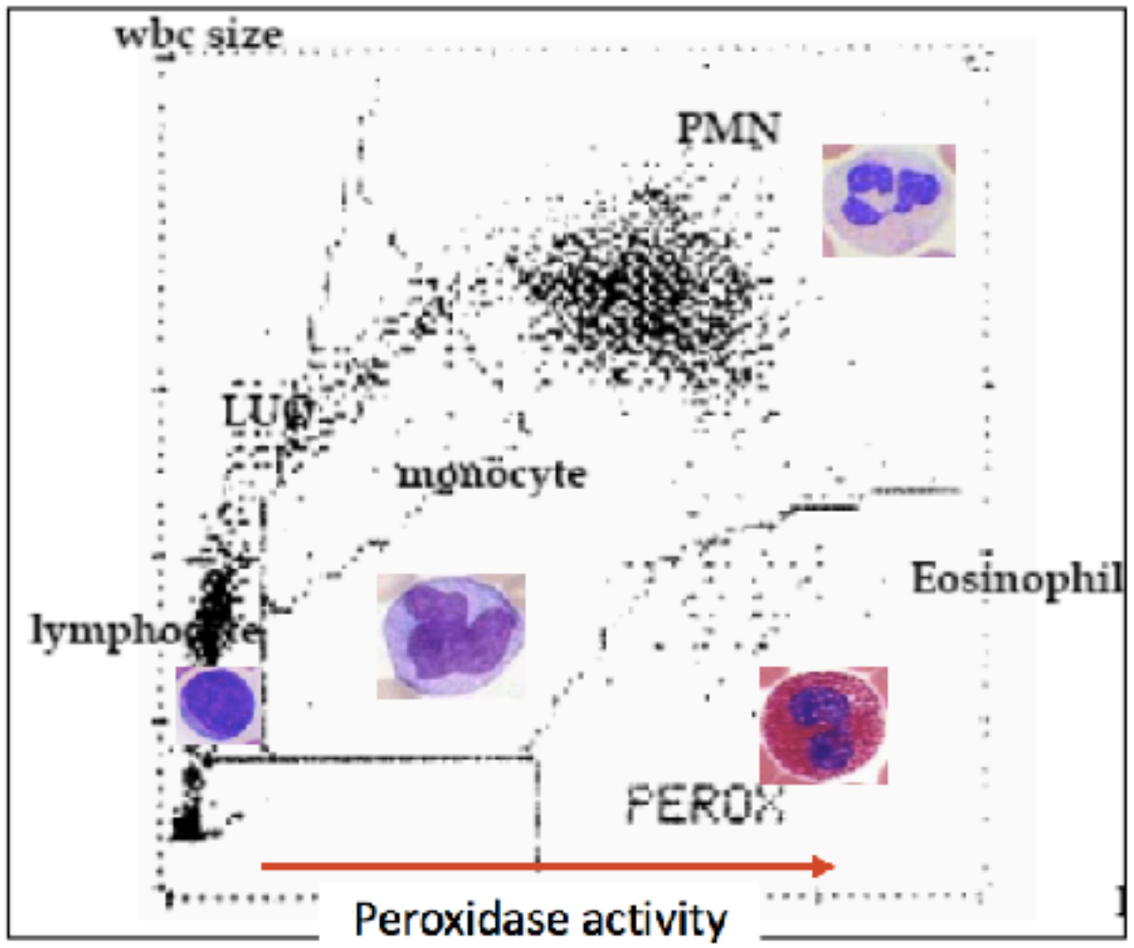
WBC Count

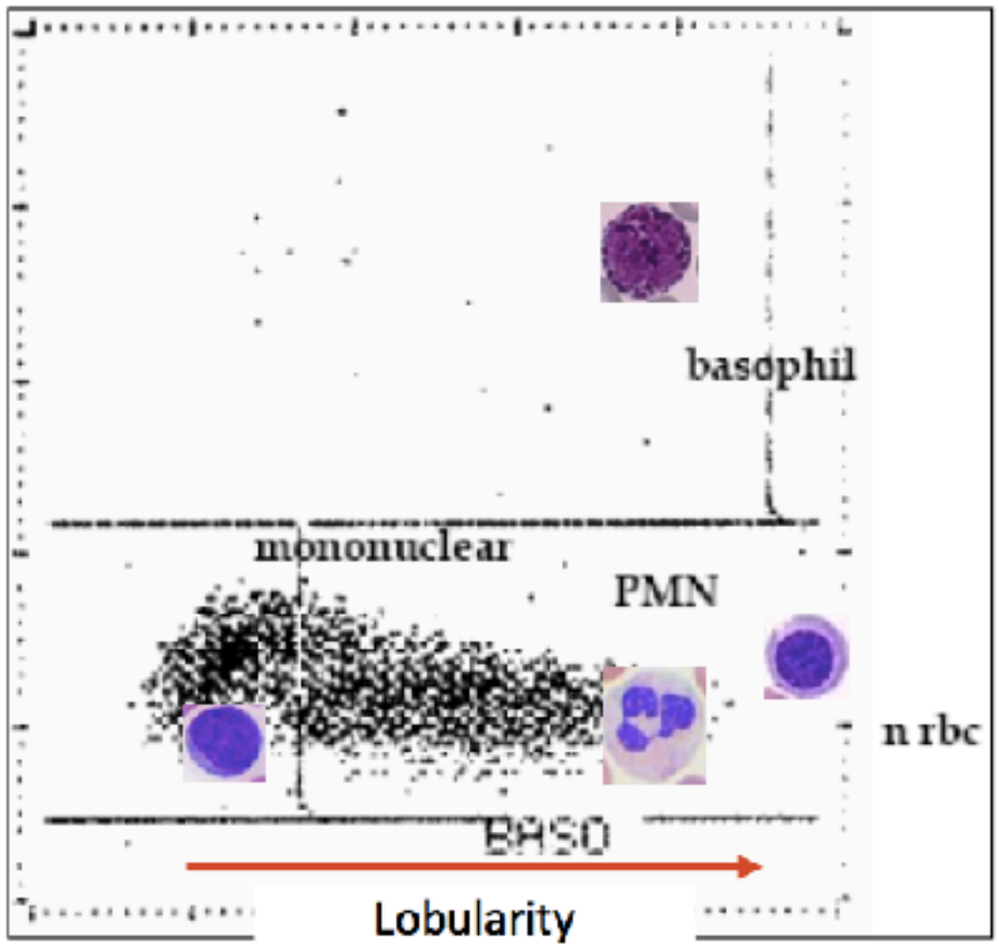
Five part differential



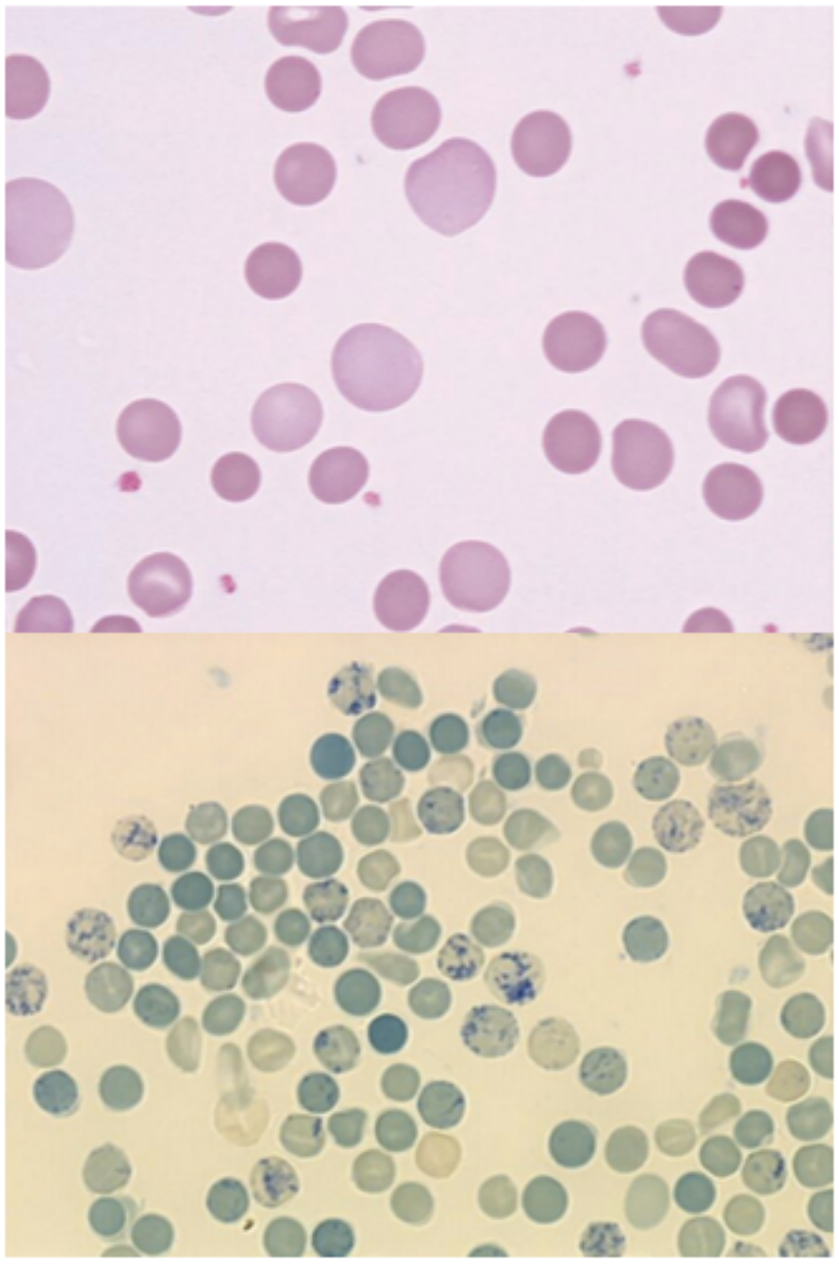
WBC Count

Six part differential





Reticulocyte count



- %Corrected (RetiC) = $\frac{\%\ \text{RetiC x patient's Hct}}{45}$
- Reticulocyte production index = $\frac{\%\text{corrected RetiC}}{\text{maturation time}}$

maturation time (days) =	(Hct ≥ 40%),
1.5	(Hct 30-40%)
2	(Hct = 20-30%)
2.5	(Hct < 20%)

Absolute reticulocyte count= %reticulocyte x RBC

- >100,000 → Appropriate bone marrow response
- < 50,000→Inappropriate bone marrow response
- 50,000-100,000→combined etiologies

Normal Value

- Hb
  - 13-18 g/dl men
  - 12-16 g/dl women
- Hct
  - 40-52 % men
  - 35-47 % women
- RBC count
  - 4.4-5.9 x10^{12}/L men
  - 3.8-5.2 x10^{12}/L women
- RDW 11.5-14.5
- MCV 80-100 fl
- MCH >27 pg
- MCHC 32-36 g/dl
- WBC
  - 3.8-10.6 x10^{9}/L men
  - 3.6-11.0 x10^{9}/L women
- PLT 150 – 440 x10^{6} /L

Special condition on CBC

Component	Disorder/Condition	Effect on cell count
Red cells	Microcytosis/schistocyte	Under estimated RBC
	Howell-Jolly bodies	Elevate platelet count
	Polycythemia	Under estimated RBC
White cells	Leukocytosis	Overestimated RBC
	Acute leukemia chronic lymphocytic leukemia viral infection	Spurious low WBC
	Chemotherapy of acute leukemia	May artificially increase platelet count
Platelets	Platelet agglutinins	Underestimated platelet count sometimes with spurious increase in WBC
Plasma	Cold agglutinins	May underestimate RBC with spurious macrocytosis
	Cryoglobulins	Variation in platelet count

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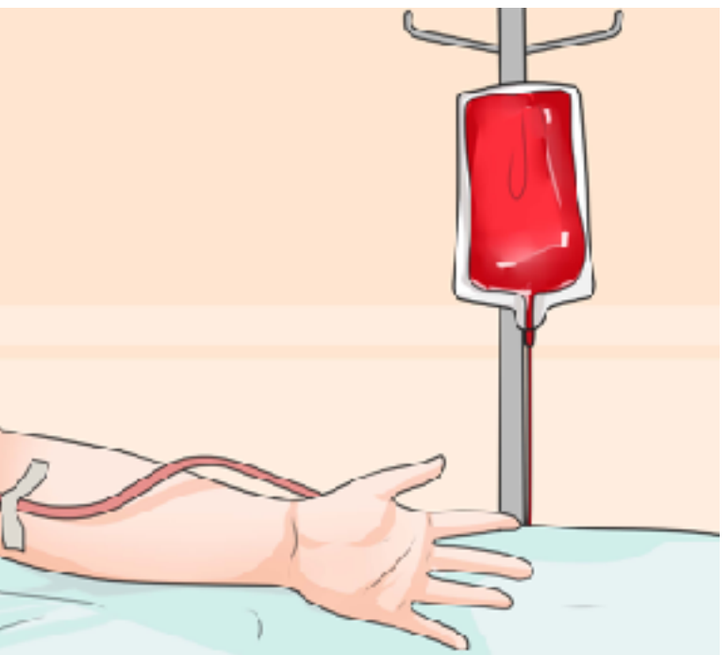

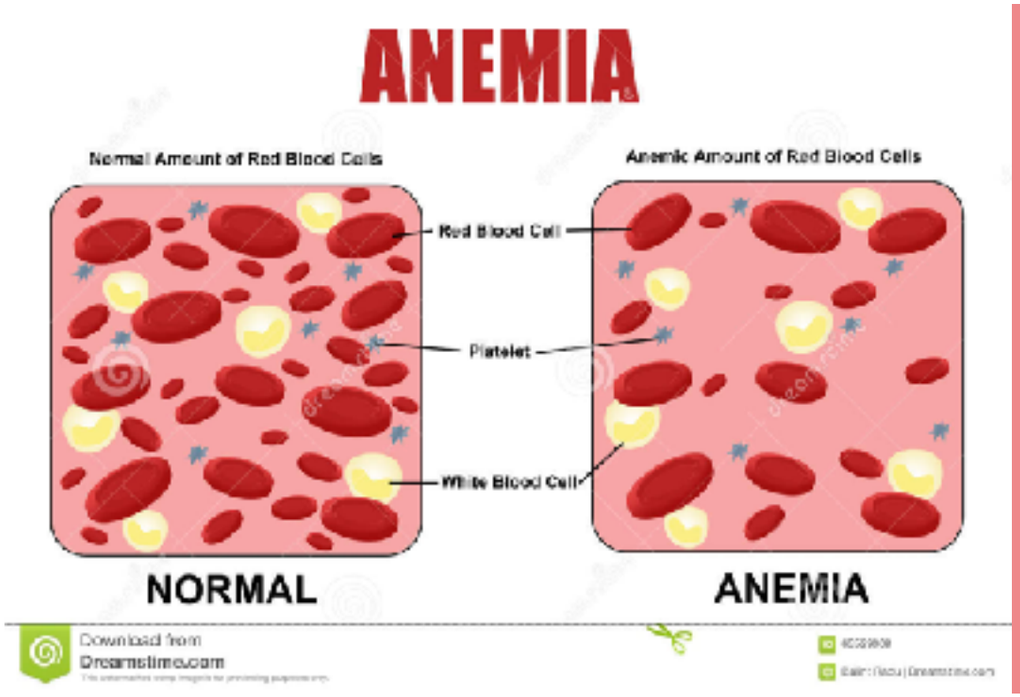

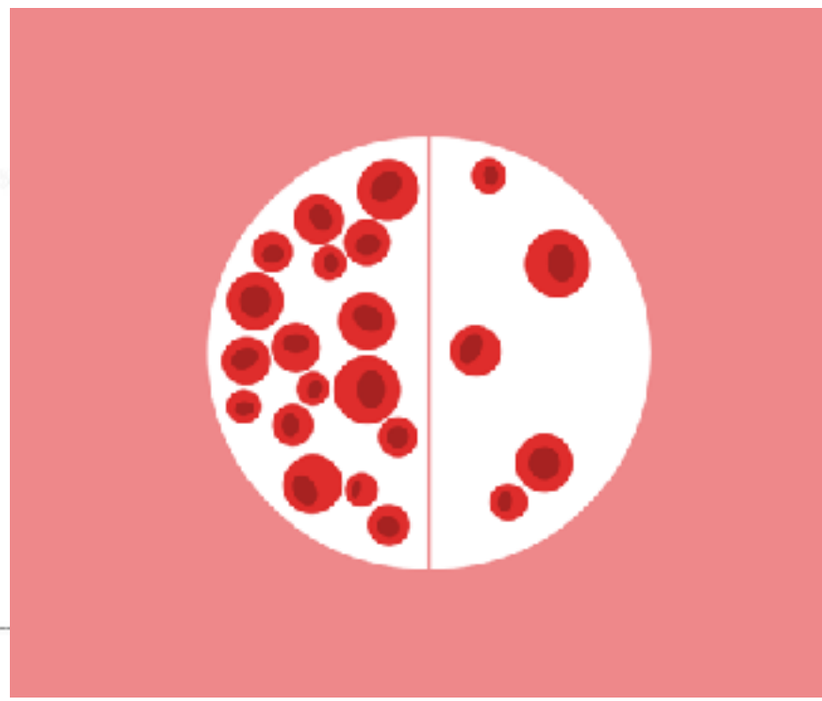

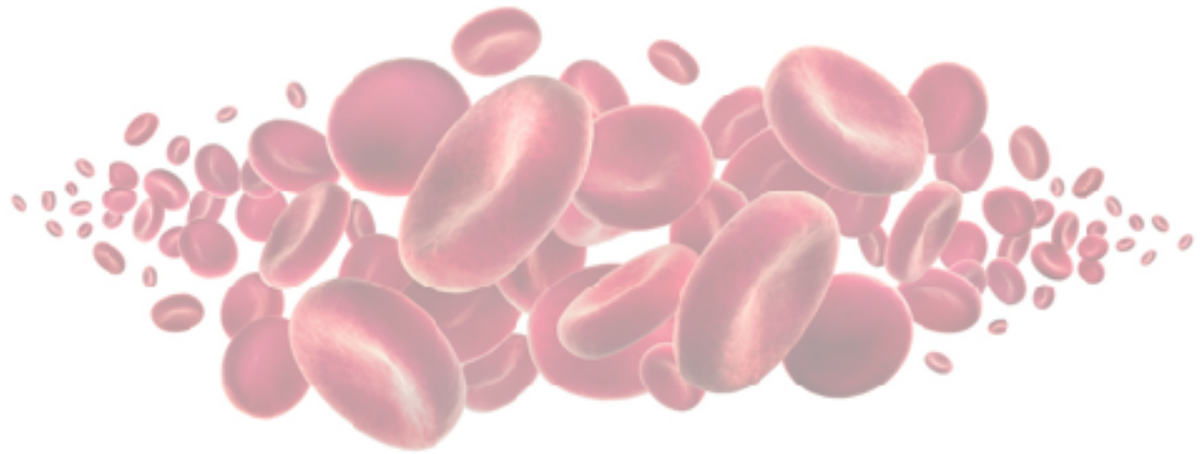

Hemoglobin Typing & Thalassemia

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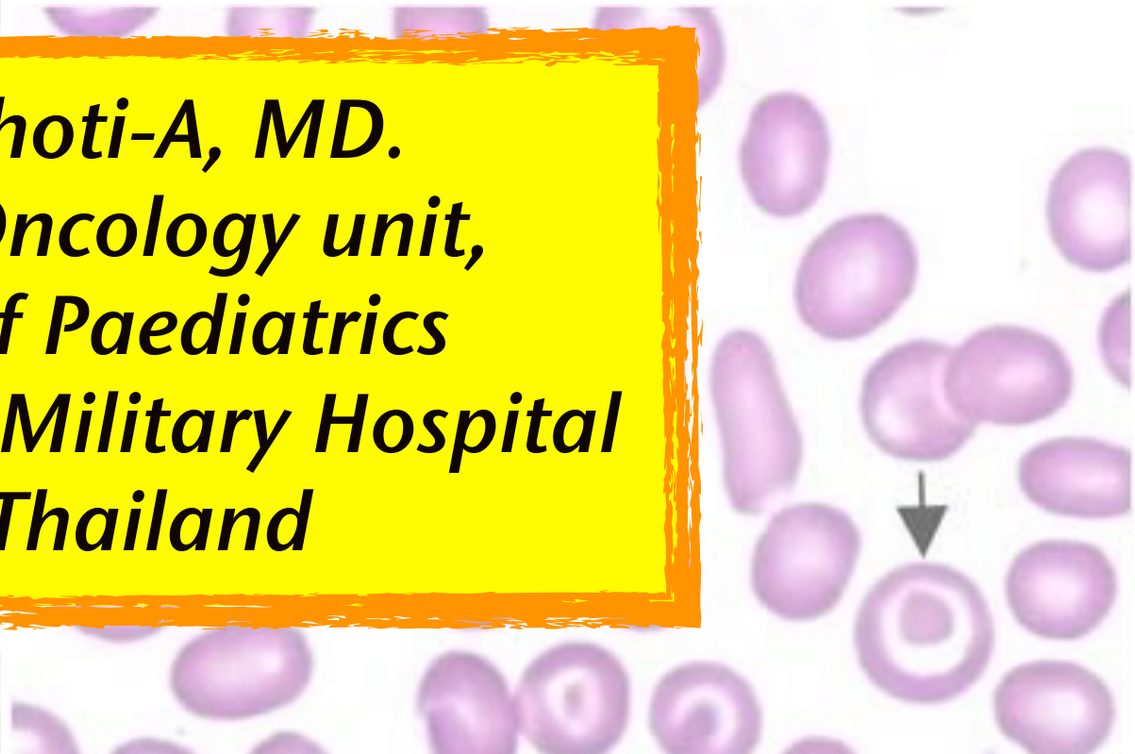
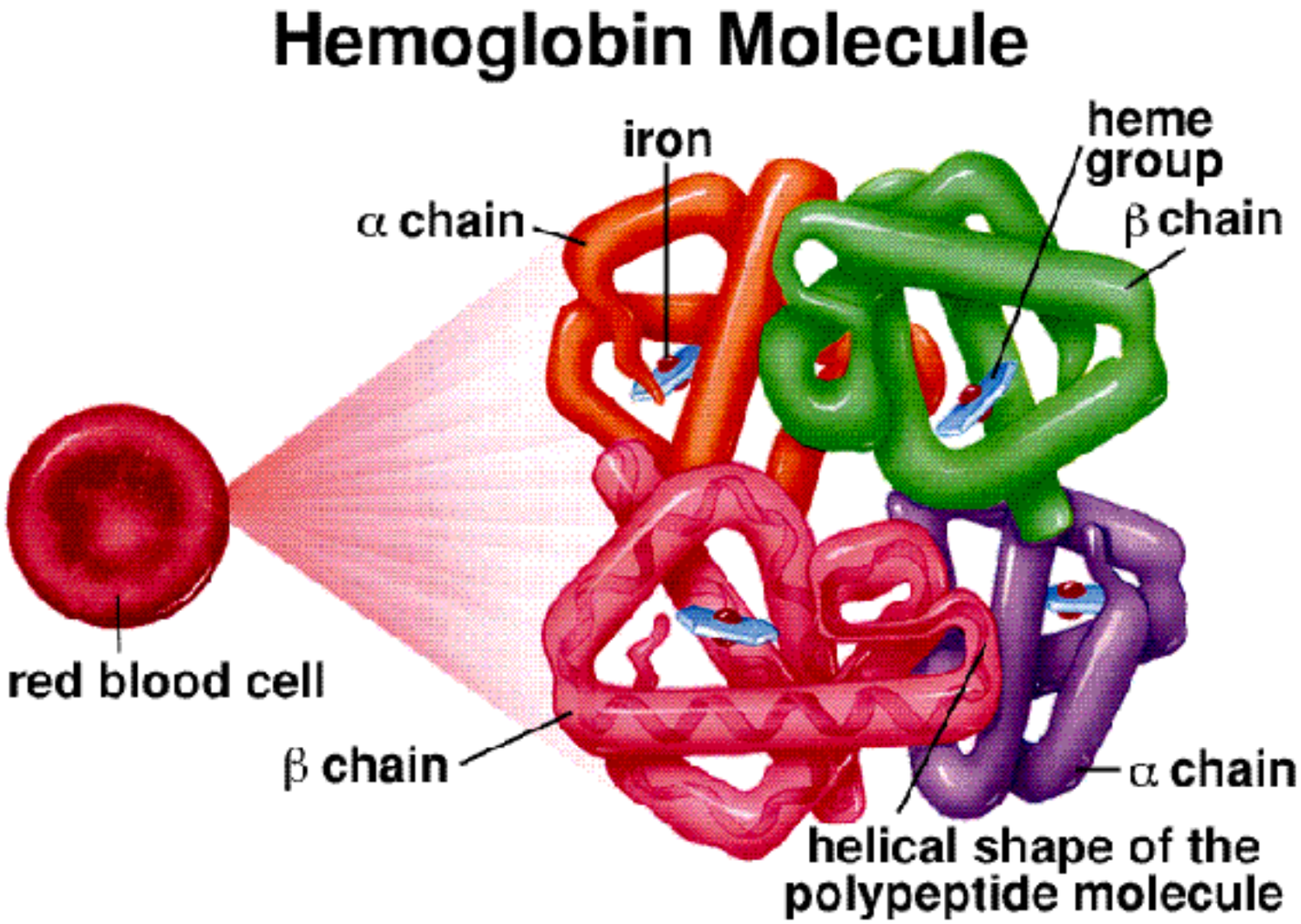


Structure of Hemoglobin



Hb consists of 4 subunits

- 2 α-chain/ α-like chain (ζ(Zeta)-globin chain)

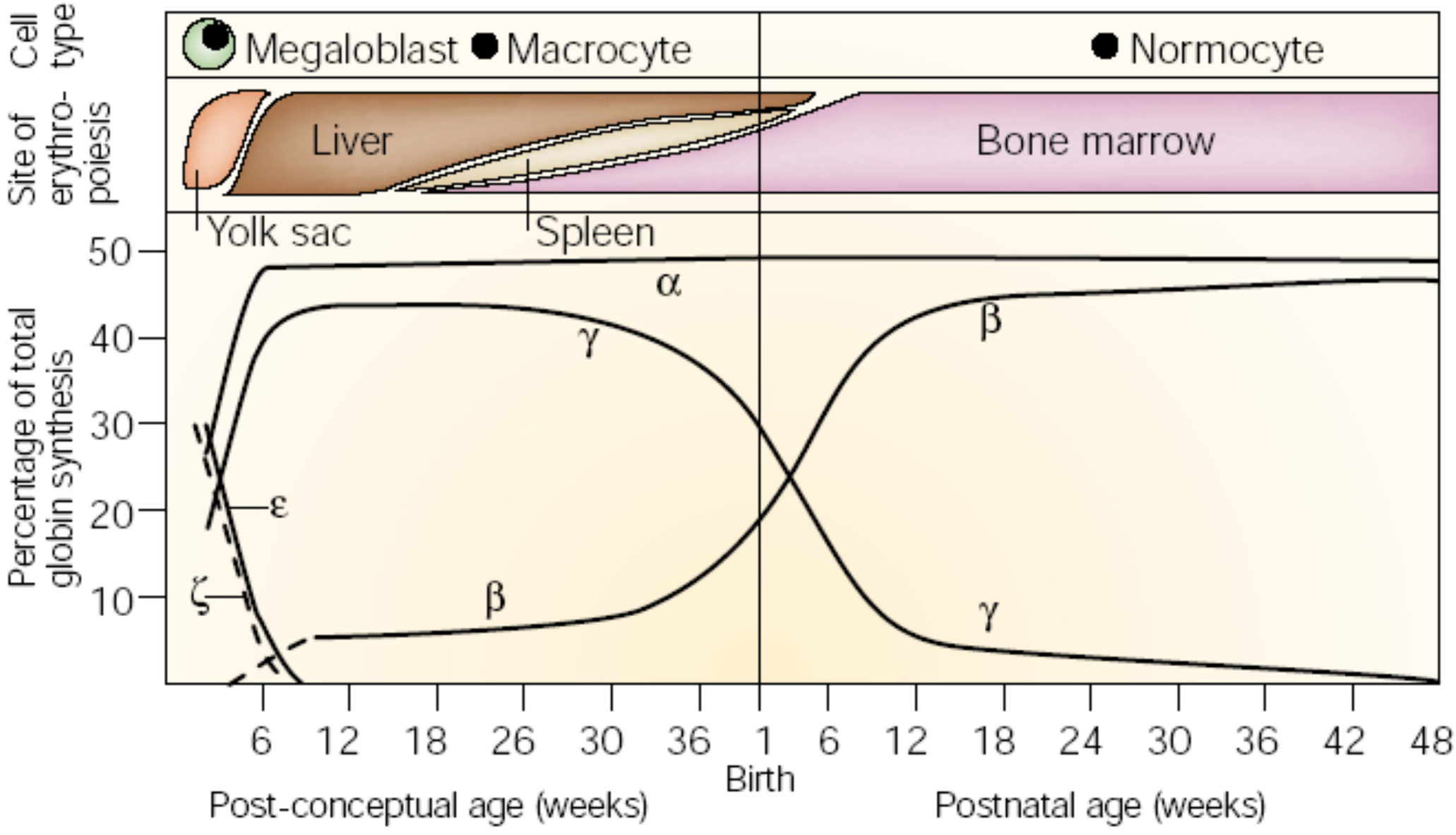
located on ***Chromosome 16***

- 2 ß-chain/ ß-like chain (ɣ(Gamma), δ(Delta), ε(Epsilon)-globin chain)

located on ***Chromosome 11***

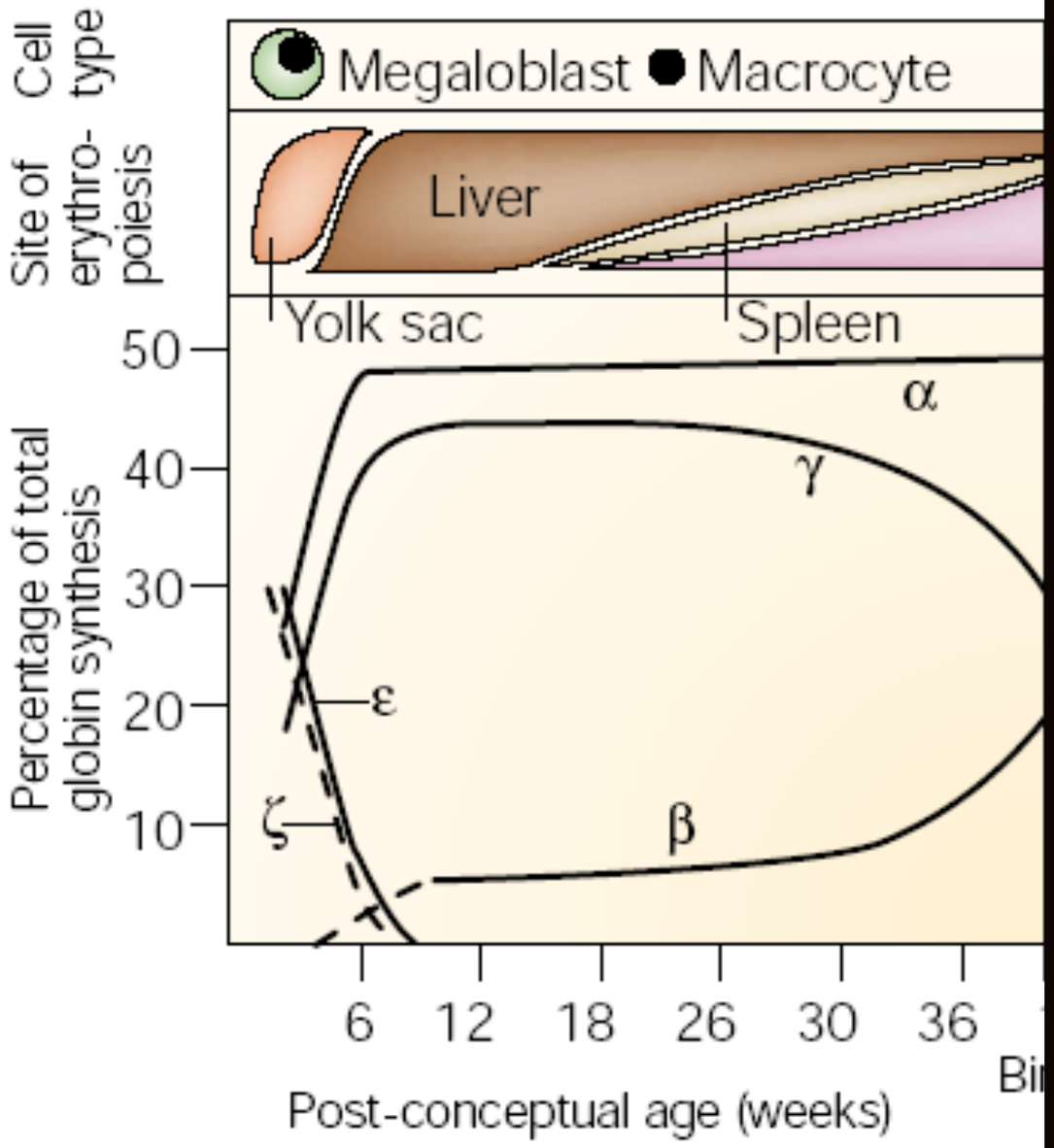
The Inquiry into life, The McGraw-Hill, 8th Edition, 1997

Globin gene synthesis



Lessons from the thalasemia. Nature Reviews, Genetics, volume2, 2001

Globin gene synthesis



Embryonic Hemoglobin

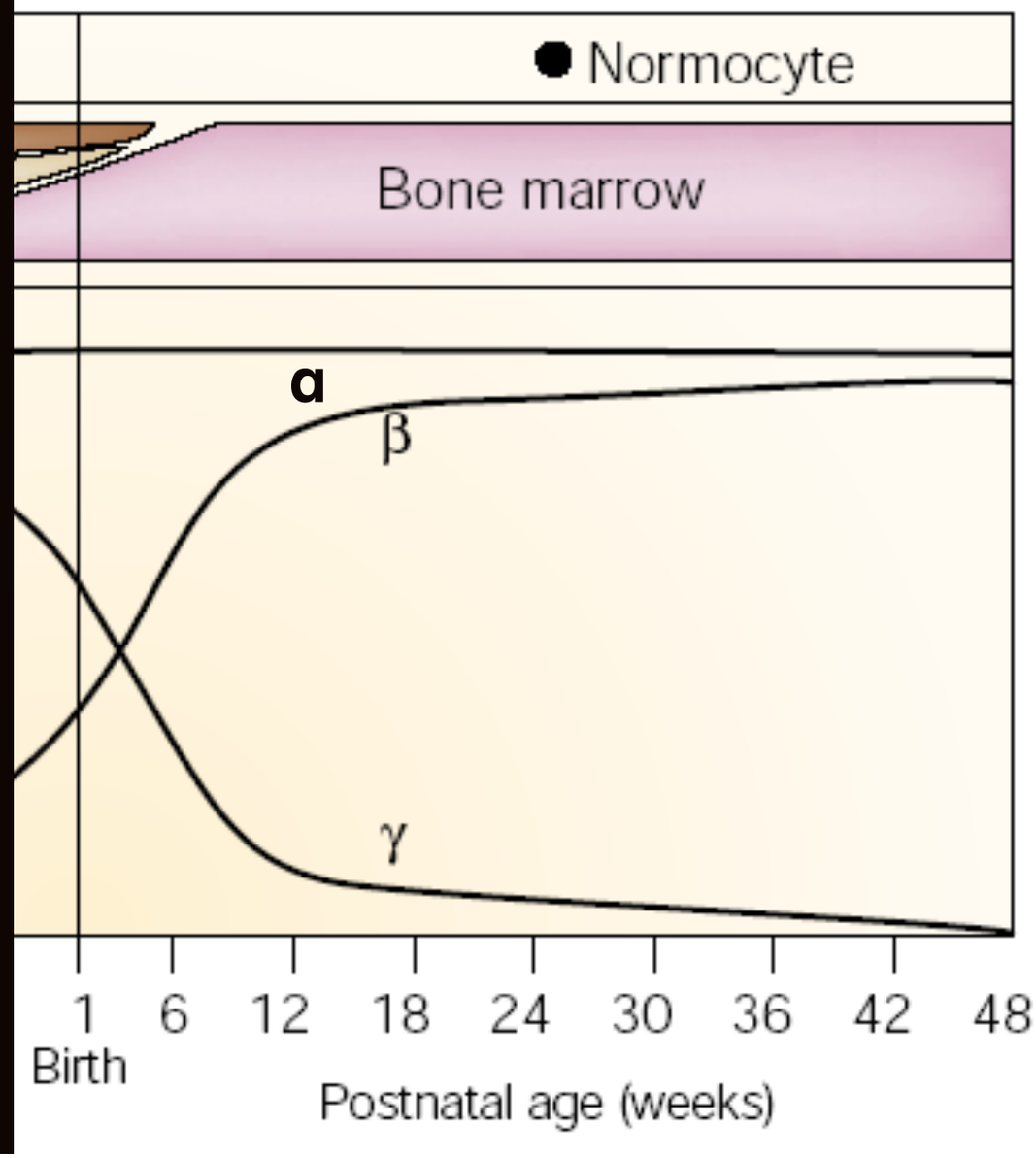
- ***Hb Gower I*** : ζ2 ε2
- ***Hb Gower II*** : α2 ε2
- ***Hb Portland*** : ζ2 ɣ2

***Lessons from the thalasemia. Nature Reviews, Genetics, volume2, 2001**

Globin gene synthesis

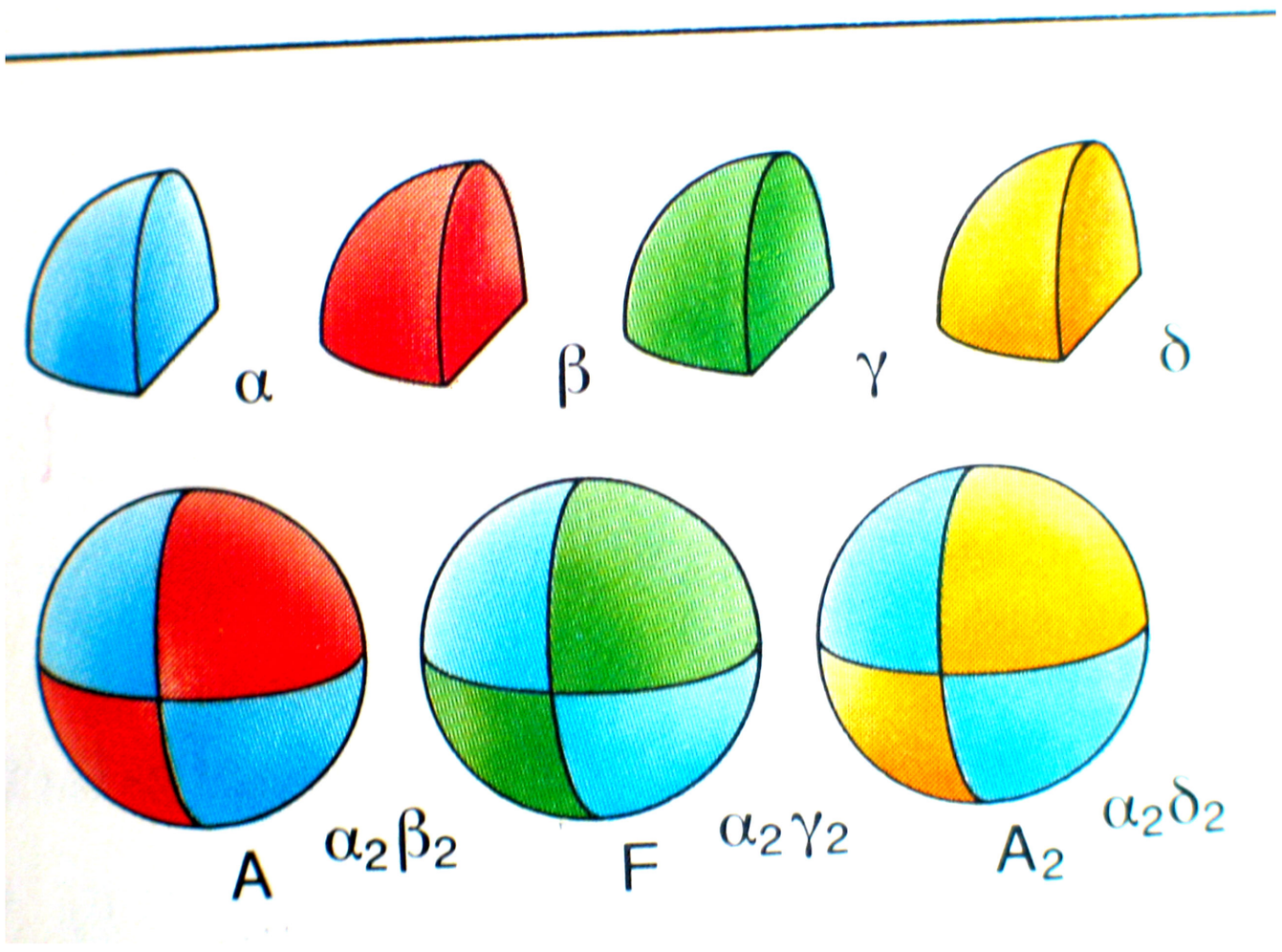
Fetal Hemoglobin

- ***Hb F*** : α2 ɣ2
- ***Hb A*** : α2 β2
- ***Hb A2*** : α2 δ2



**Lessons from the thalasemia. Nature Reviews, Genetics, volume2, 2001*

Normal Adult Hemoglobin



HbA **96-98%**

- HbA : α2 β2

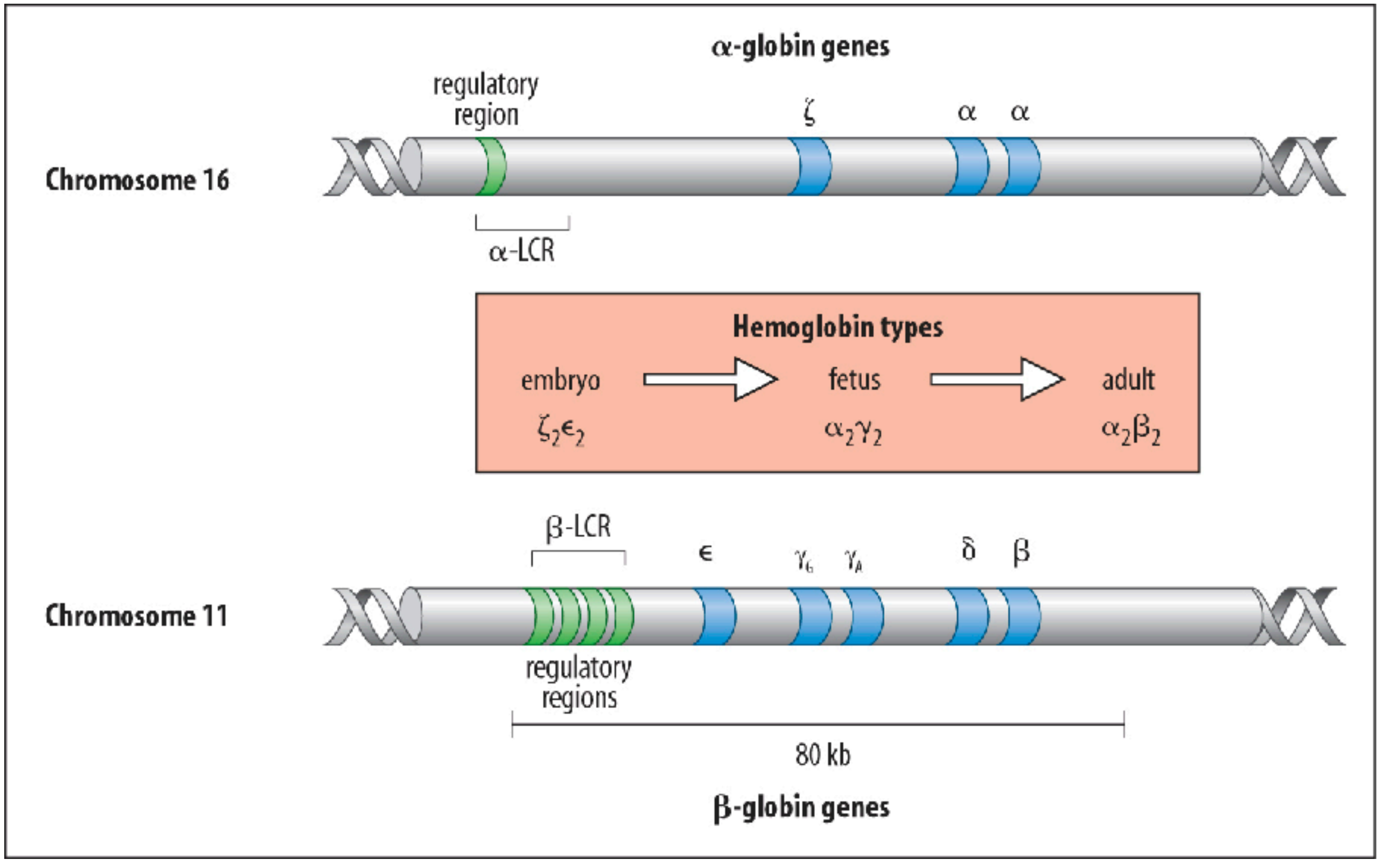
HbA2 **1-3%**

- HbA2 : α2 δ2

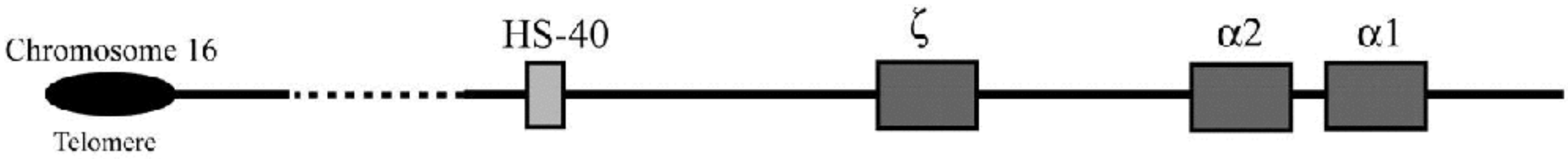
HbF **0.5-1%**

- HbF : α2 ɣ2

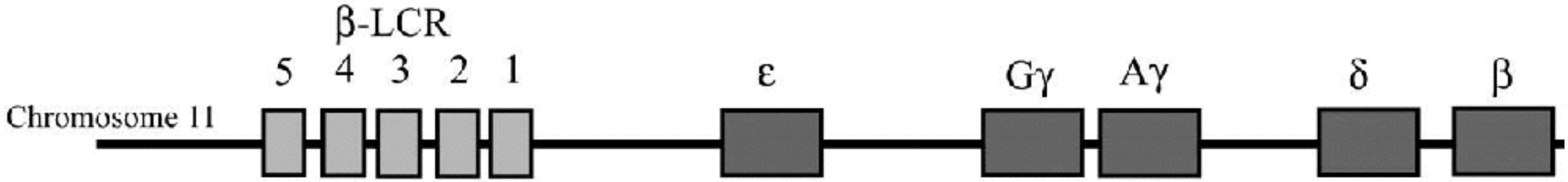
Globin genes



Globin genes



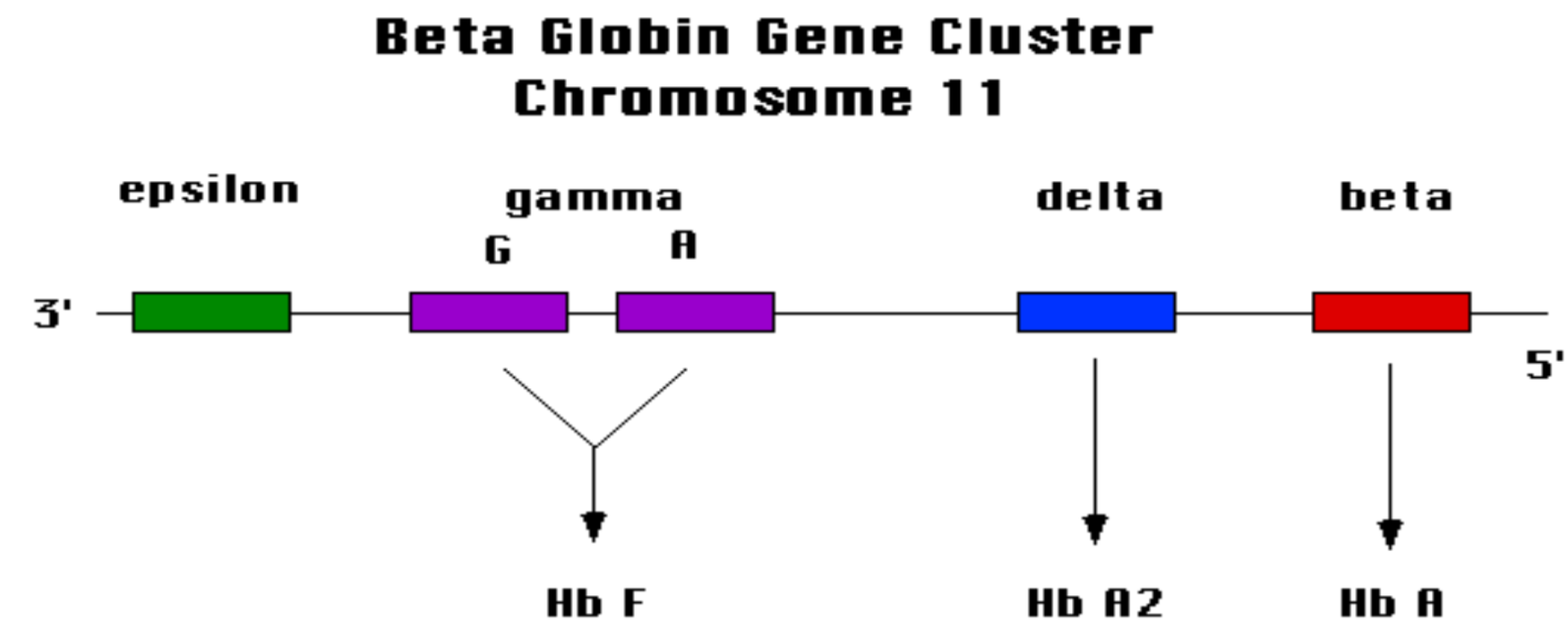
EMBRYO	FETUS	ADULT
ζ2ε2	α2γ2	α2β2
α2ε2		α2δ2
ζ2γ2		

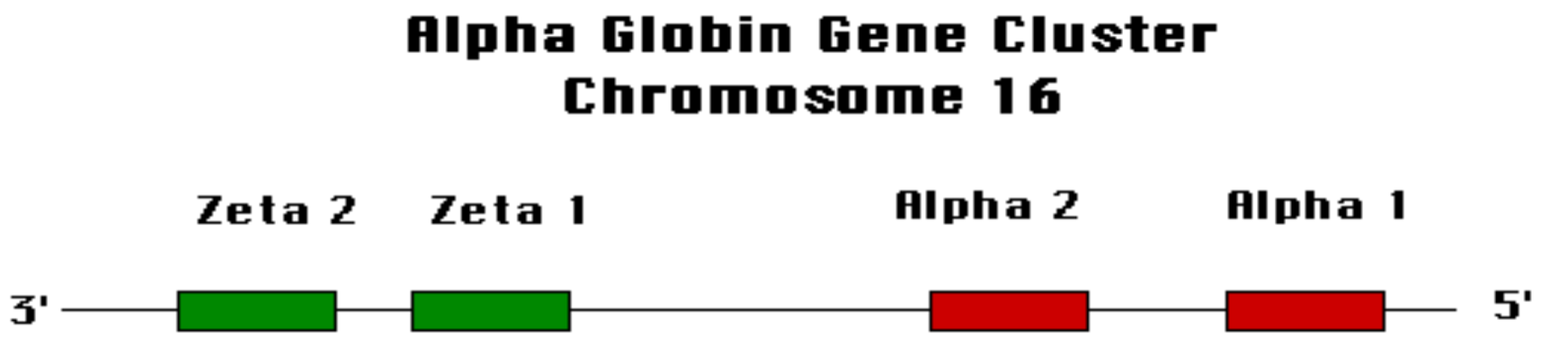


Approximate scale:

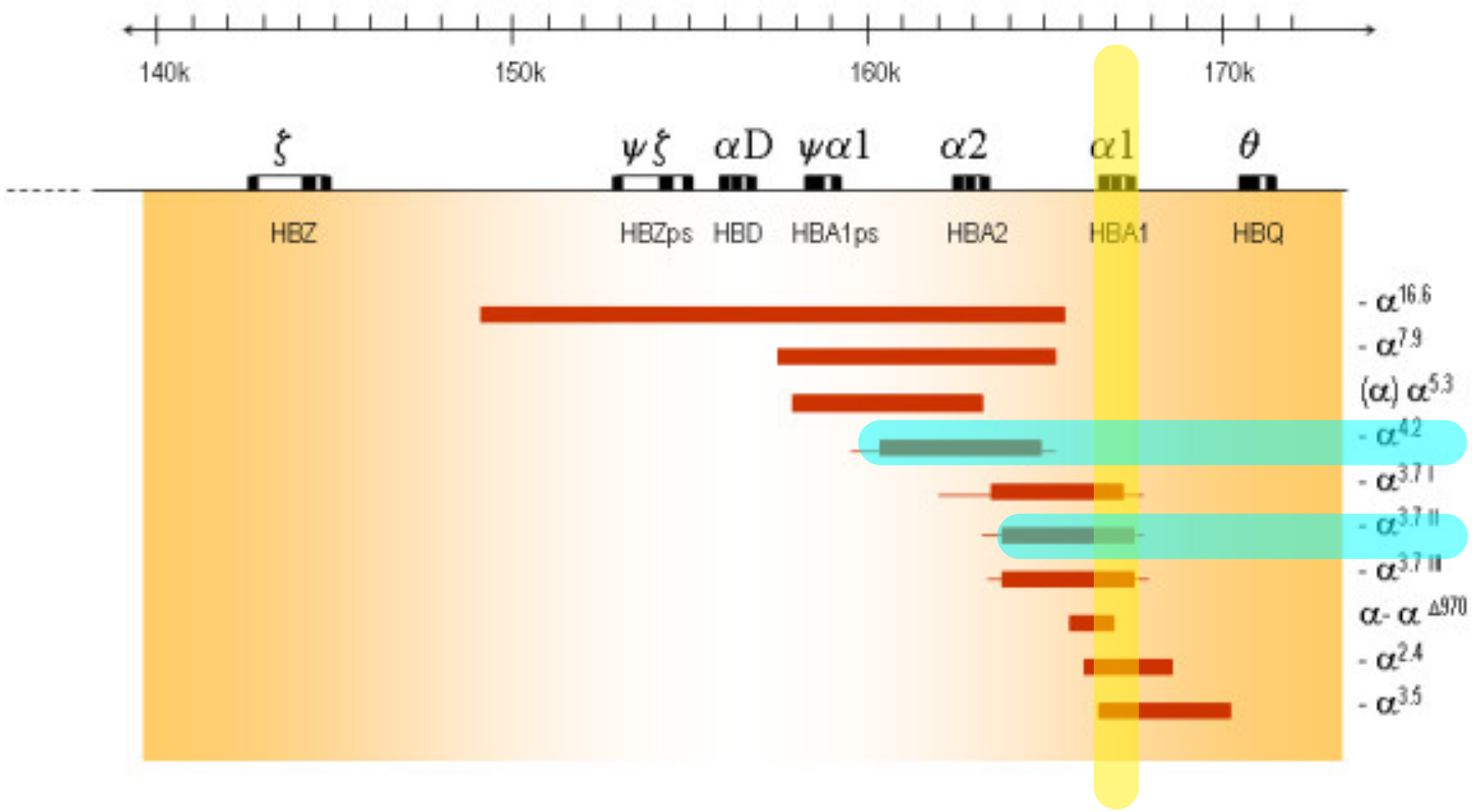
10 kb

Cluster of Globin gene





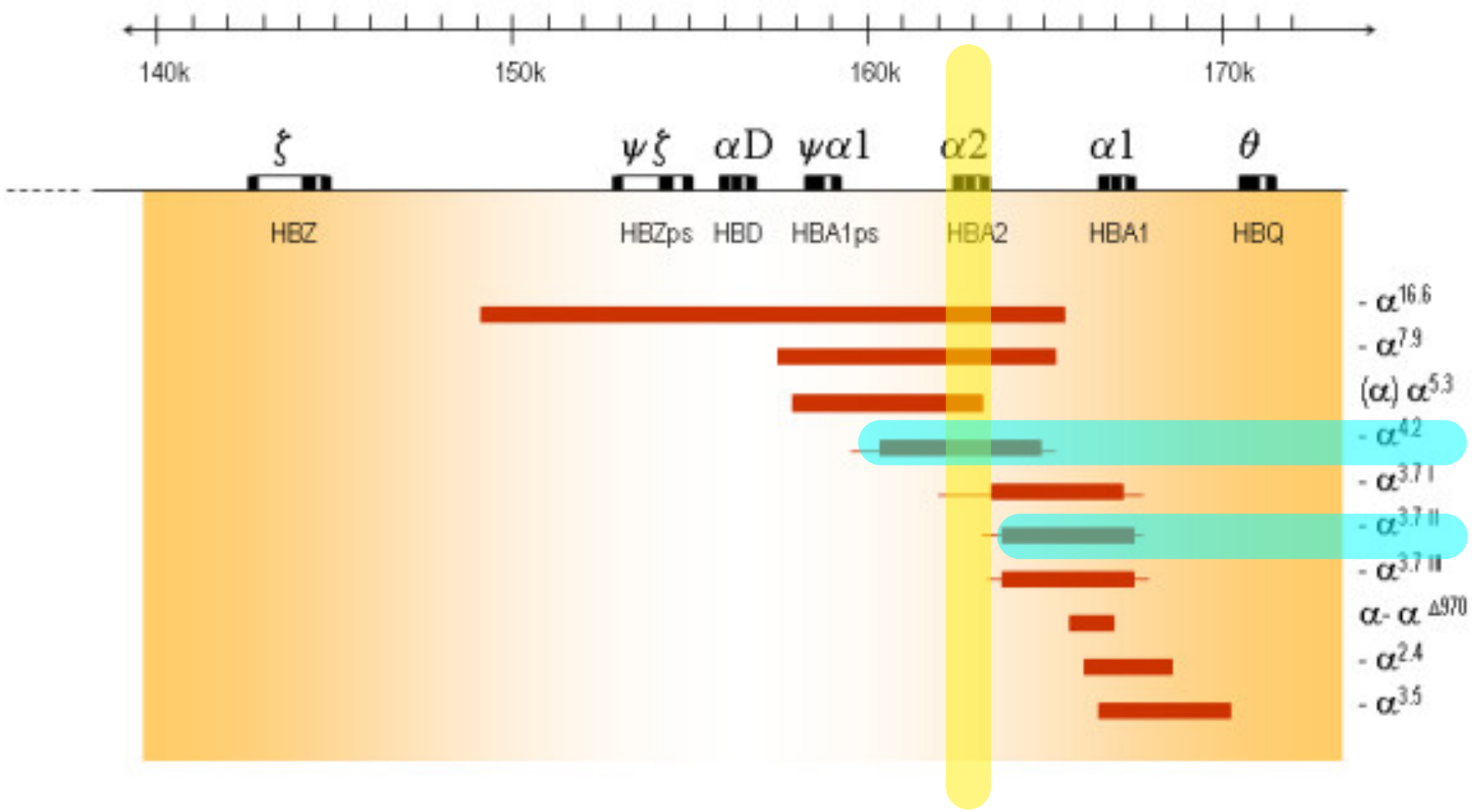
Deletion of alpha globin gene



- **Deletion of one alpha gene giving rise to alpha+-thalassemia / alpha thal-2 deletion**

- **Ref : Harteveld CL and Higgs DR, Orphanet J Rare 2010;5;13-33.**

Deletion of alpha globin gene



- **Deletion of one alpha gene giving rise to alpha+-thalassemia / alpha thal-2 deletion**

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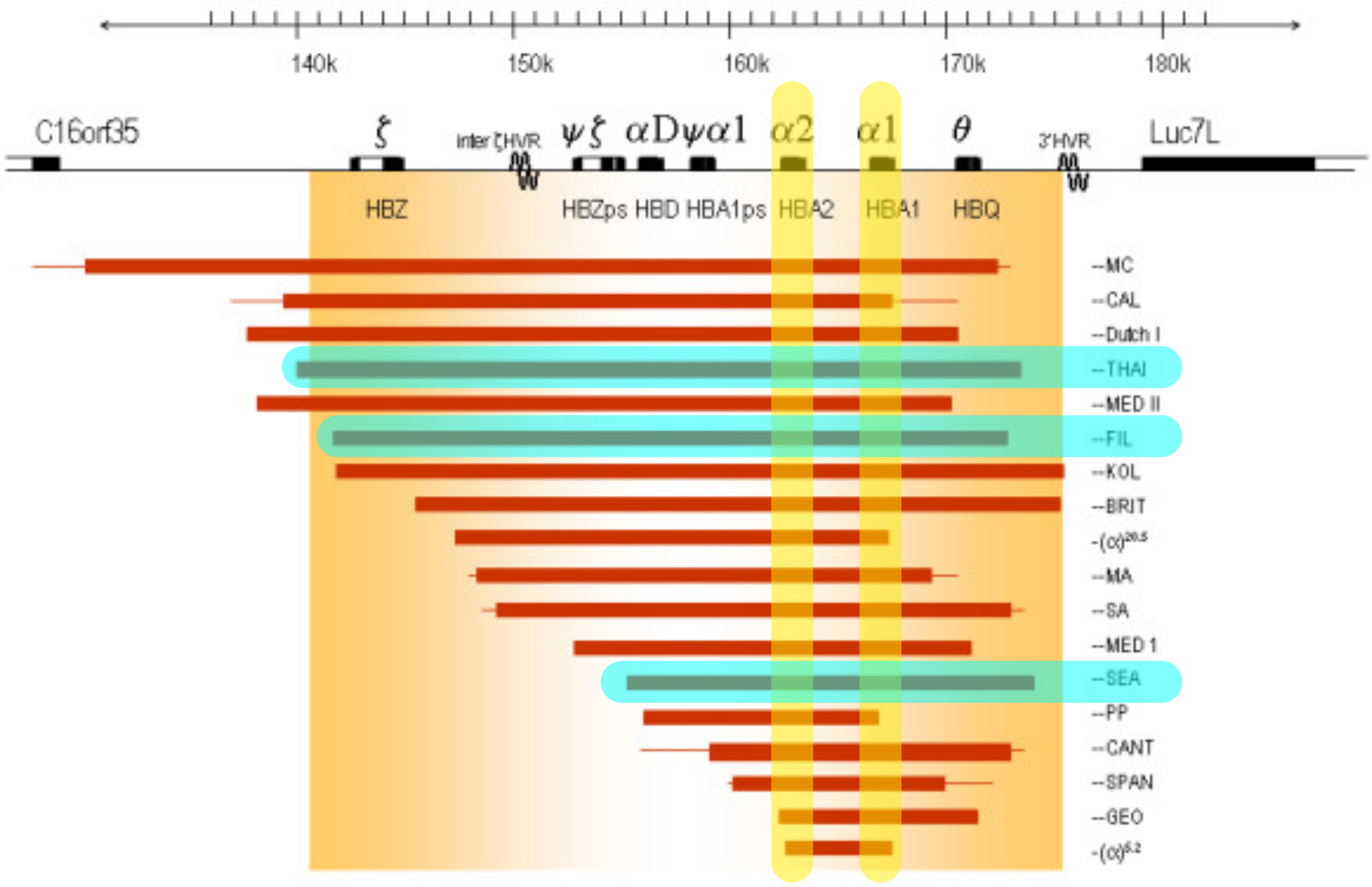
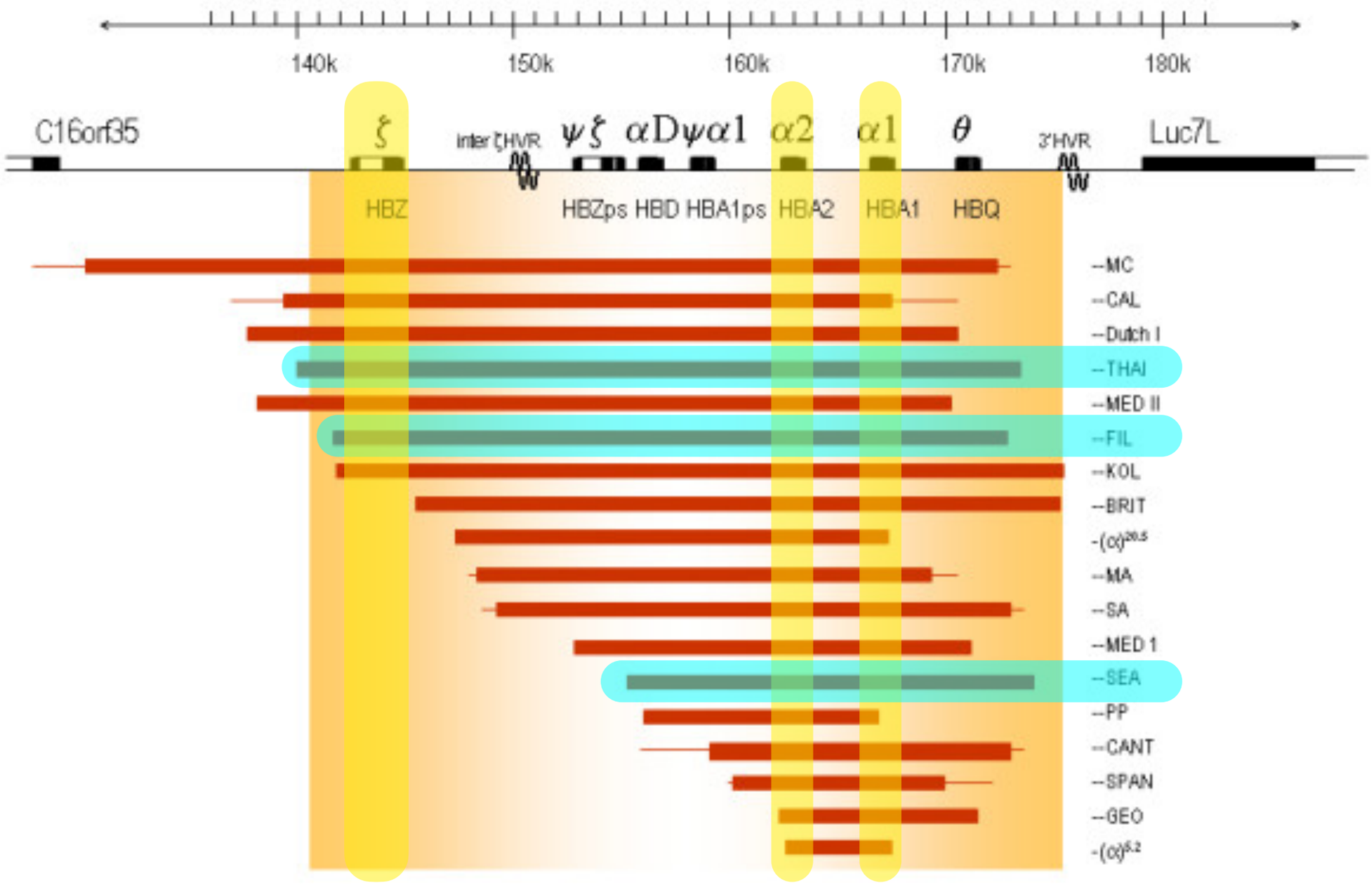
Deletion of alpha globin gene



- **Deletion of two alpha gene giving rise to alpha^{0}-thalassemia / alpha thal-1 deletion**

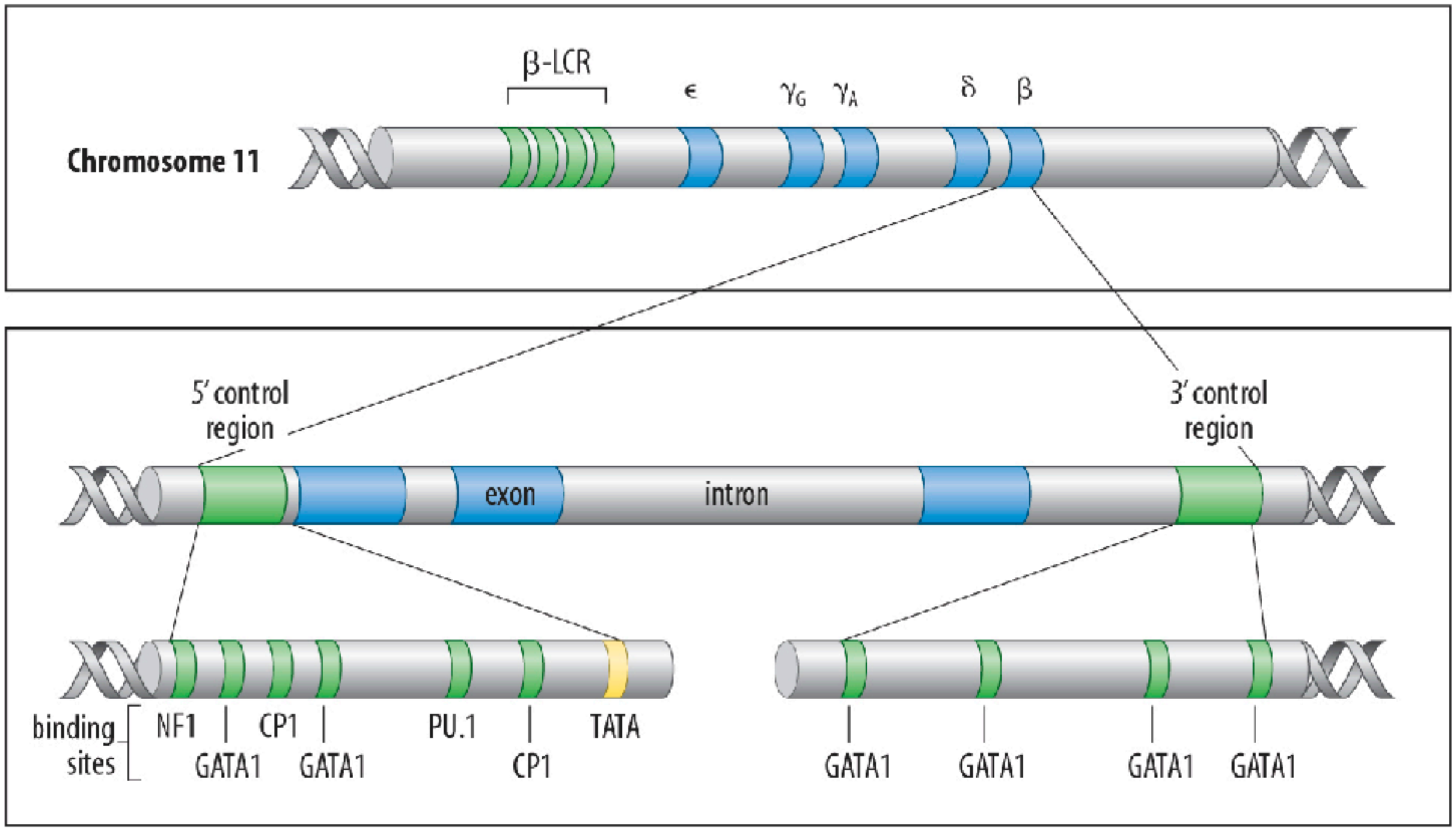
- **Ref : Harteveld CL and Higgs DR, Orphanet J Rare 2010;5;13-33.**

Deletion of alpha globin gene



- **Deletion of two alpha gene giving rise to alpha^0-thalassemia / alpha thal-1 deletion**
  - **Ref : Harteveld CL and Higgs DR, Orphanet J Rare 2010;5;13-33.**

Mutation of Beta globin gene



Thalassemia disease

Thalassemia Major

Tranfusion-Dependent Thalassemia
TDT

- **Severe anemia presenting early in life**
- **Require lifelong RBC transfusion**
- **If untreated, lead to dead in first decade of life**

Homozygous Beta thalassemia (β^0/β^0)

Beta thalassemia/Hb E (β^0/β^E)

Bart's hydrops fetalis

Thalassemia Intermedia

Non-Transfusion-Dependent Thalassemia
NTDT

- **Mild anemia**
- **Diagnosed usually in late childhood**
- **Occasional blood transfusion may be required**

Beta thalassemia intermedia (β^0/β^+ or β^+/β^+)

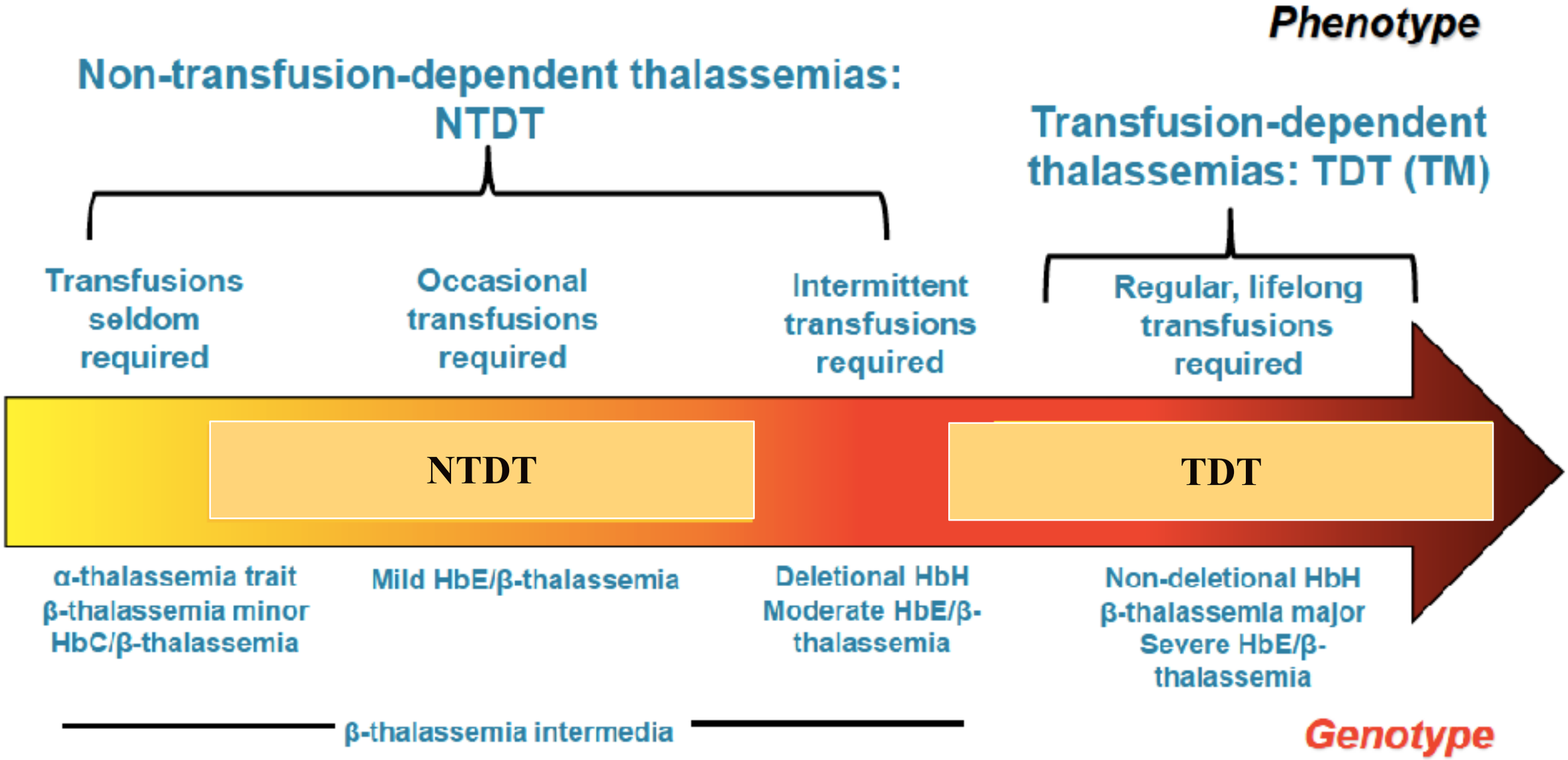
Beta thalassemia/Hb E (β^0/β^Eor β^+/β^E)

Hemoglobin H disease

Beta thalassemia

β^0-thalassemia mutation	*β^+-thalassemia mutation*
Codon 41/42 deletion (-CTTT)	*Codon 19 (A>G)*
Codon 17 (A>T)	*IVS I nt-5 (G>C)*
Codon 35 (C>A)	*IVS II nt-654 (C>T)*
Codon 71/72 (+A)	*Codon 26 (G>A)*

Spectrum of Thalassemia



[1]Muncie HL *et al. Am Fam Physician* 2009;80:339; [2]Galanello R *et al. Orphanet J Rare Dis* 2010, 5:11; [3]Harteveld C *et al. Orphanet J Rare Dis* 2010, 5:13; [4]Cohen AR *et al. Hematology Am Soc Hematol Educ Program* 2004;14

Point of Hemoglobin Typing

Must know!!!			
Hb A	**α2 β2**	***Hb H***	**β4**
Hb A2	**α2 δ2**	***Hb Bart's***	**ɣ4**
Hb F	**α2 ɣ2**		

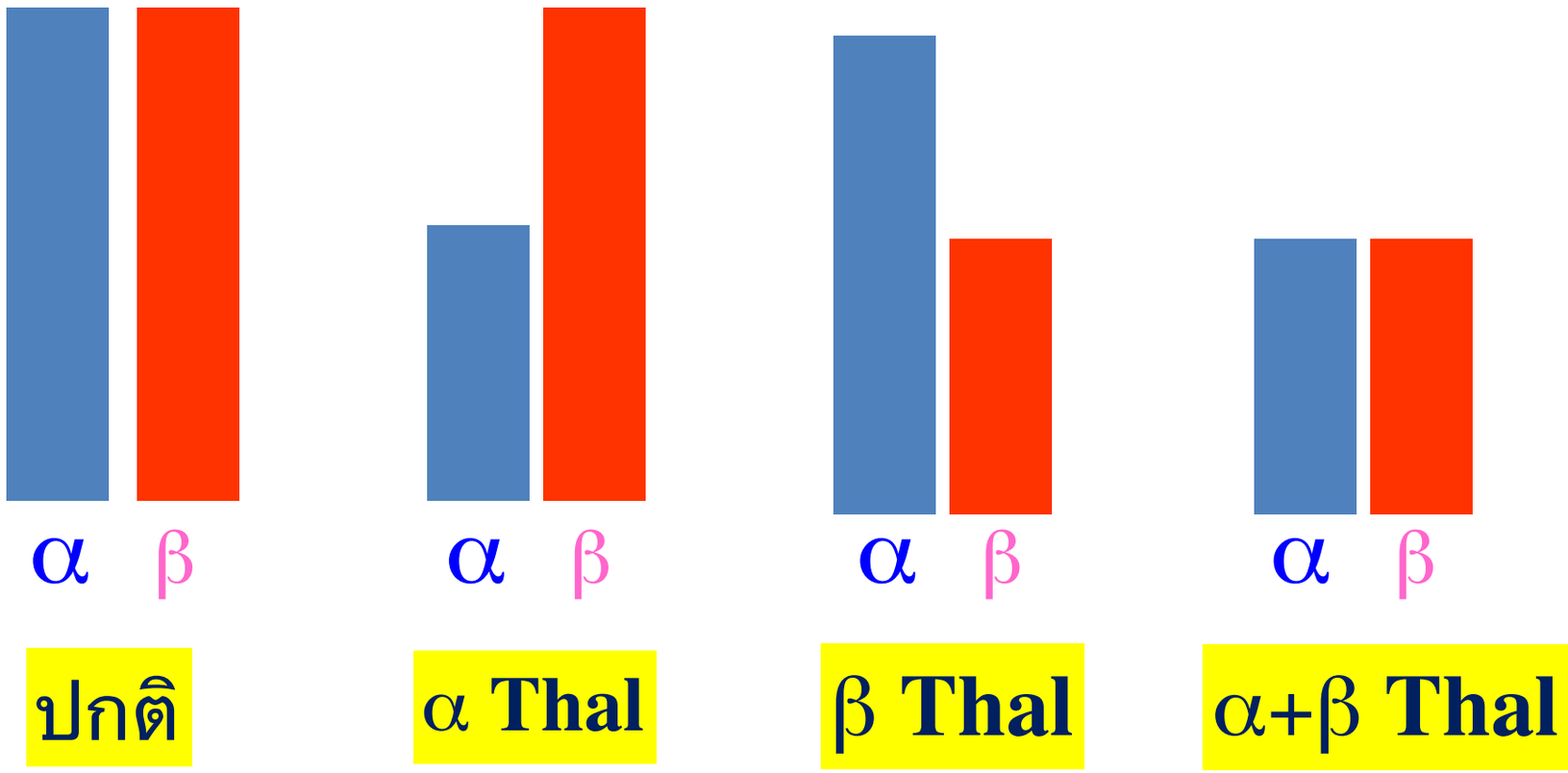
Normal : (αα,αα)(β,β)

A. Decrease production *:* ปริมาณ *Hb*

B. Abnormal production *(amino acid)*

Variant Hb Ex; Hb E, Hb Cs

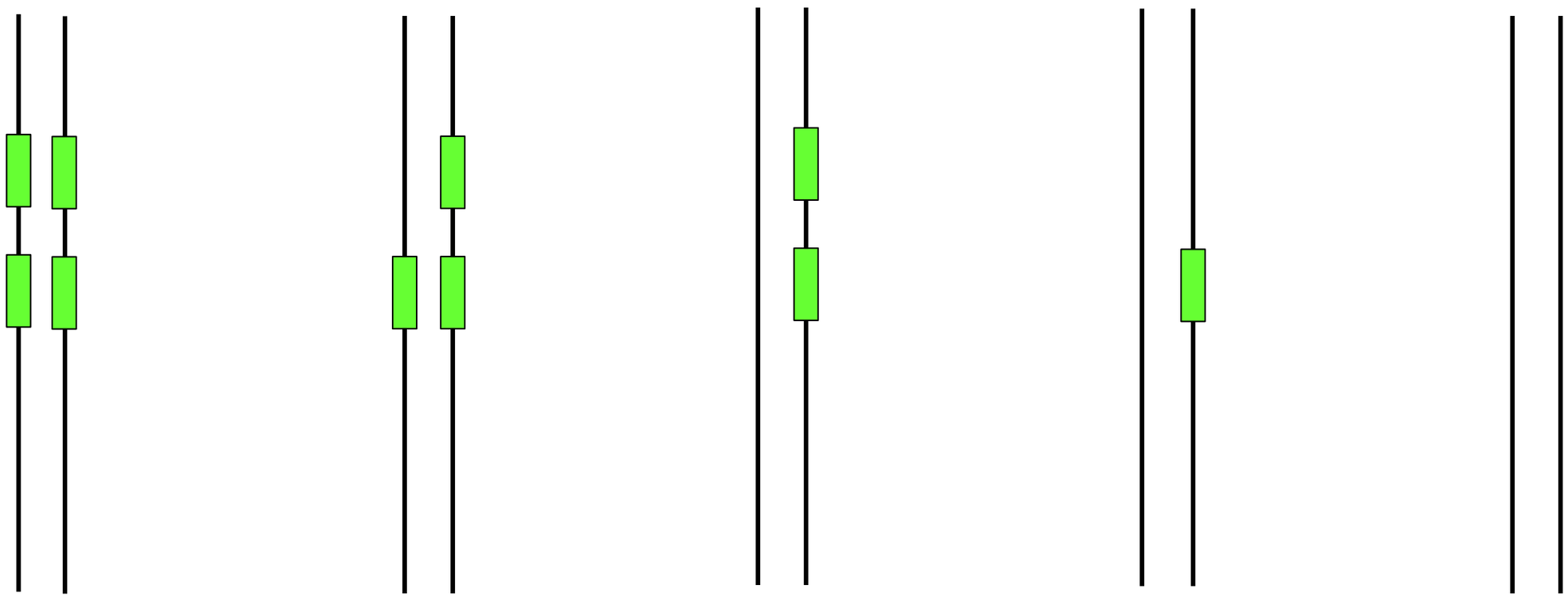
Principle of thalassemia



ปกติ | α Thal | β Thal | α+β Thal

Alpha globin

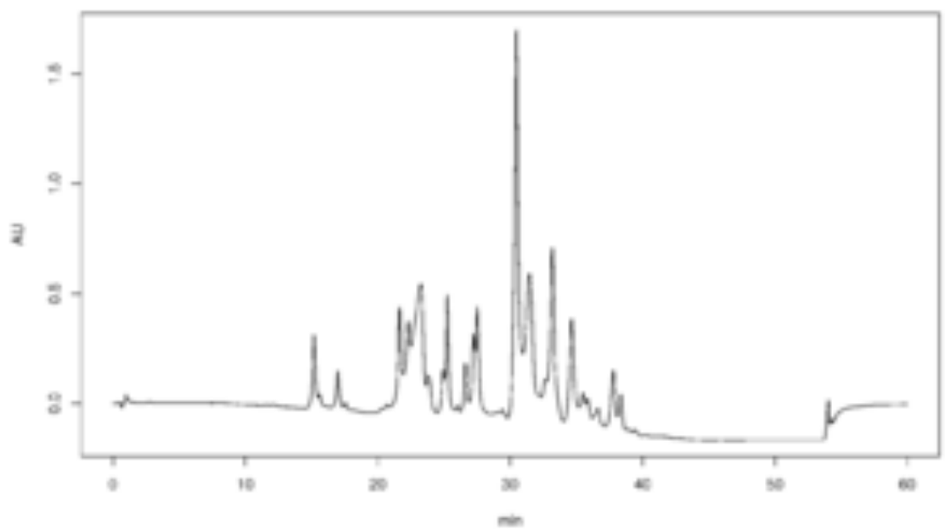
	-25 %	-50%	-75%	-100%
Normal	α thal 2	α thal 1	H diseases	Bart's hydrops



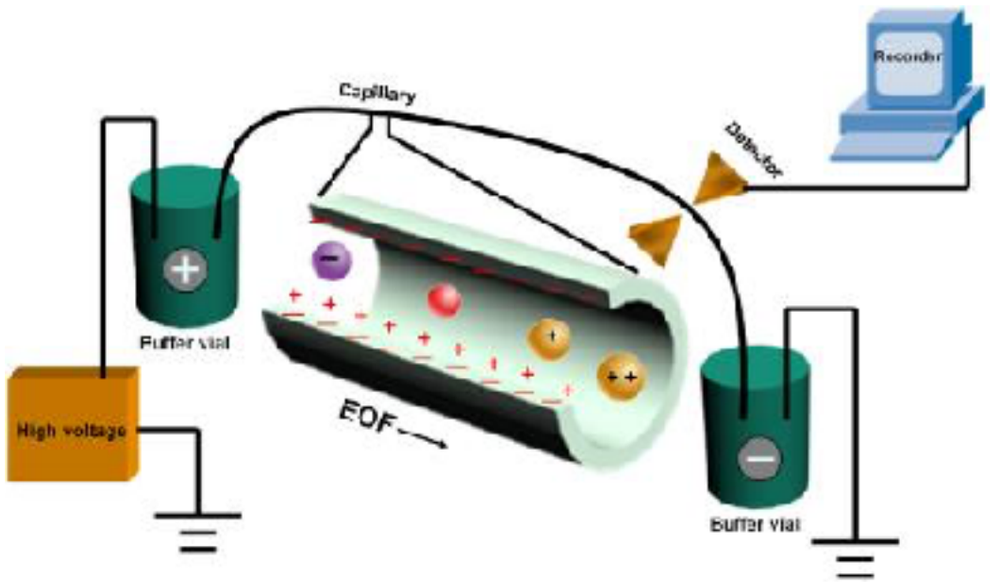
Alpha globin genotype ***Chain Deletion***

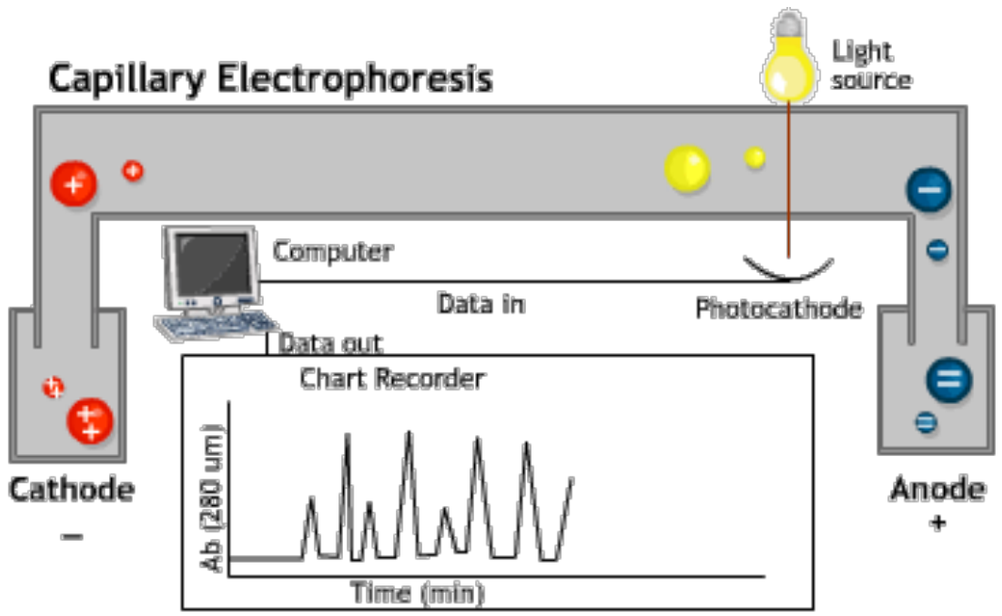
Hemoglobin typing

HPLC



Capillary Electropheresis





%Hb F = 0.9	% Hb A_2 = 6.0	Hb-typing : A_2A(High A_2)

แปลผล : : beta thal trait

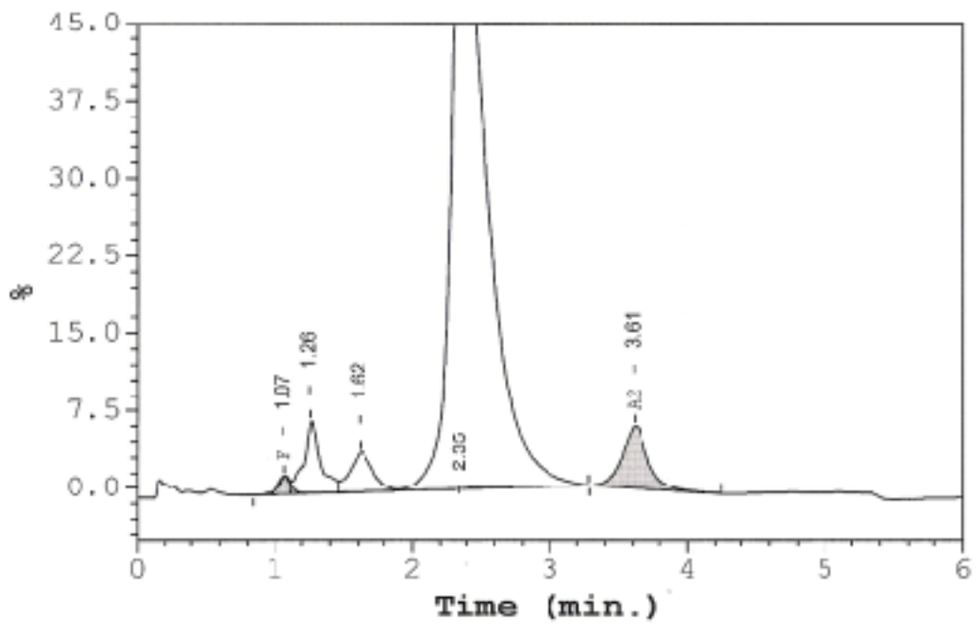
Peak Name	Calibrated Area%	Area%	Retention Time (min.)	Peak Area
F	0.9	---	1.07	14915
P2	---	4.5	1.26	79750
P3	---	3.6	1.62	63911
Ao	---	85.1	2.35	1517026
A2	6.0*	---	3.61	107770

Total Area :1,783,372

F Concentration = 0.9 %
A2 Concentration =6.0* %

Analysis comments:

*Values outside of expected ranges



Genotype β/β⁰ หรือ β/β+

ตรวจ β –mutation DNA

ตรวจ α –thal 1 DNA

%Hb F = 0.7	% Hb E = 26.2	Hb-typing : EA

แปลผล: Hb E trait

หมายเหตุ : % E > 25 %Always not have α thal 1 included

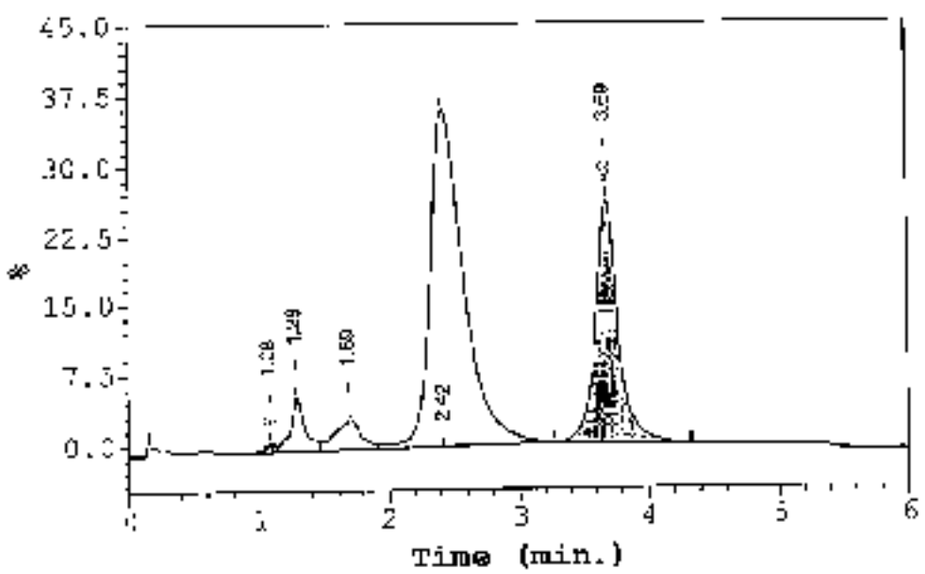
F	0.7	—	1.08	13442
P2	—	4.2	1.28	96380
P3	—	4.4	1.69	100671
Ao	—	60.6	2.42	1385584
A2	26.2*	—	3.69	691959

Total Area :2,238,036

F Concentration = 0.7 %
A2 Concentration =26.2* %

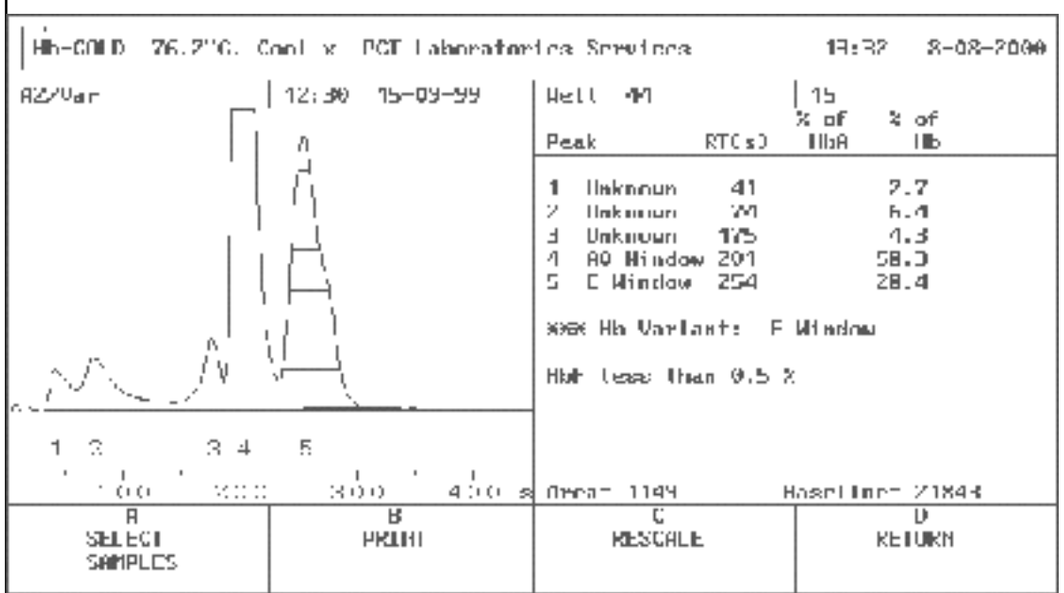
Analysis comments:

*Values outside of expected ranges



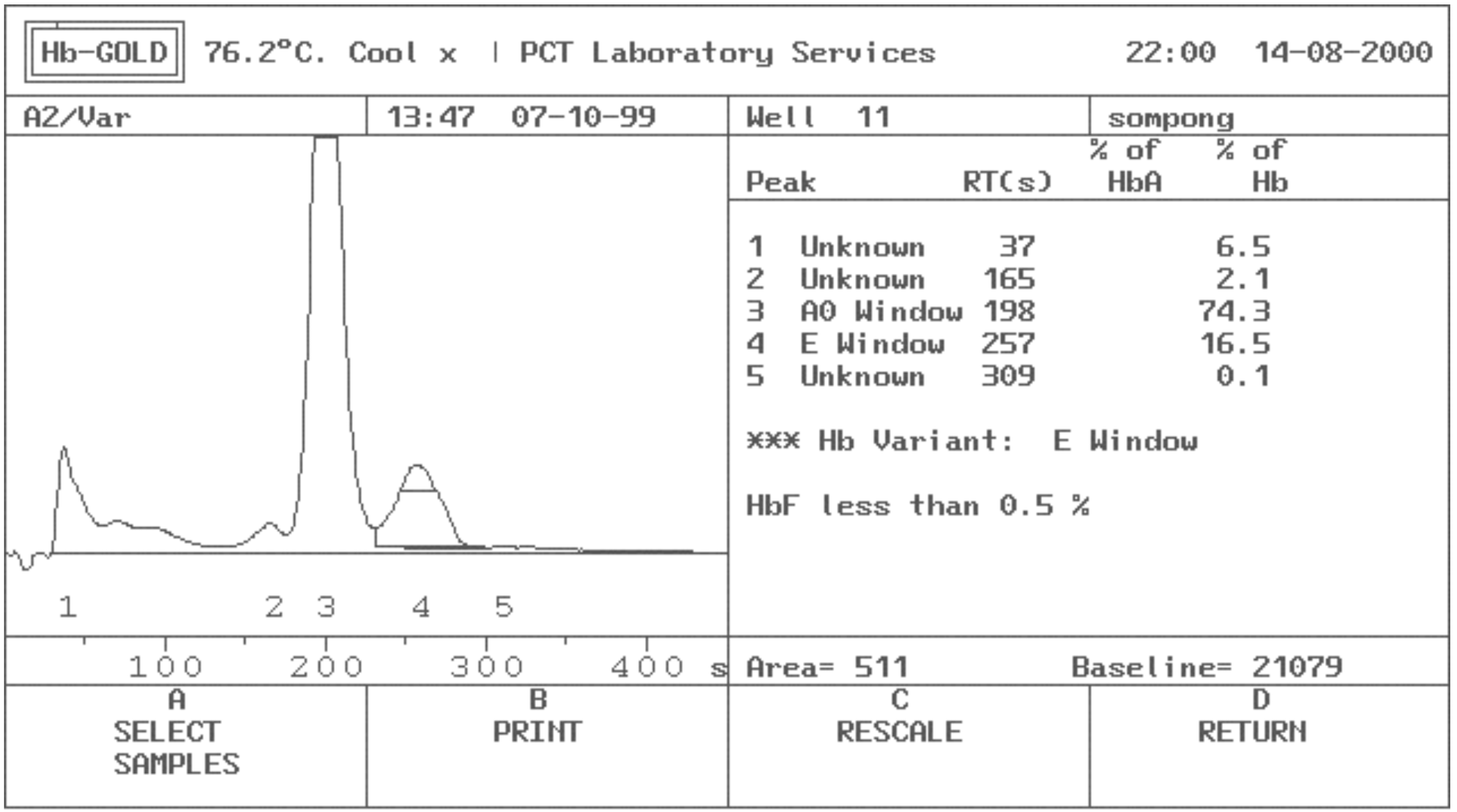
Genotype β/β^{E}

กรณี EA , %HbE>25 ที่คู่สมรสเป็น Hb typing ชนิดใดก็ตามจะ<u>ไม่ตรวจ α **–Thal1 DNA**</u>



Hb E + Hemoglobin H disease

(β^{E}/β + α *thal-1/*α *thal-2)*



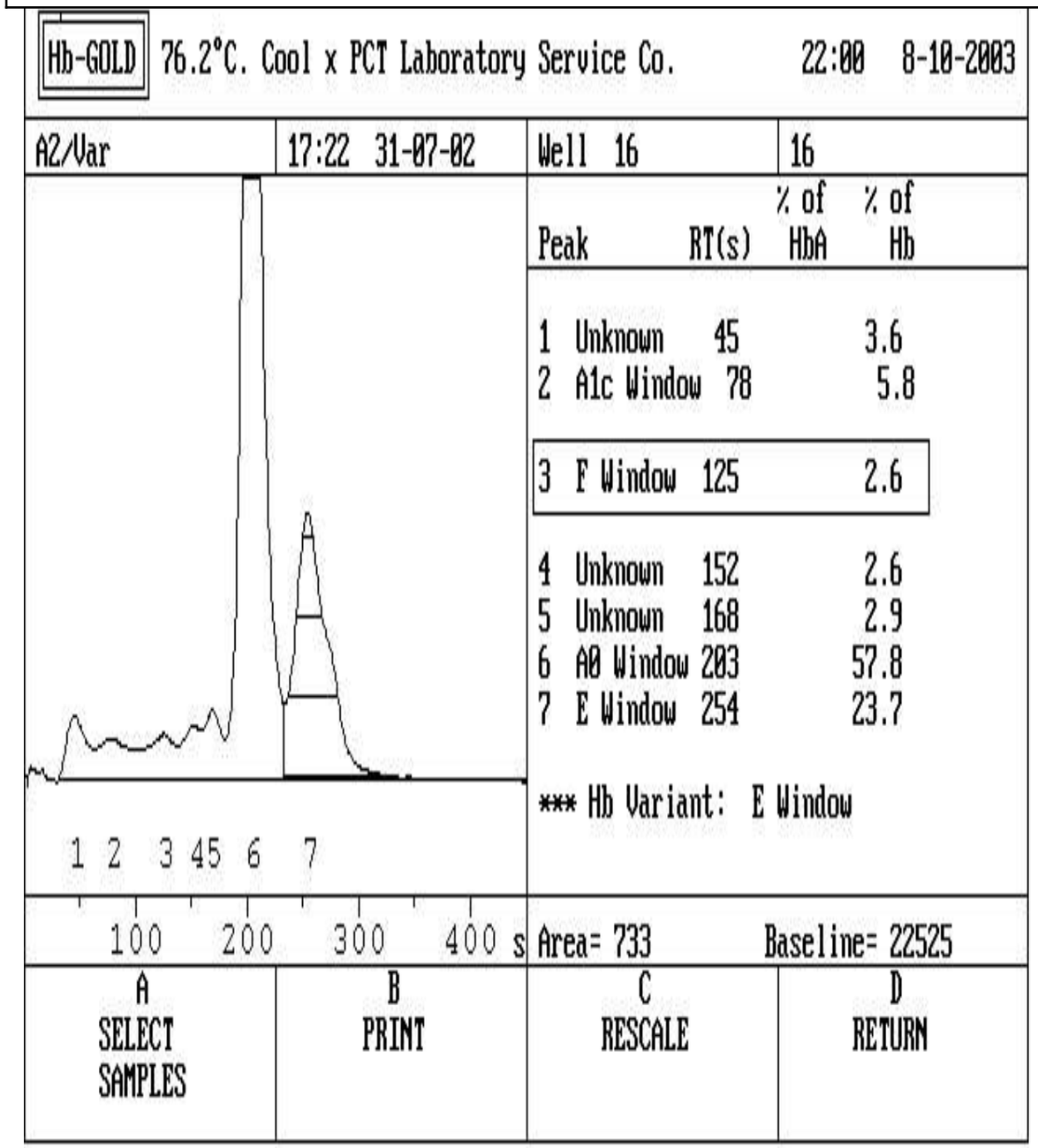
% E < 20 %

Always have α **thal disease included**

%Hb F = 2.6	% Hb E = 23.7	Hb-typing : EA

แปลผล: Hb E trait(Prob with α **thal 1)**

Hb E Trait ที่%HbE < 25



Genotype β / β ^{E}

ตรวจ
α –thal 1 DNA

%Hb F = 1.4	% Hb E = 84.0	Hb-typing : EE
แปลผล: Homozygous Hb E		

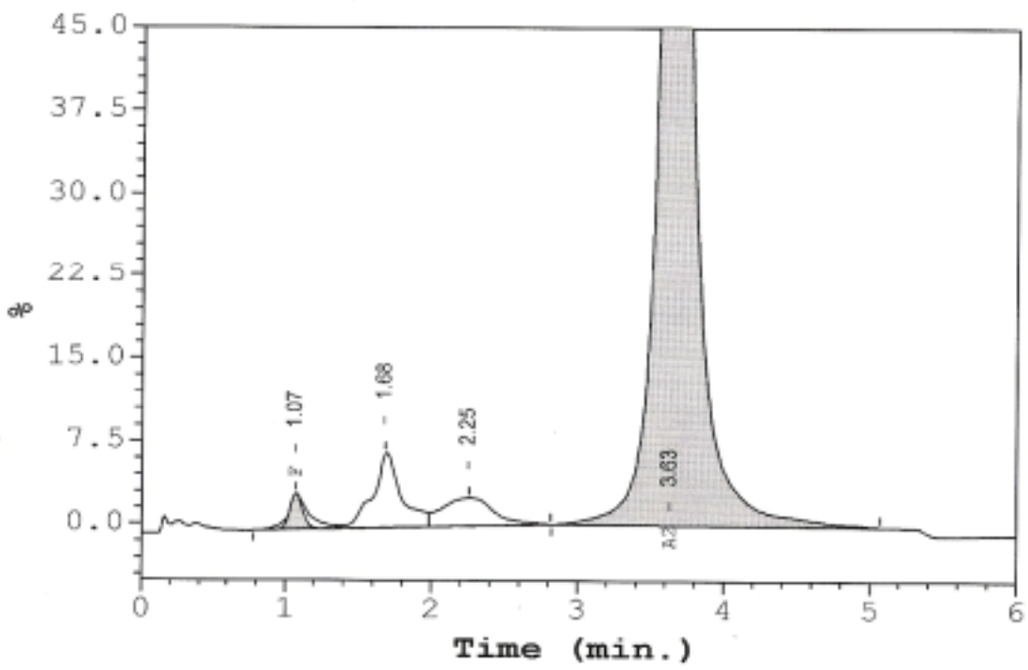
Peak Name	Area%	Area%	Time (min.)	Area
F	1.4*	---	1.07	27973
P3	---	5.8	1.68	130033
Ao	---	3.9	2.25	87840
A2	84.0*	---	3.63	2009977

Total Area :2,255,823

F Concentration = 1.4*%
A2 Concentration =84.0* %

Analysis comments:

*Values outside of expected ranges



Genotype β^{E}/β^{E}

ตรวจ
α –thal 1 DNA

%Hb Bart's Present	%Hb F = 2.2	% Hb E = 14.7	%Hb CS = 2.6	Hb-typing : CSEABart's
แปลผล: EA Bart's diseases with Hb CS				

Peak Name	Calibrated Area%	Area%	Retention Time (min.)	Peak Area
F	2.2*	---	1.08	32236
Unknown	---	3.1	1.25	53327
P3	---	3.1	1.61	52537
Ao	---	73.2	2.45	1241976
A2	14.7*	---	3.67	273128
Unknown	---	0.5	4.66	8135
C-window	---	2.1	4.92	36159

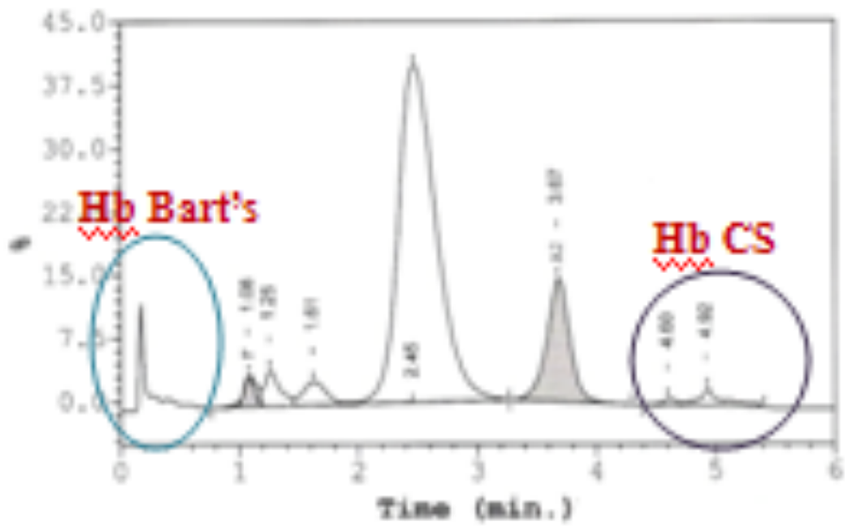
Hb CS = 2.6%

Total Area :1,697,498

F Concentration = 2.2*%
A2 Concentration =14.7* %

Analysis comments:

*Values outside of expected ranges



Genotype β / β^{E}

Genotype α-thal1 / α^{cs}

%Hb Bart's H Present	%Hb F 0.4	% Hb A_2 1.5	Hb-typing : A_2ABart's H
แปลผล: Hemoglobin H disease			

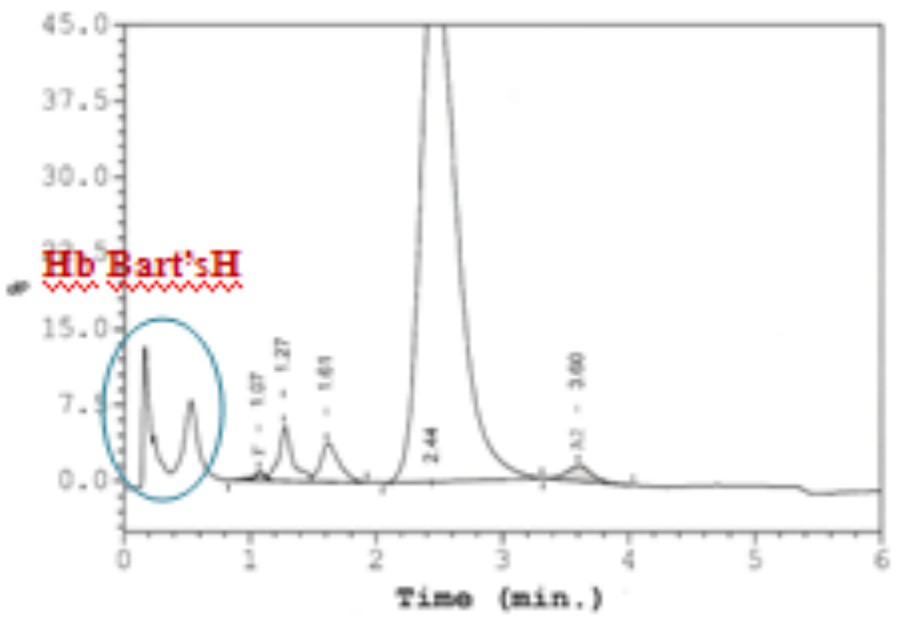
Peak Name	Calibrated Area%	Area%	Retention Time (min.)	Peak Area
F	0.4	---	1.07	7544
P2	---	3.6	1.27	63280
P3	---	3.3	1.61	57175
Ao	---	90.8	2.44	1578815
A2	1.5*	---	3.60	32266

Total Area :1,739,080

F Concentration = 0.4 %
A2 Concentration =1.5* %

Analysis comments:

*Values outside of expected ranges



Genotype β / β

α-Thal1 / α-Thal2

%Hb A = 33.5	%HbF 50.8	% Hb E 14.0	Hb-typing : EFA
แปลผล: beta thal /Hb E			

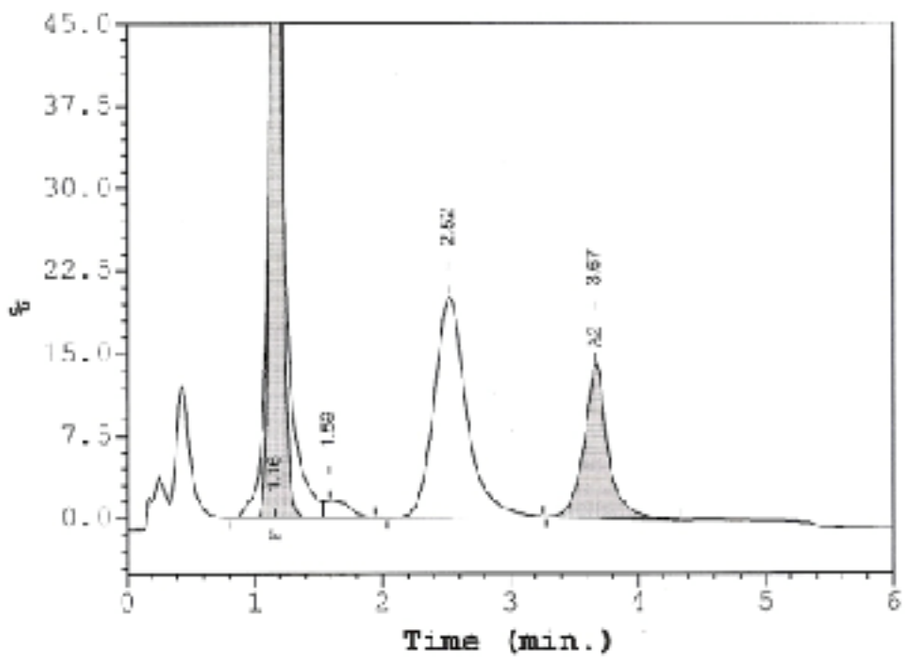
Peak Name	Calibrated Area%	Area%	Retention Time (min.)	Peak Area
F	50.8*	---	1.16	573247
P3	---	1.9	1.59	23120
Ao	---	33.5	2.52	402178
A2	14.0*	---	3.67	202552

Total Area :1,201,097

F Concentration = 50.8* %
A2 Concentration =14.0* %

Analysis comments:

*Values outside of expected ranges



Genotype β^{+} / β^{E}

ตรวจ
α –thal 1 DNA

%Hb F	% Hb A2	%Hb Bart's	Hb-typing : Bart's
0.0	0.0	100%	
แปลผล: Hb Bart's hydrop fetalis			

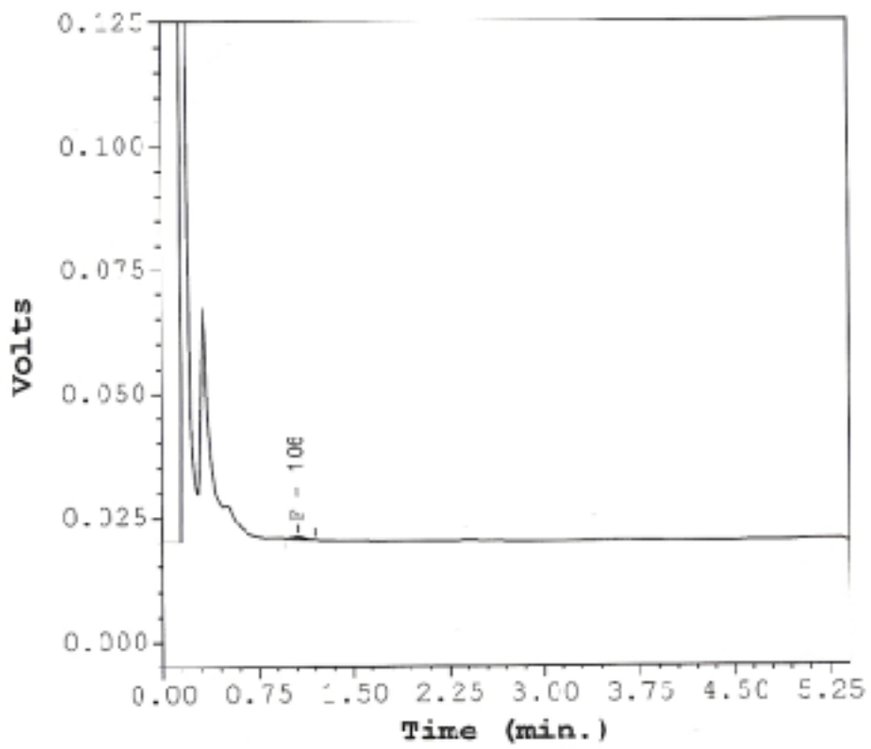
Peak Name	Calibrated Area%	Area%	Retention Time (min.)	Peak Area
F	111.2*	---	1.06	2673

Total Area :2,673*

F Concentration = 111.2* %
A2 Concentration = %

Analysis comments:

*Values outside of expected ranges



α-Thal1 / α-Thal1

red cell indices and Hb typing

normocyte

HbA2+A

Dx:Normal
α-thal2 trait

Hb A+CS

Dx:Hb CS

HbE+A

HbE Determination

HbE=25-35%

Dx:HbE trait
α-thal2/HbE

HbE<21%

Hb<10g/dl

Dx:HbE trait
with iron def.
α-thal1/HbE
with iron def.

Hb>10g/dl

Dx: α-thal1/HbE

microcyte

Hb EE

Dx:Homo HbE

Thal.Dz

β-thal Homo
β-thal/HbE
HbH
AEBart's
EFBart's

HbA2+A

HbA2
Determination

HbA2>3.5%

Dx: β-thal tr.

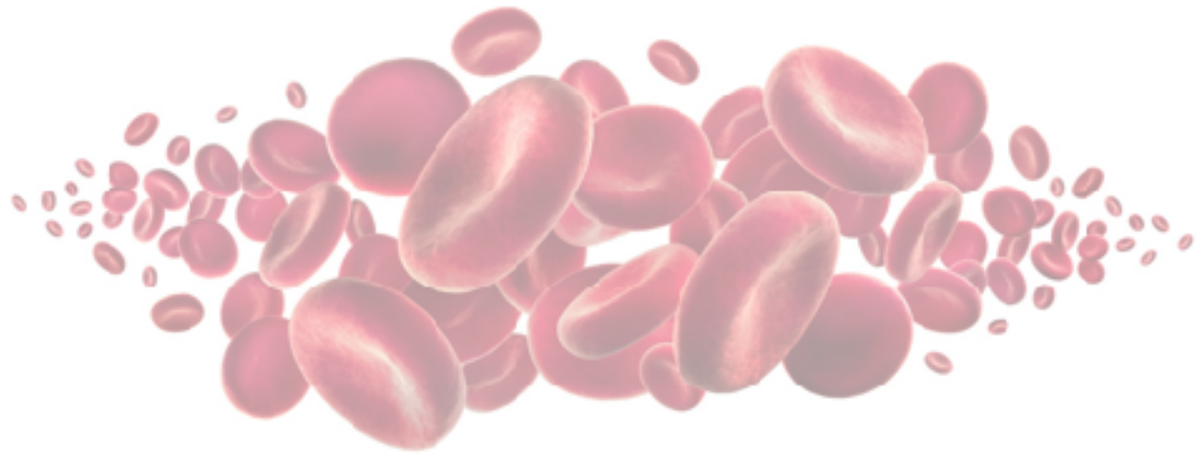
HbA2<3.5%

Hb<10g/dl

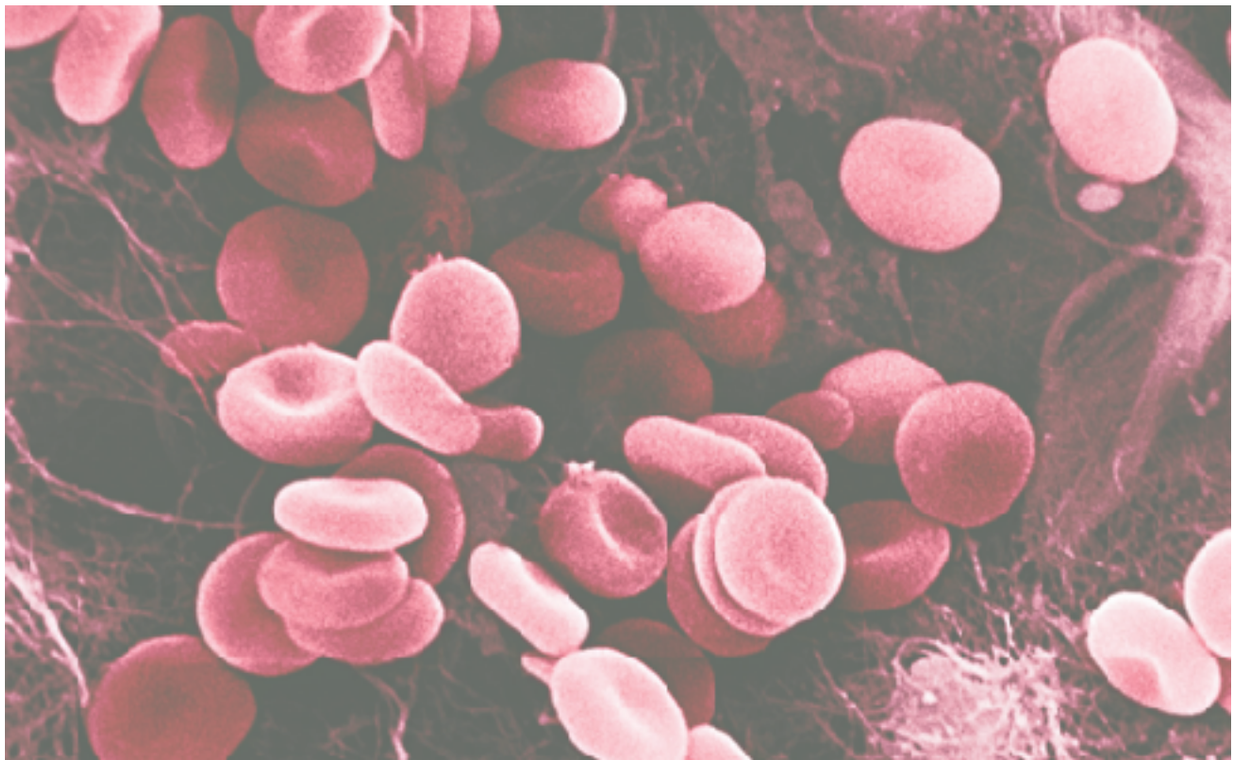
Dx: iron def.
iron def. on
top of thal tr.

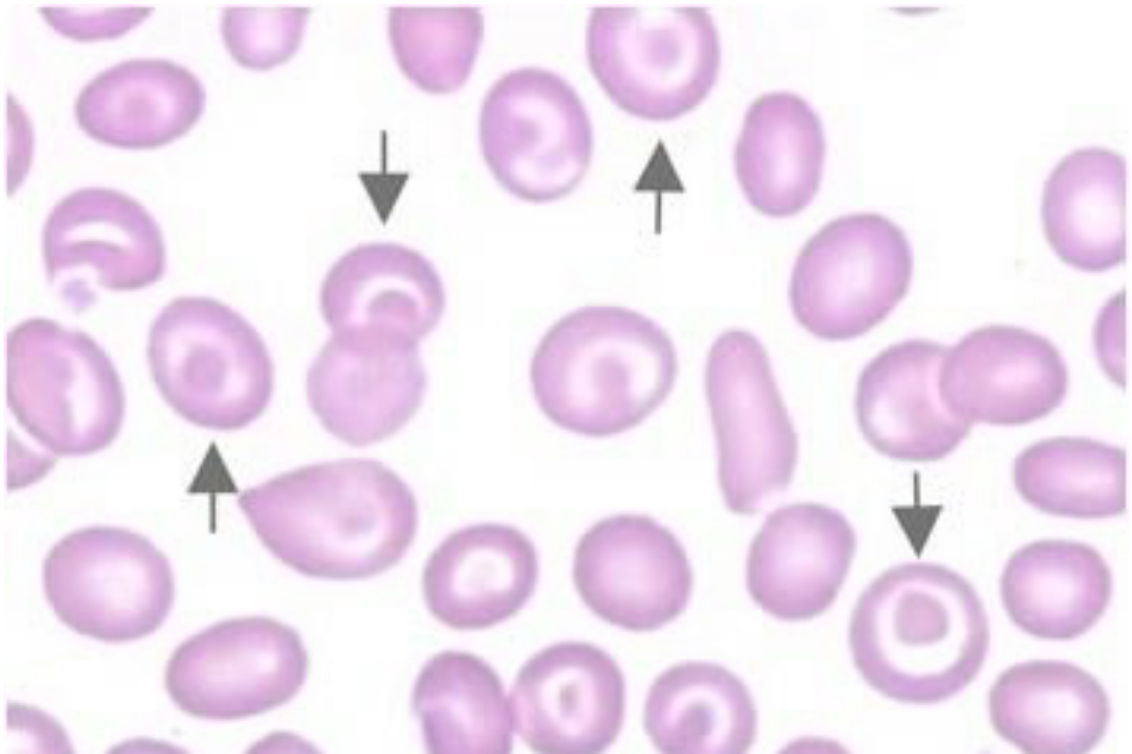
Hb>10g/dl

Dx: α-thal1 tr.

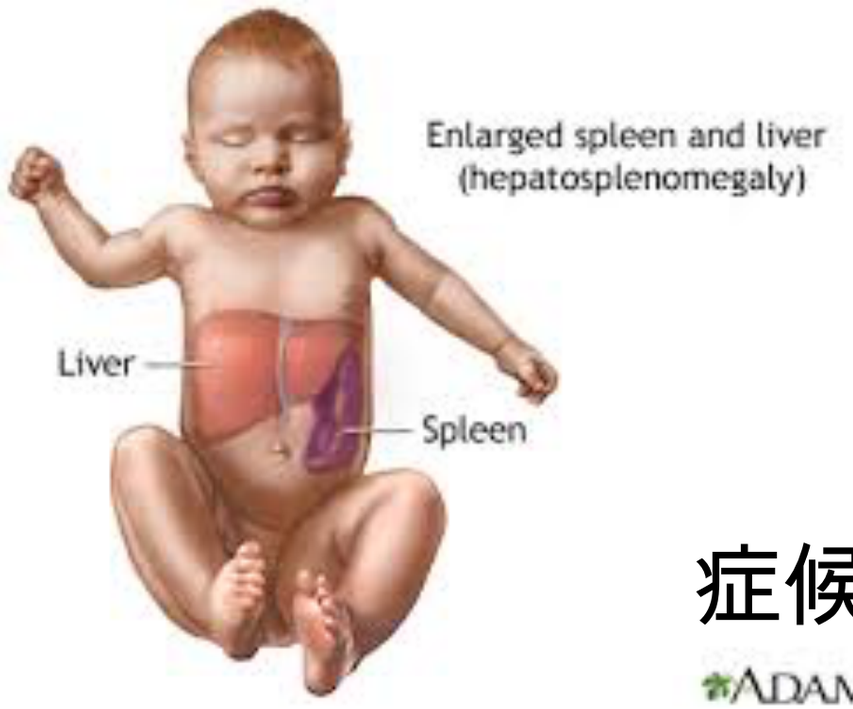


Clinical features

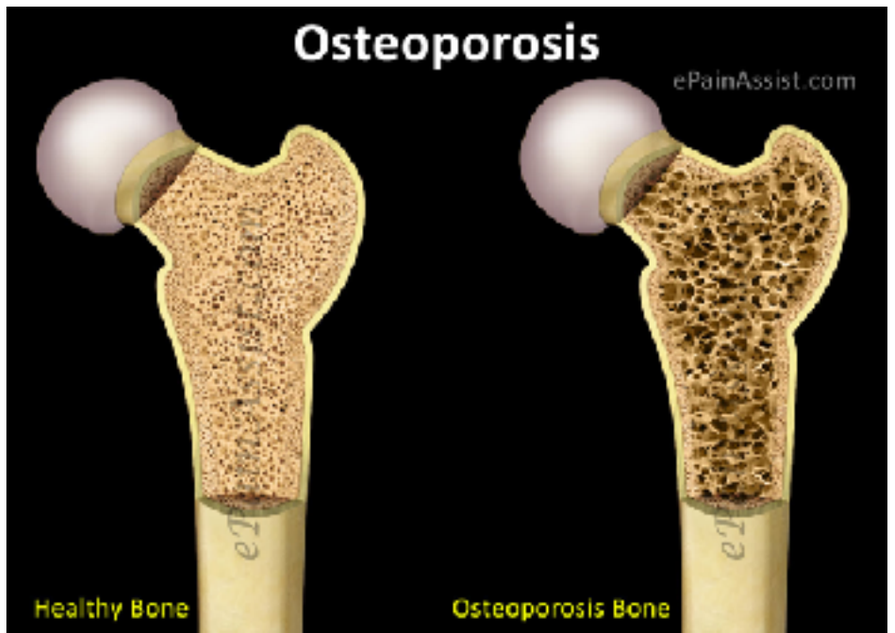




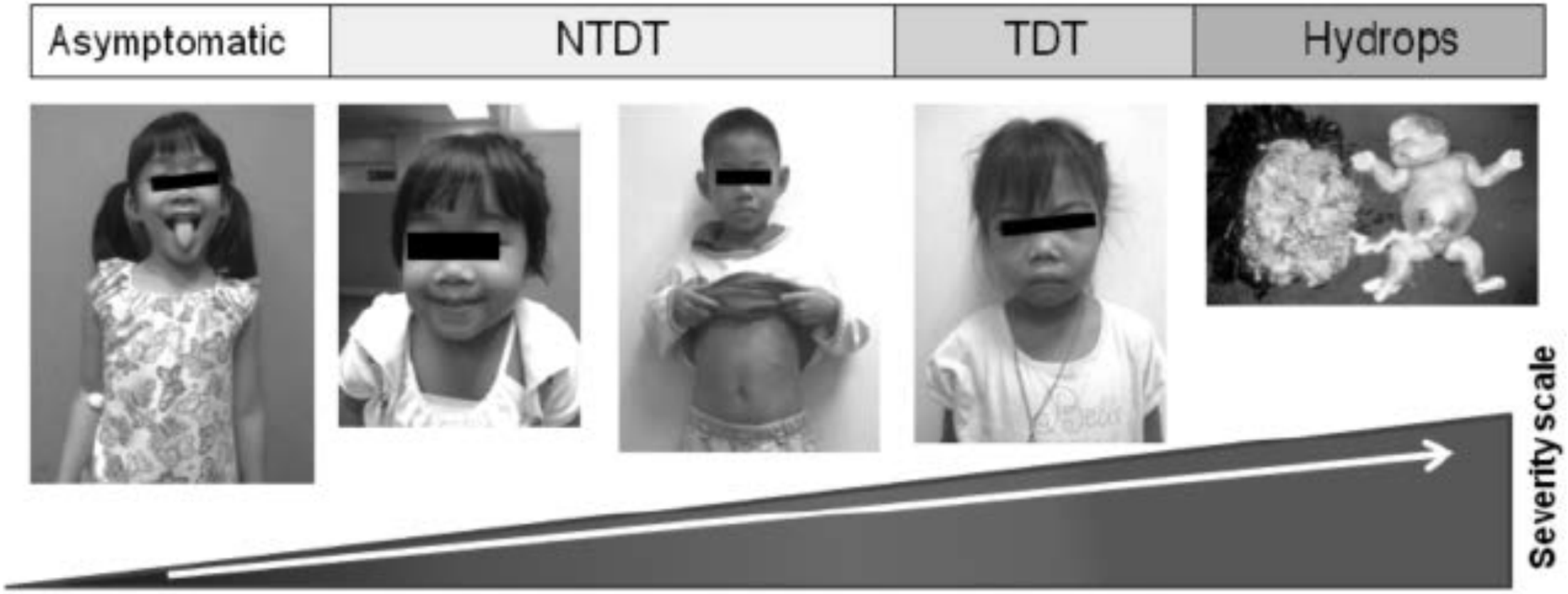
Beta Thalassemia Major – bone changes



症候



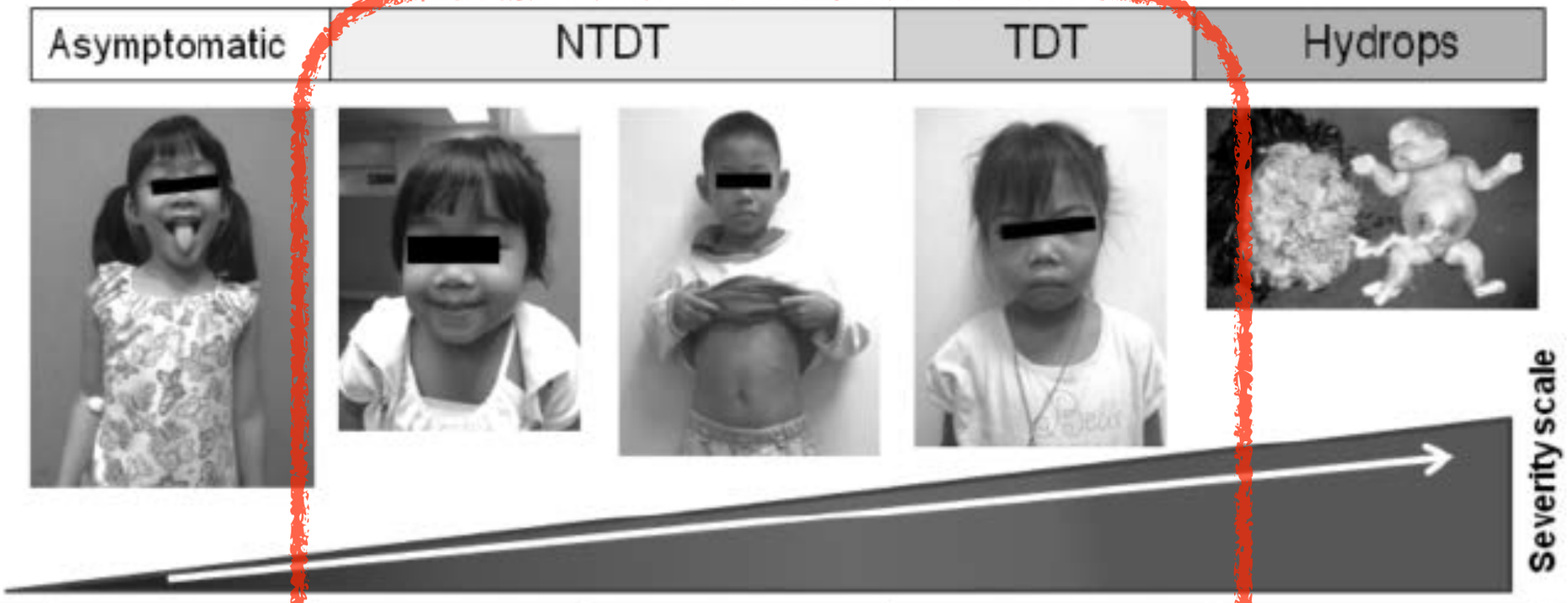
Thalassemia syndromes



Genotypes	Silent HbH	Deletional HbH	Non-deletional HbH	Transfusion – dependent HbH		HbH hydrops
1°	--/-α --/$\alpha^T\alpha$	--/-α	--/$\alpha^T\alpha$ --/$\alpha\alpha^T$	--/-α	--/$\alpha^T\alpha$ --/$\alpha\alpha^T$ $\alpha^T\alpha/\alpha^T\alpha$	--/$\alpha^T\alpha$ $\alpha^T\alpha/\alpha^T\alpha$
2°	β thal	+ β globin variants such as Hb E, Hb C, Hb S, homozygous Hb E, β thal/Hb E etc.		-		-
3°	-			KLF-1		-

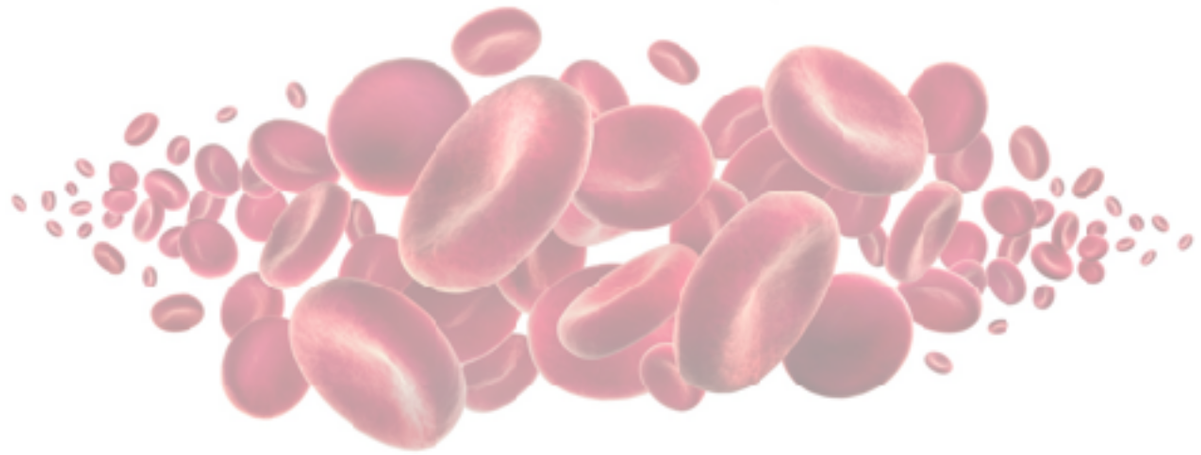
* V. Viprakasit et al., *Alpha-thalassemia syndromes: from clinical and molecular diagnosis to bedside management*

Thalassemia syndromes

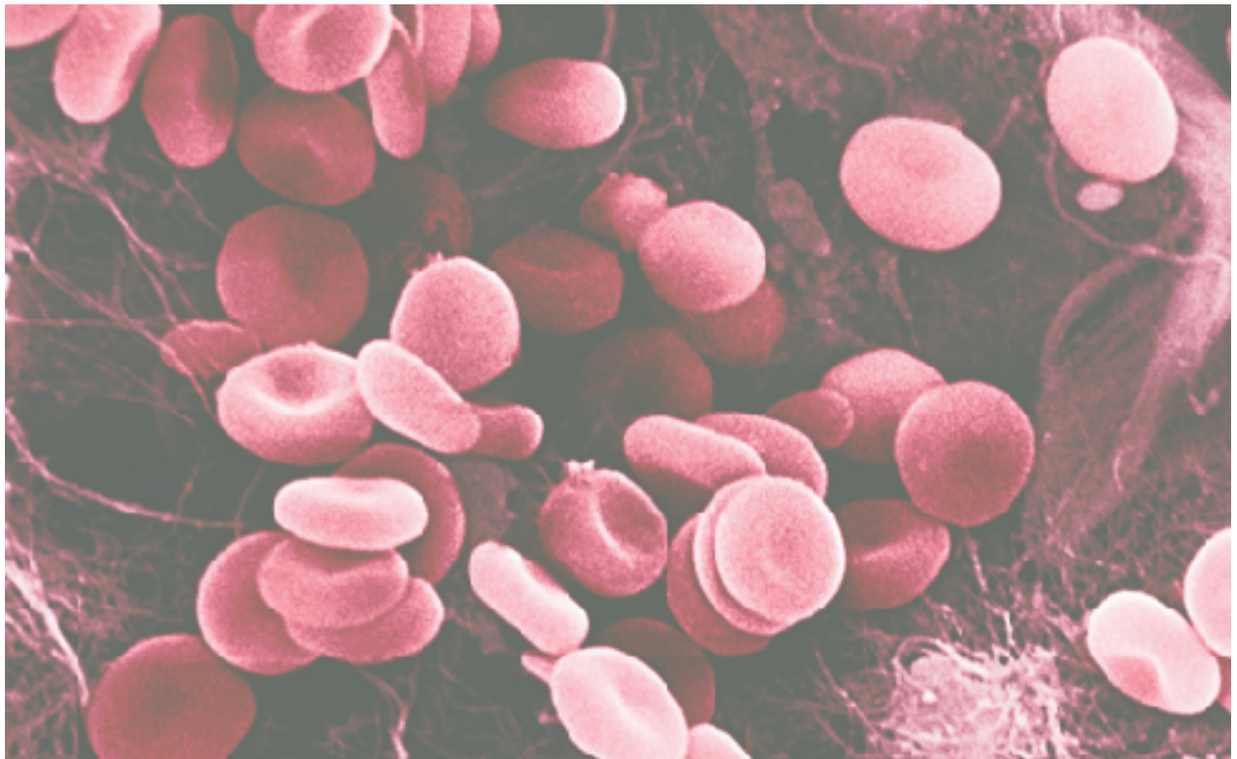


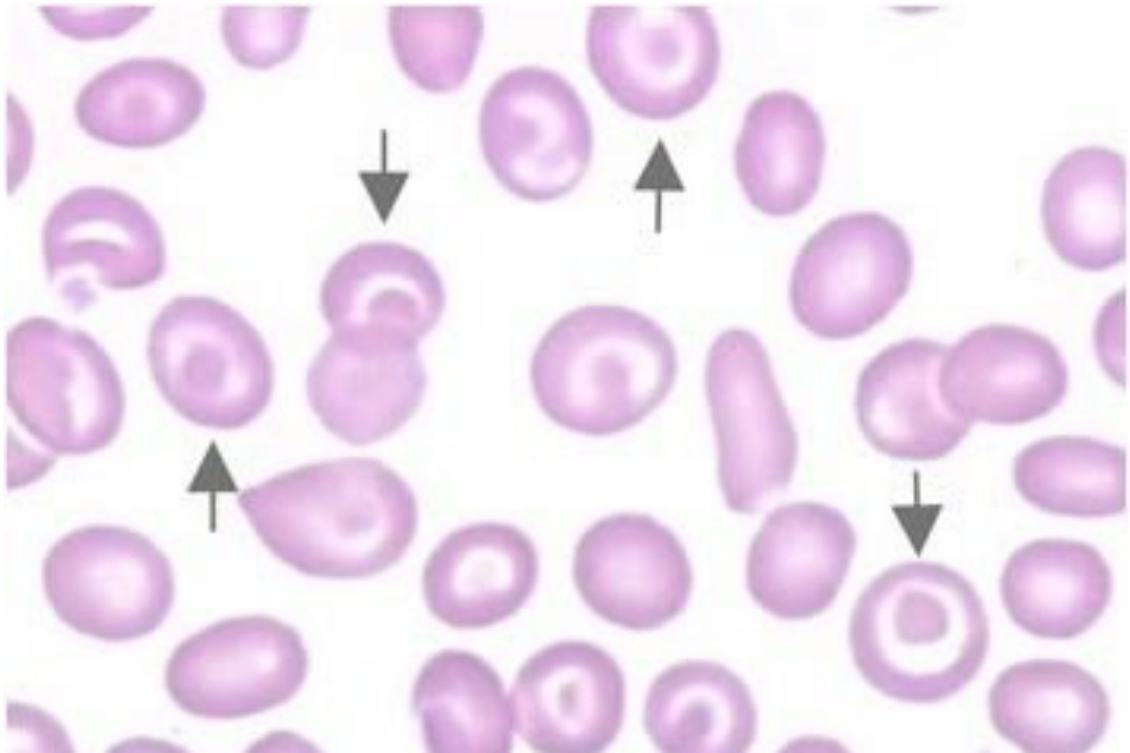
Genotypes	Silent HbH	Deletional HbH	Non-deletional HbH	Transfusion – dependent HbH		HbH hydrops
1°	--/-α --/α^{T}α	--/-α	--/α^{T}α --/αα^T	--/-α	--/α^{T}α --/αα^T α^{T}α/α^{T}α	--/α^{T}α α^{T}α/α^{T}α
2°	β thal	+ β globin variants such as Hb E, Hb C, Hb S, homozygous Hb E, β thal/Hb E etc.		-		-
3°		-		KLF-1		-

* V. Viprakasit et al., *Alpha-thalassemia syndromes: from clinical and molecular diagnosis to bedside management*



Diagnosis





Screening test for Thalassemia

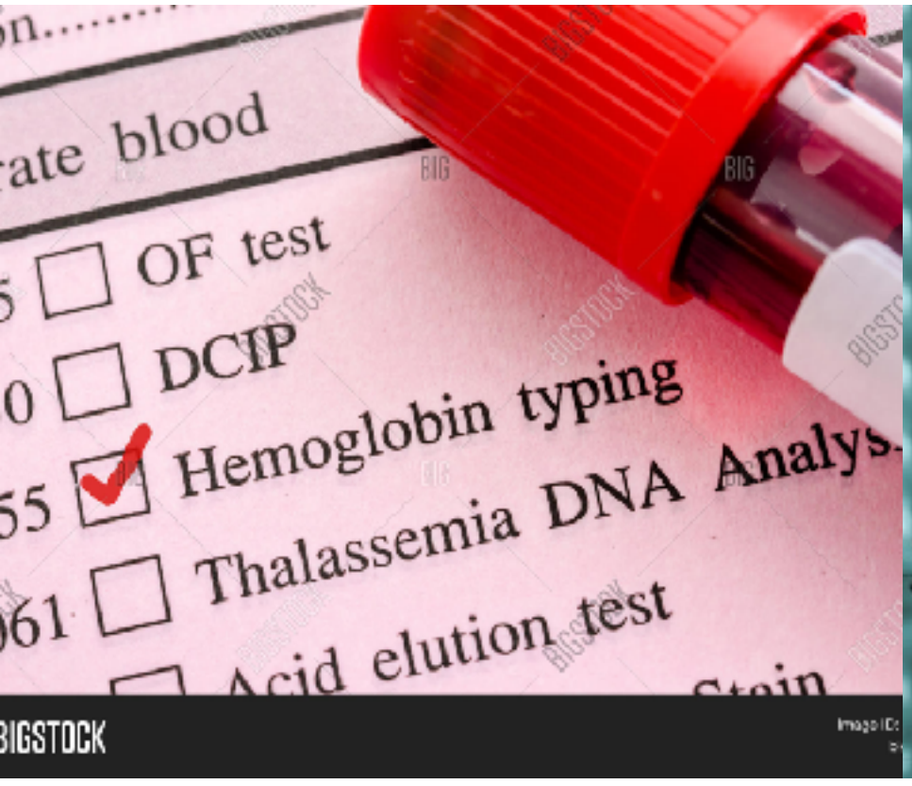
Screening test for Thalassemia *(2 tests for screening)*

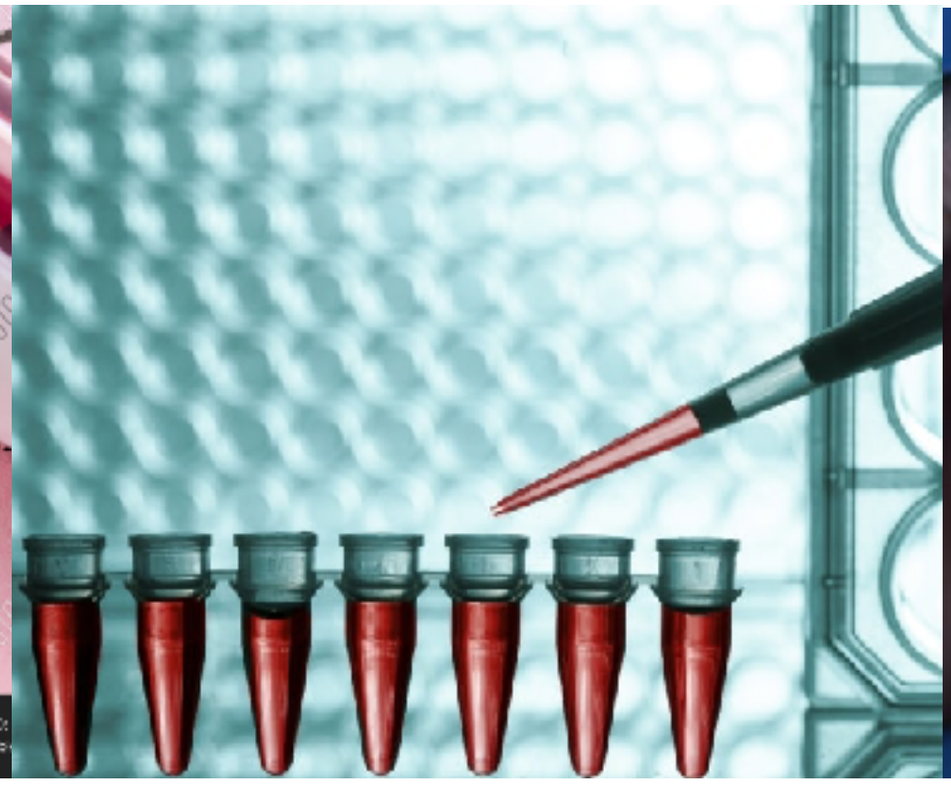
- **Red cell Indices (MCV, MCH)**
- **One tube osmotic fragility test**
- **DCIP - DiChlorophenolIndoPhenol preparation test**
- **Hb E screen**

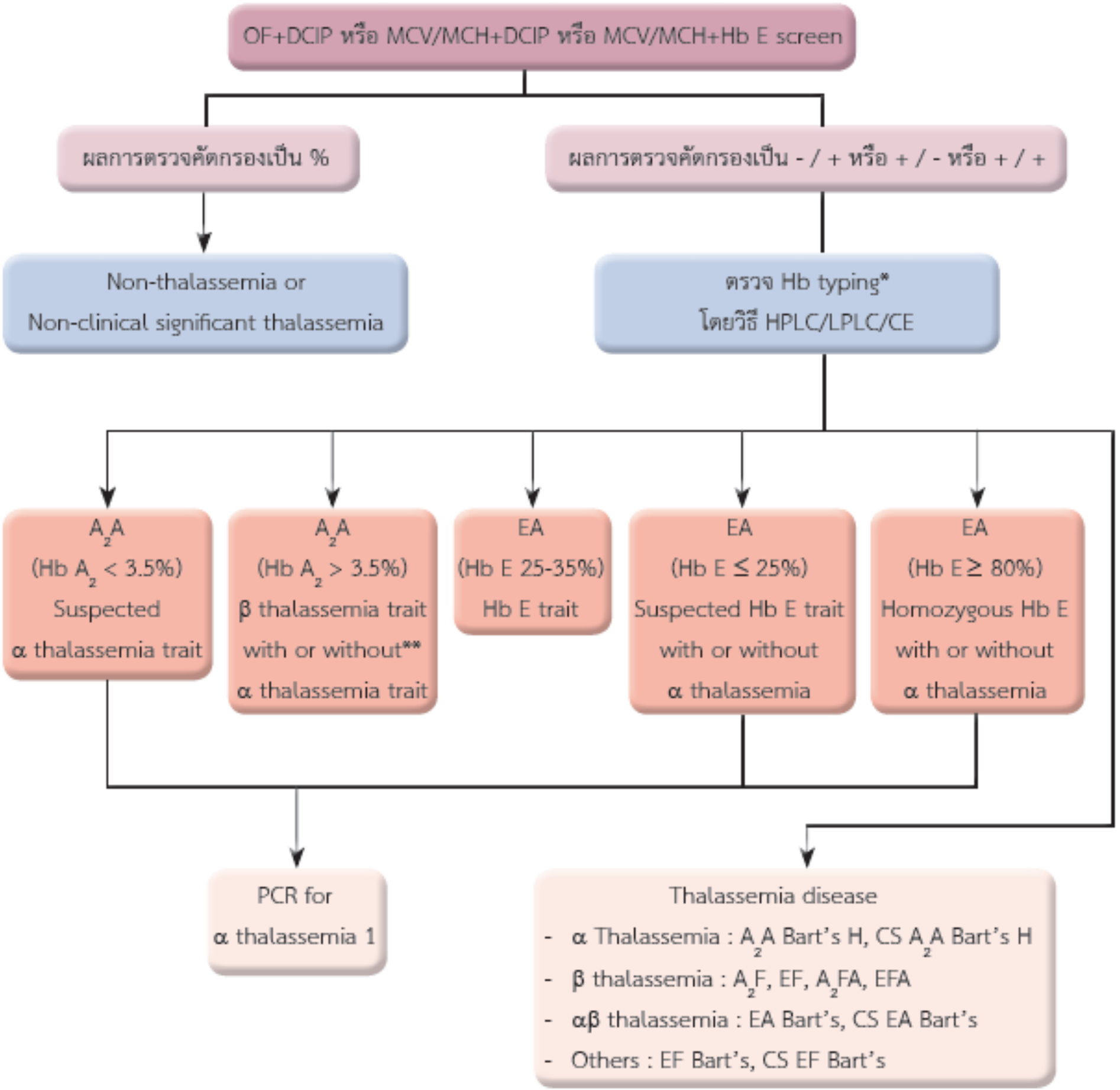
Confirmation test for Thalassemia

Confirmation test for Thalassemia

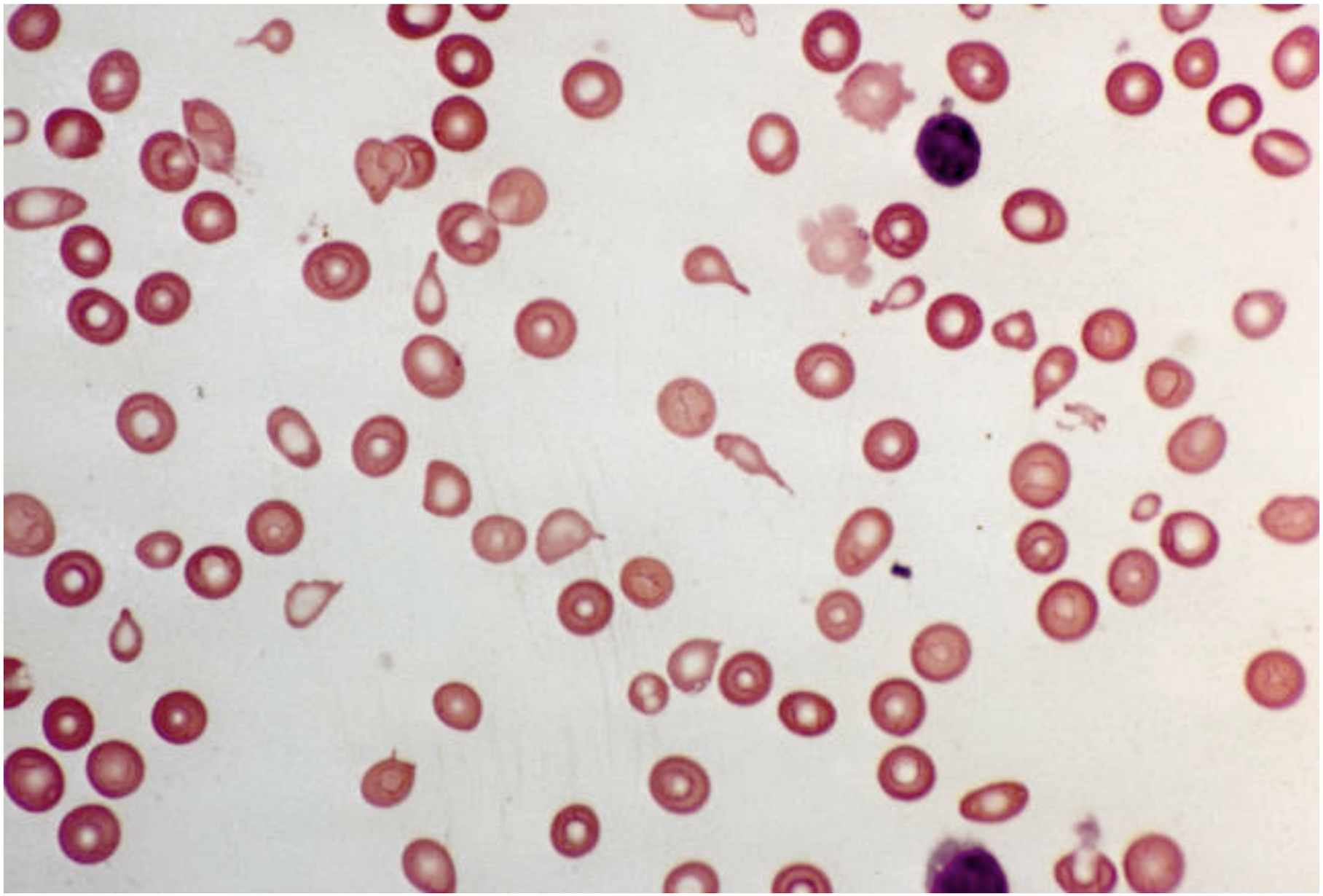
- **Hemoglobin typing**
- **Molecular testing**






OF+DCIP หรือ MCV/MCH+DCIP หรือ MCV/MCH+Hb E screen
ผลการตรวจคัดกรองเป็น %
ผลการตรวจคัดกรองเป็น - / + หรือ + / - หรือ + / +
Non-thalassemia or
Non-clinical significant thalassemia
ตรวจ Hb typing*
โดยวิธี HPLC/LPLC/CE
A2A
(Hb A2 < 3.5%)
Suspected
α thalassemia trait
A2A
(Hb A2 > 3.5%)
β thalassemia trait
with or without**
α thalassemia trait
EA
(Hb E 25-35%)
Hb E trait
EA
(Hb E ≤ 25%)
Suspected Hb E trait
with or without
α thalassemia
EA
(Hb E ≥ 80%)
Homozygous Hb E
with or without
α thalassemia
PCR for
α thalassemia 1
Thalassemia disease
- α Thalassemia : A2A Bart's H, CS A2A Bart's H
- β thalassemia : A2F, EF, A2FA, EFA
- αβ thalassemia : EA Bart's, CS EA Bart's
- Others : EF Bart's, CS EF Bart's

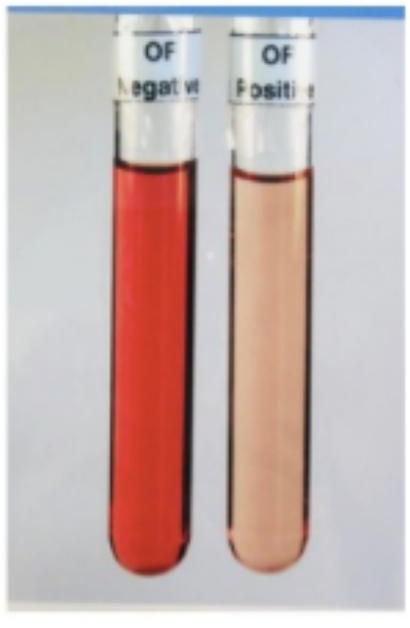
RBC Indices & PBS



One tube osmotic fragility test

ใส่ 0.36% NaCl 5 ml ในหลอดทดลอง
ขนาด 13x100 mm

ปิเปต EDTA blood 20 µl ใส่ในหลอดทดลอง ผสมเลือดกับน้ำยา
โดย mix หลอดไปมาเบาๆ ตั้งทิ้งไว้ที่อุณหภูมิห้อง 5 นาที อ่านผล



Negative : สารละลายใสสีแดง ไม่เป็นธาลัสซีเมีย หรืออาจเป็นธาลัสซีเมียชนิดไม่รุนแรง เช่น α-Thalassemia 2 , Hb Constant Spring

Positive : สารละลายขุ่น อาจเป็น α-Thalassemia หรือ β-Thalassemia เมื่อผลตรวจเป็น Positive ต้องส่งตัวอย่างเลือดตรวจ Hb typing และ DNA analysis ต่อไปเพื่อจะได้ทราบว่าเป็นธาลัสซีเมียชนิดไหน

DCIP - DiChlorophenolIndoPhenol test

วิธีทำ

นำ EDTA blood ปั่นในเครื่อง centrifuge ที่ 1000 rpm/min 10 นาที และทิ้งส่วนที่เป็นพลาสมา

↓

เติม DCIP reagent 5 ml ลงในหลอดทดลองขนาด 16x100 mm

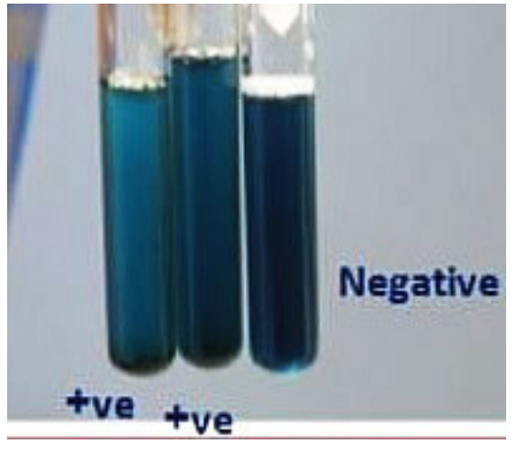
↓

นำ Pack Red cell 20 µl ลงในหลอดที่มีน้ำยา DCIP ผสมให้เข้ากัน โดย mix หลอดทดลองไปมาเบาๆ

↓

นำหลอดทดลองไป incubate ที่ 37°c นาน 1 ชั่วโมง และอ่านผลการทดสอบโดยดูการตกตะกอนที่เกิดขึ้นด้วยตาเปล่า

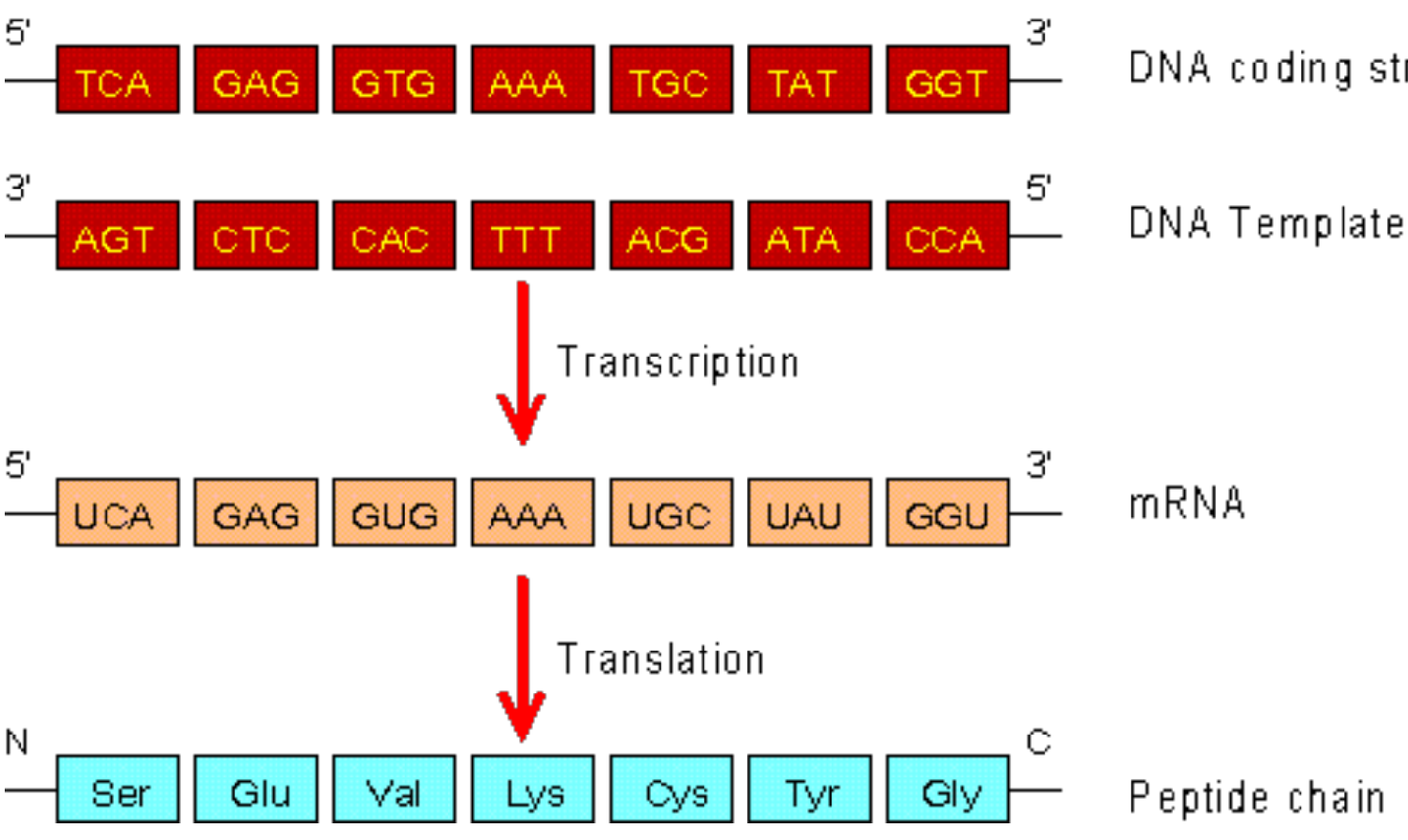
↓



Negative : สารละลายใส

Positive : สารละลายตกตะกอน อาจเป็นผู้ที่มี Hb E ซึ่งพบได้ทั้งพาหะ Hb E, Homozygous Hb E หรือ α-Thalassemia หรือ β-Thalassemia ที่มี Hb E ร่วมด้วย Hb H, Hb Bart's อาจทำให้ขุ่นเล็กน้อยได้

Confirmation test for Thalassemia



Genetic testing

Hb typing

Molecular testing for Thalassemia

Genetic testing

α-thal → Deletion

Multiplex gap-PCR

β-thal → Point mutation

Direct DNA sequencing of β-globin gene (3 exons)

Multiplex amplification refractory mutation system (Multiplex-ARMS)

Treatment

Curative treatment

- **Hematopoietic stem cell transplantation**
- **Gene therapy**

Standard treatment

- **Education and genetic counseling**
- **Regular blood transfusion to maintain Hb 13-14 g/dL post transfusion and 9-10 at all time**
- **Supportive treatment : Folic acid, MTV**
- **Iron chelation**

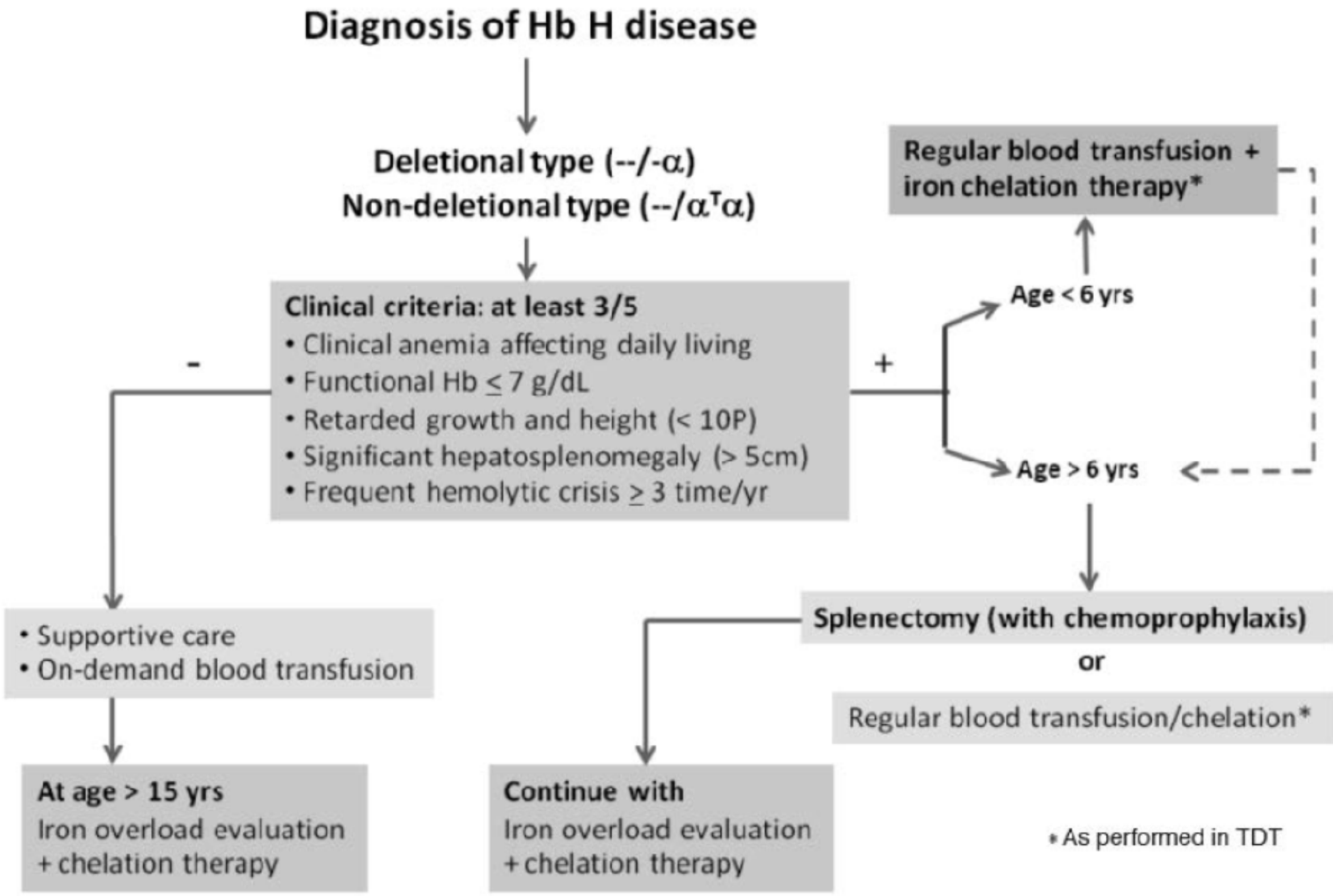
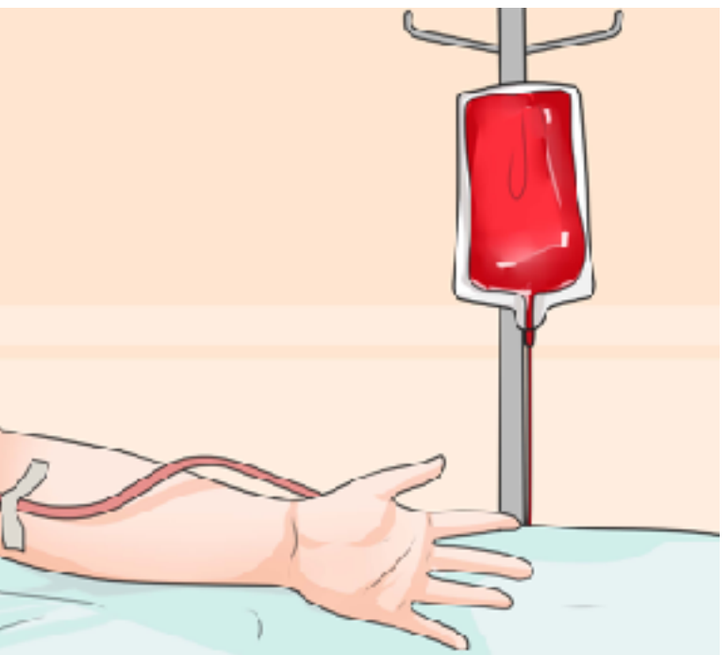


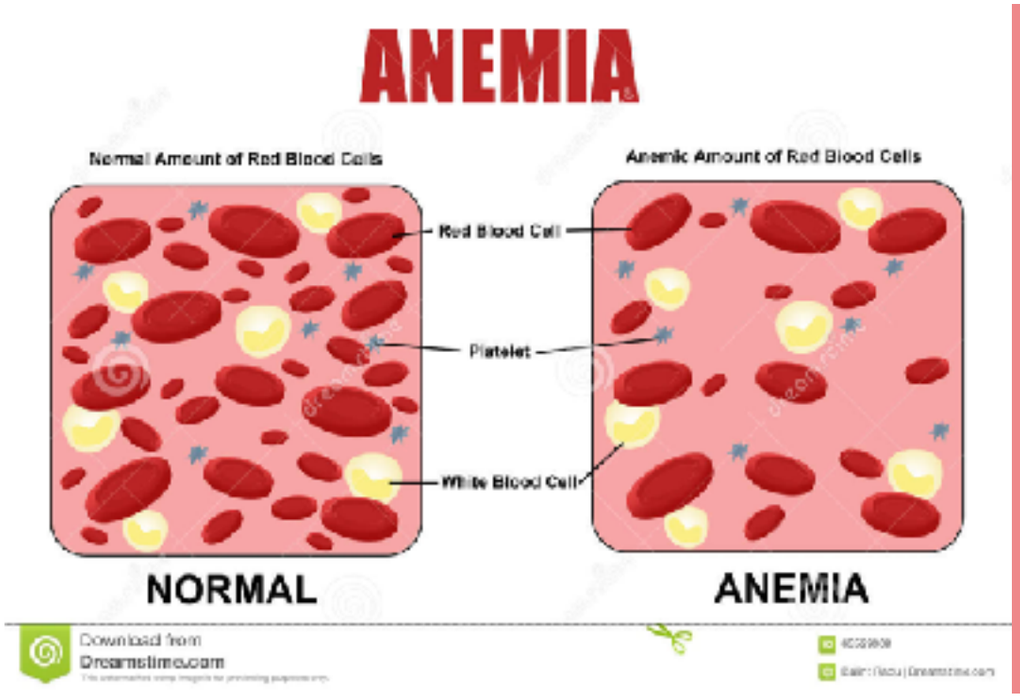
Figure 4. Management guideline in a new patient with Hb H disease. Diagnosis of Hb H disease requires a comprehensive hematology, hemoglobin and DNA analyses. [+] = with ≥3 and [-] = with <3 out of 5 criteria.

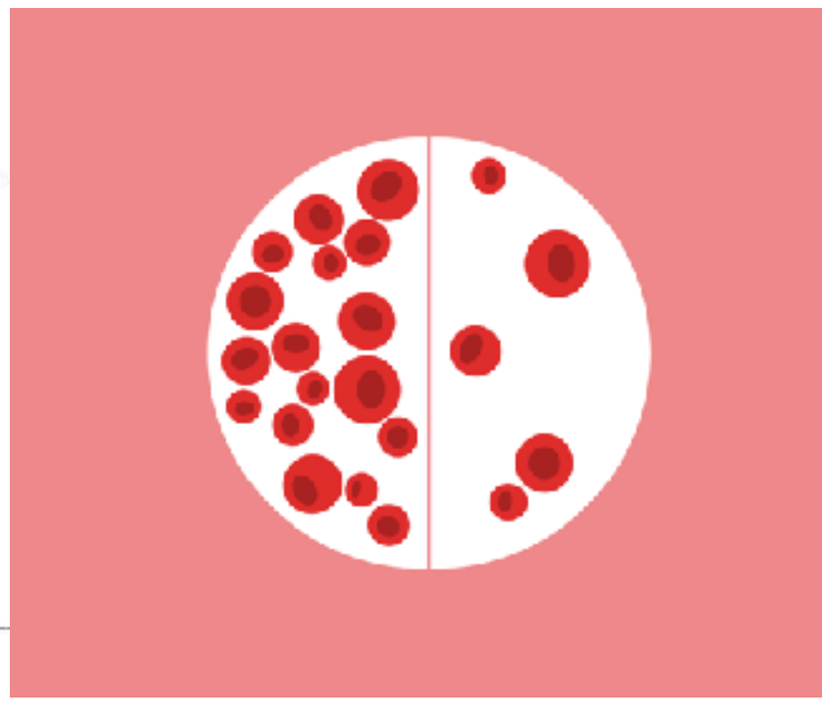
* V. Viprakasit et al., *Alpha-thalassemia syndromes: from clinical and molecular diagnosis to bedside management*

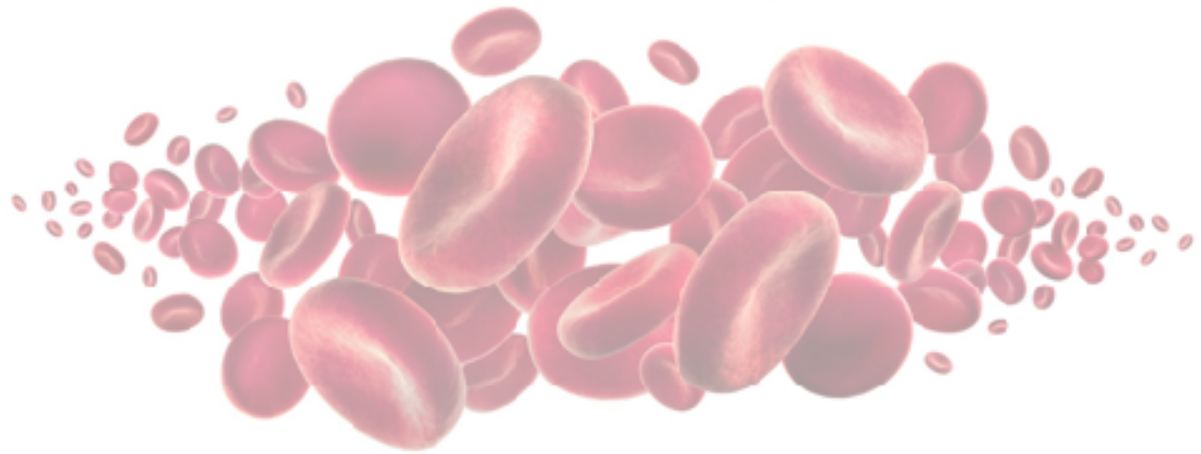
Learning objectives

- **Approach to anemia**
- **What you should know about “70 baht” CBC**
- **Hemoglobin typing**
- **Thalassemia**
- **Rational use of blood component - The concept**





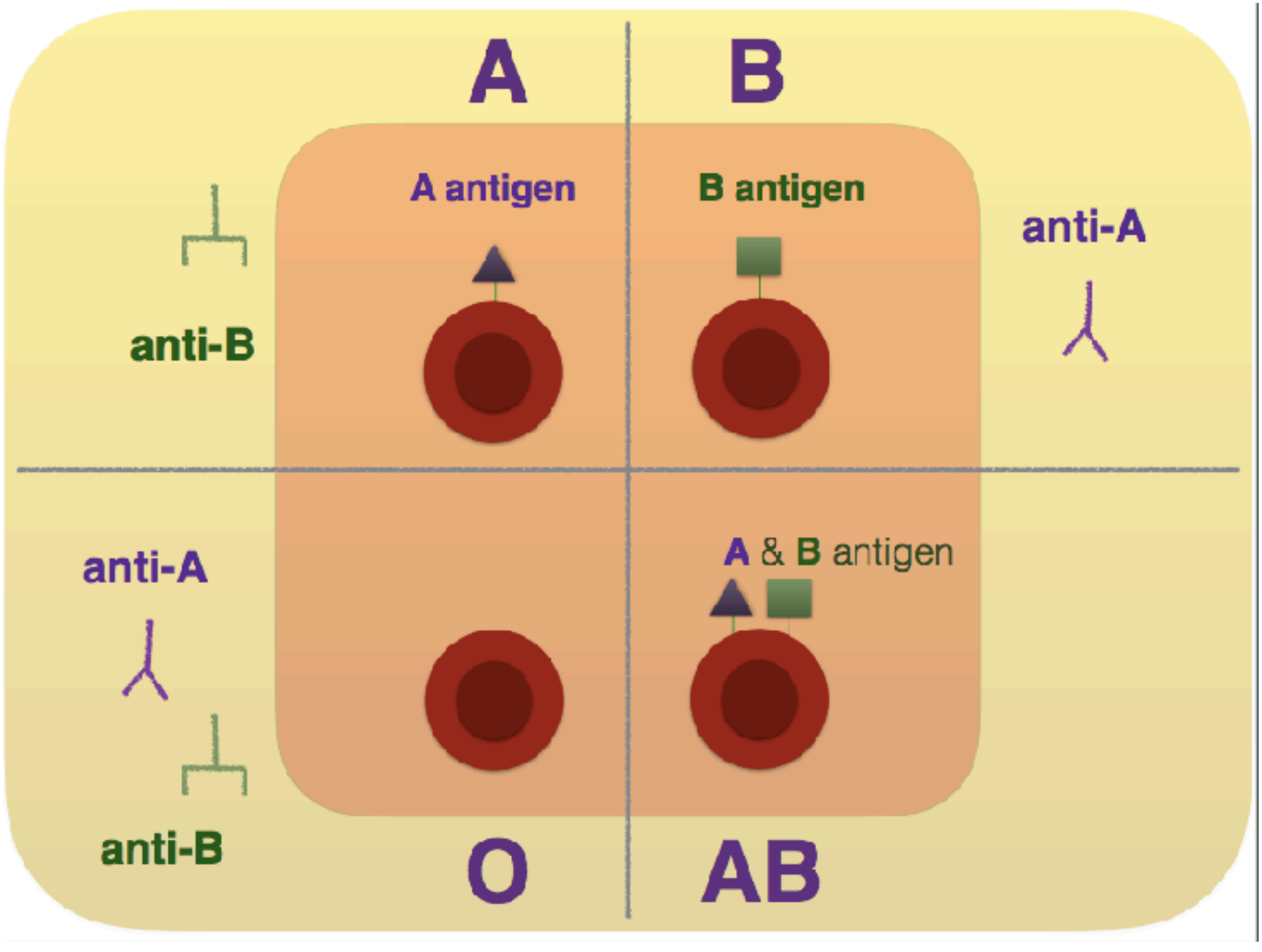




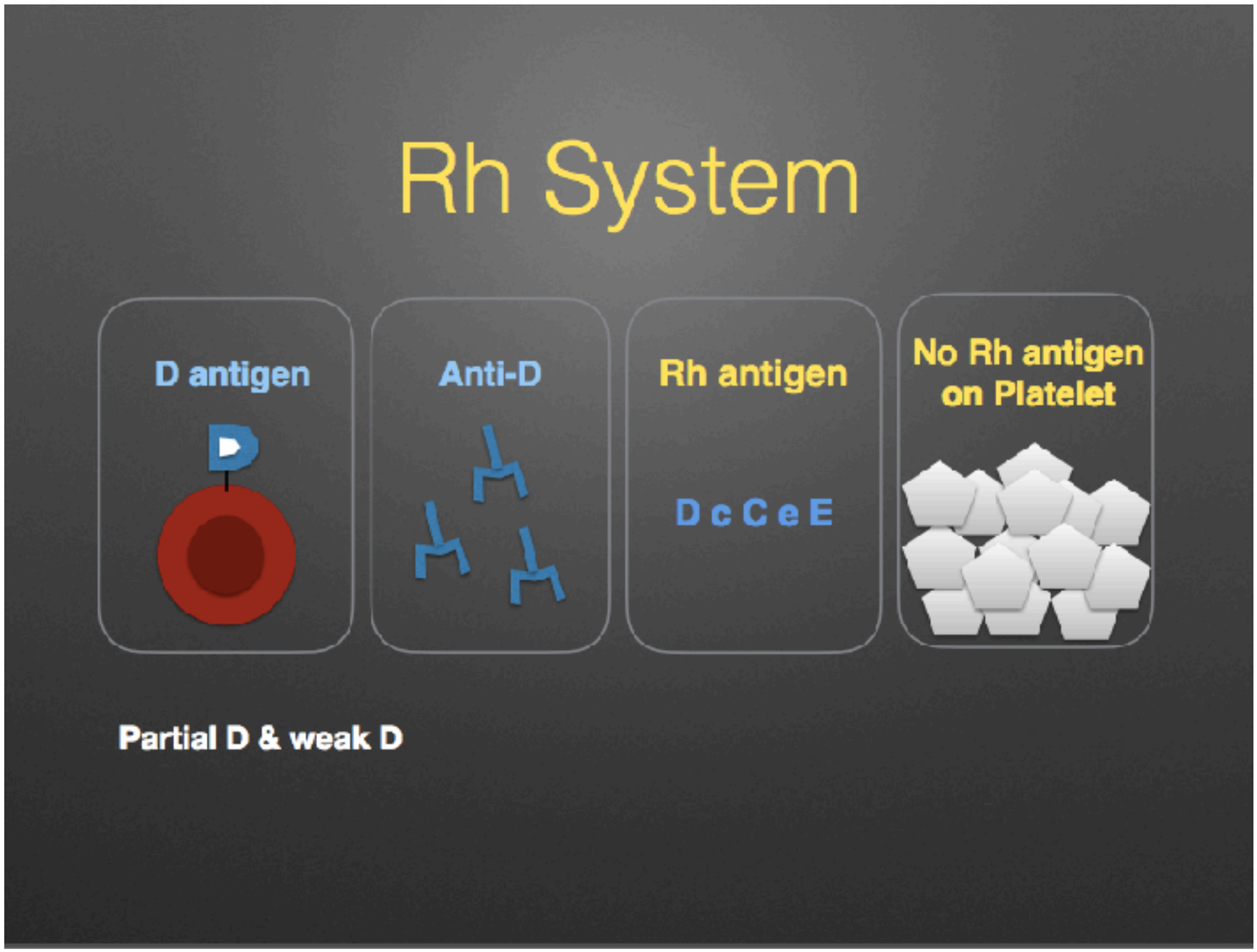
Rational use of blood component
“The concept”

F2.Apichat Photi-A, MD.
Hematology /Oncology unit,
Department of Paediatrics
Phramongkutklao Military Hospital
Bangkok, Thailand

ABO Blood group



Rh D Blood group

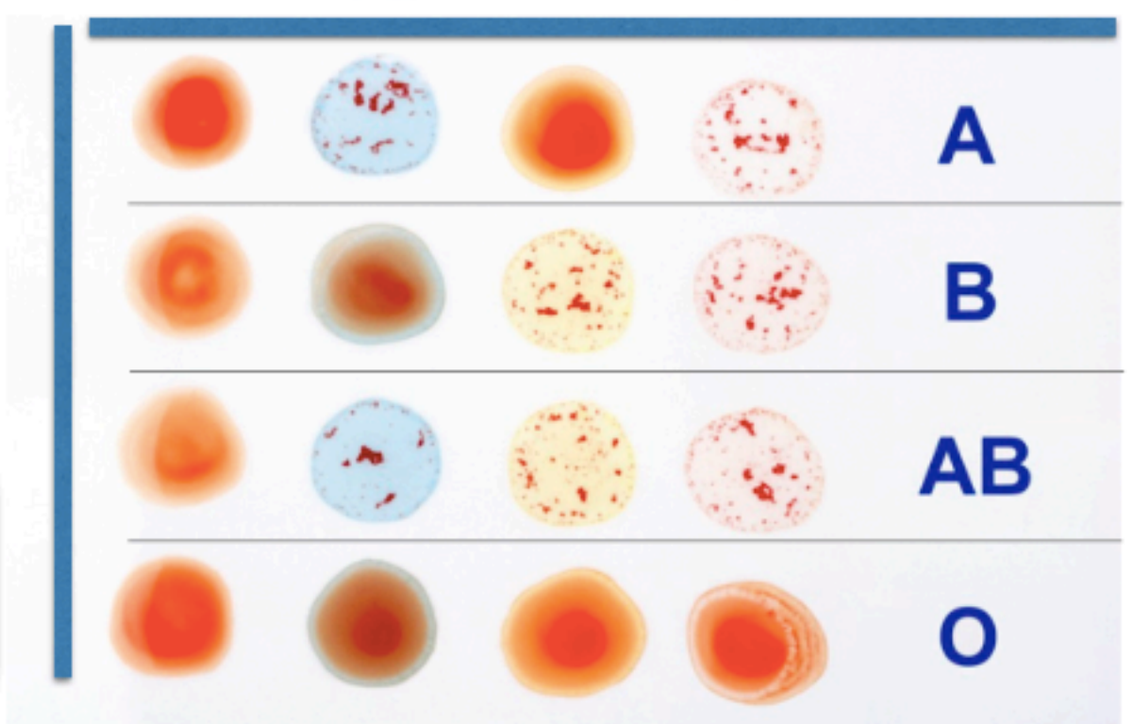


Pretransfusion testing

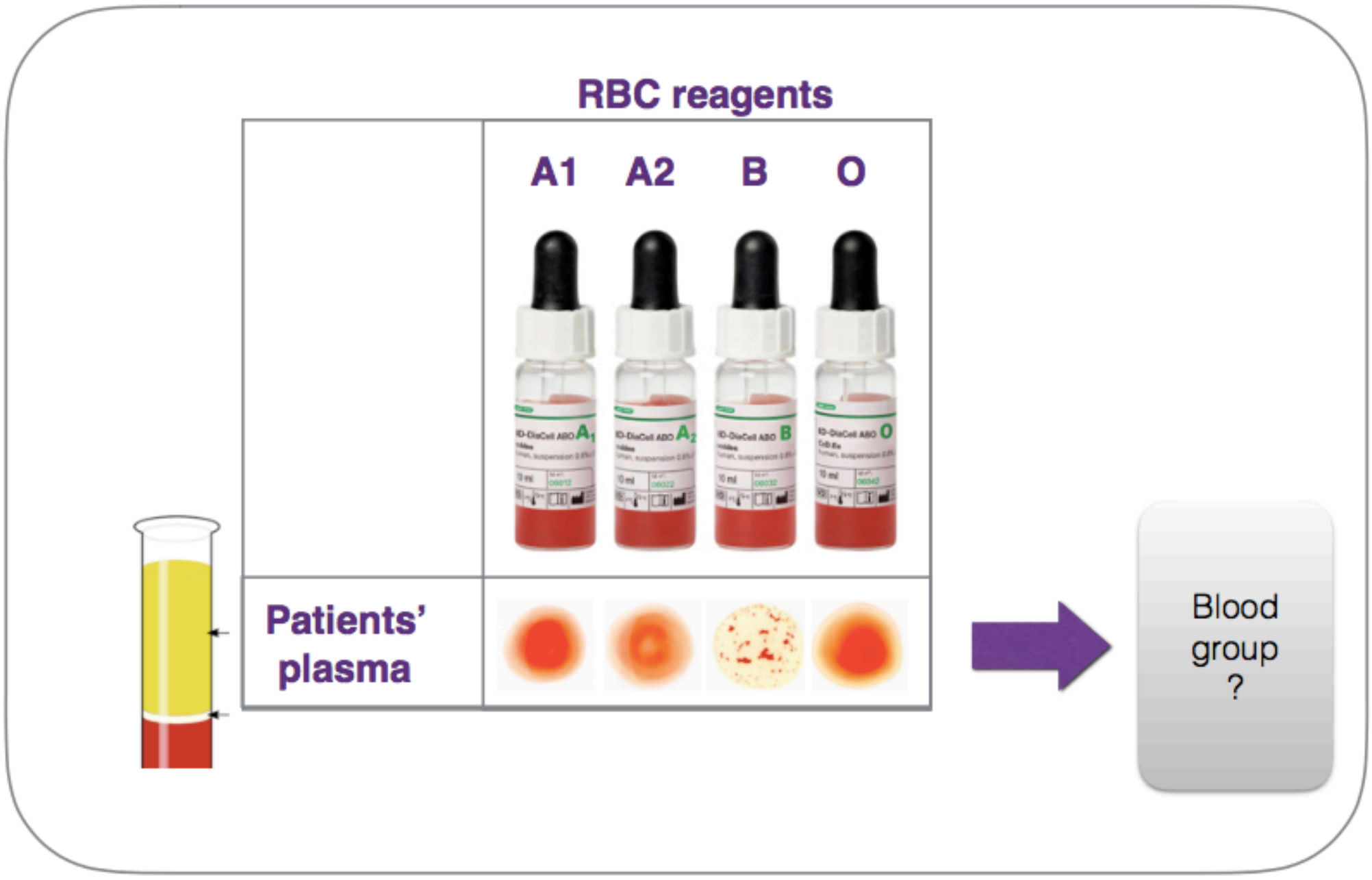
- Type and Screen
  - ABO/Rh system
    - Forward typing: cell grouping
    - Reverse typing: serum grouping
  - Others: Screening for RBC alloantibody
- Crossmatching

Cell grouping

Patient's Blood (RBC)



Serum grouping

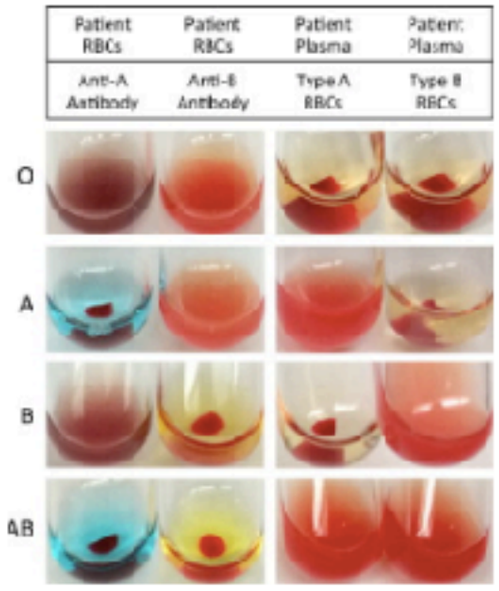


Interpretation

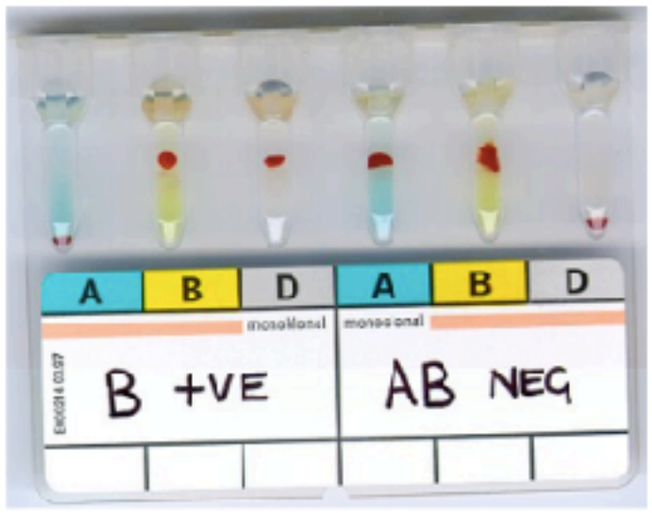
Forward & reverse ABO blood grouping

	Reaction of Cells Tested With		Reaction of Serum Tested Against		ABO Group
	Anti-A	Anti-B	A_1 Cells	B Cells	
1	0	0	+	+	O
2	+	0	0	+	A
3	0	+	+	0	B
4	+	+	0	0	AB

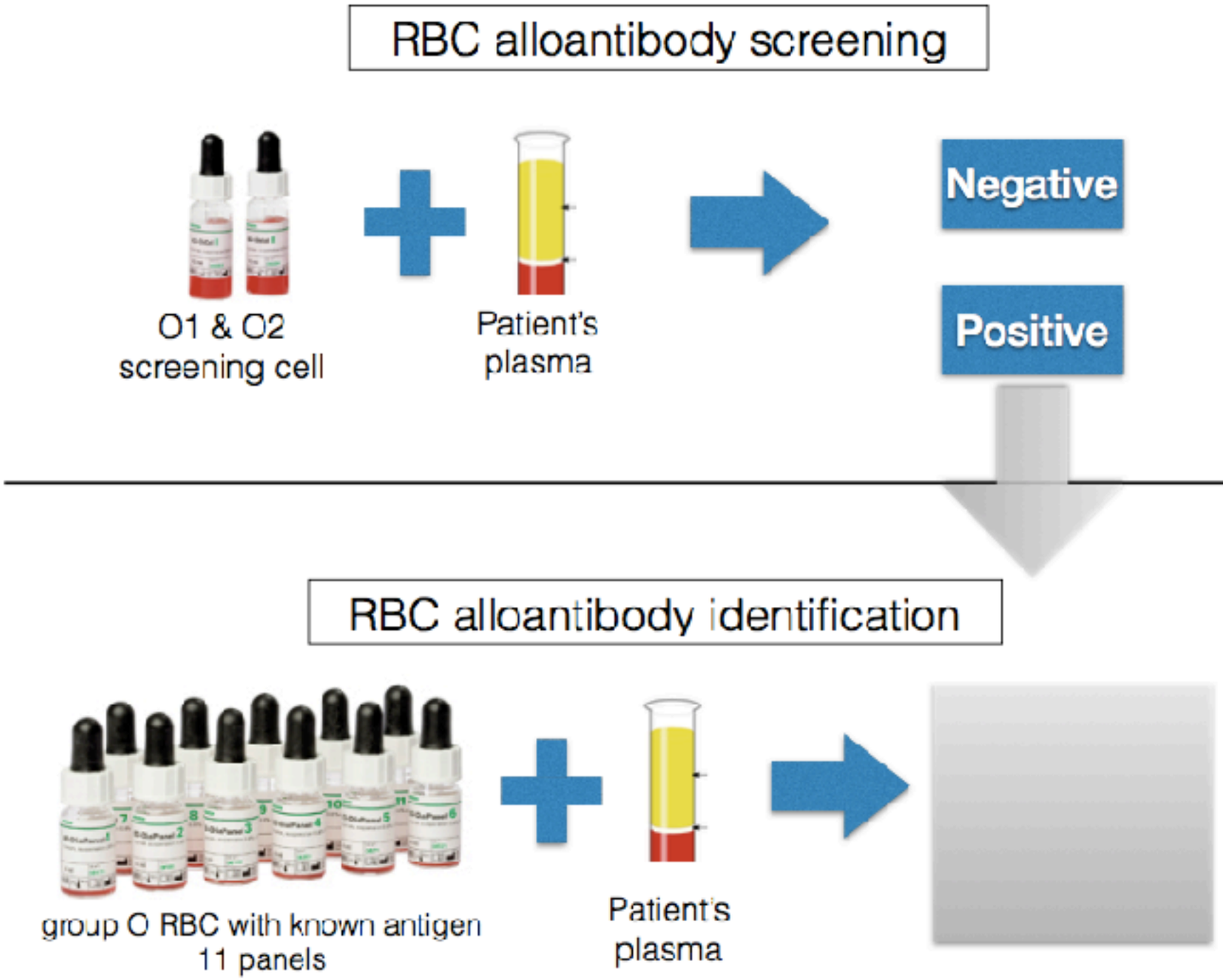
Tube method



Gel method



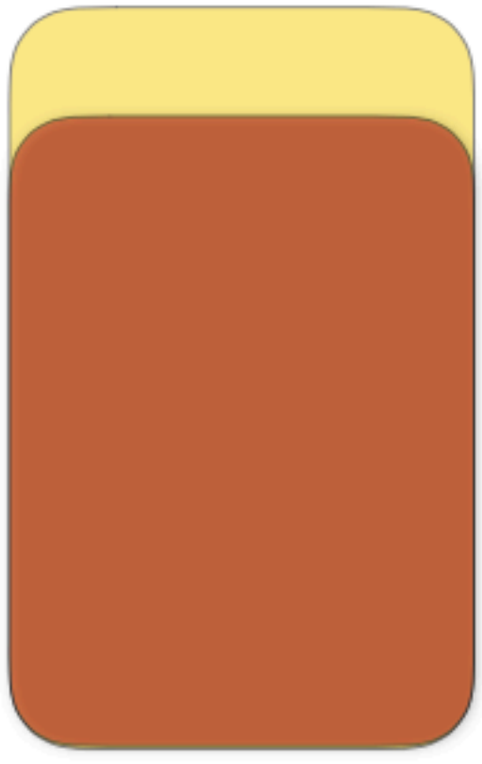
RBC Alloantibody screening



Blood components

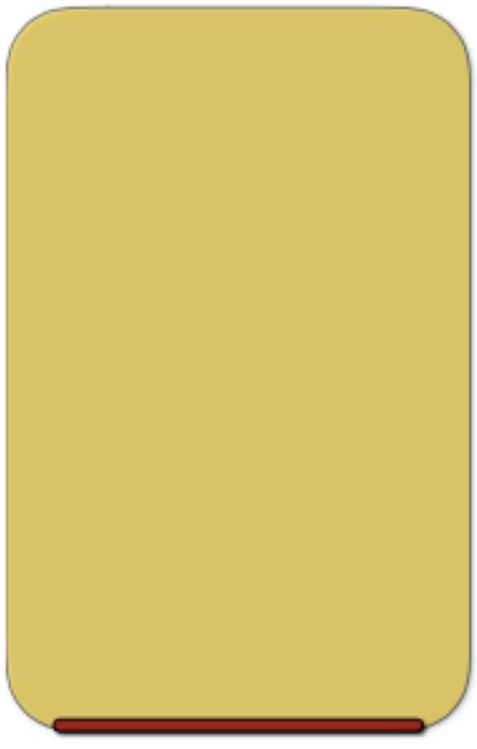
PRC

- 250-300 ml
- Hct ~ 70 - 80%
- Store at 4'C
- up to 42 days



FFP

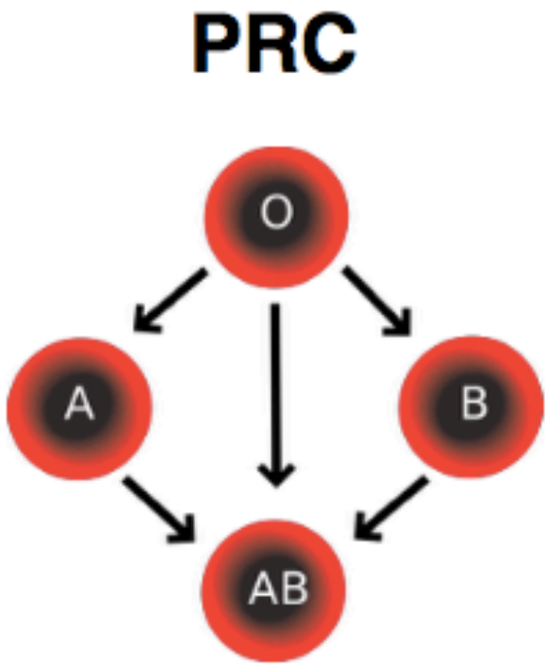
- Frozen within 8 hrs of collection
- 200-250 ml
- Stored at -18'C
- Up to 1 year



Platelet

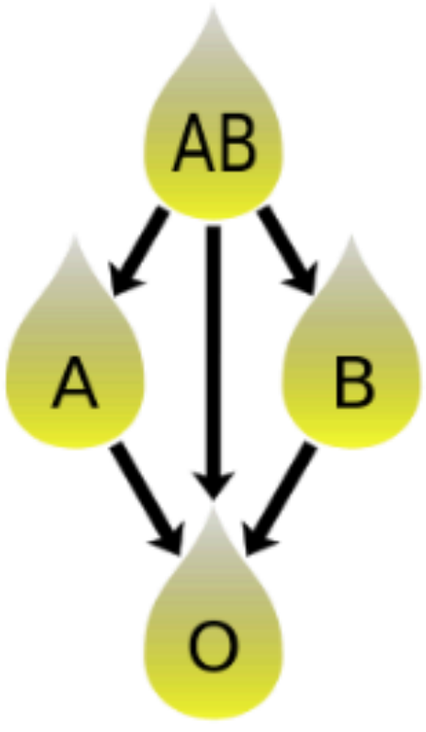
- Plt conc ~ 50 ml/unit
- Plt conc 4-6 u
- raise plt cout ~ 20,000-30,000 /cu.mm.
- Plt express ABO and HLA class I antigen
- No Rh or HLA class II antigens

RBC compatibility



Recipient	Donor							
	O-	O+	A-	A+	B-	B+	AB-	AB+
O-	✓	✗	✗	✗	✗	✗	✗	✗
O+	✓	✓	✗	✗	✗	✗	✗	✗
A-	✓	✗	✓	✗	✗	✗	✗	✗
A+	✓	✓	✓	✓	✗	✗	✗	✗
B-	✓	✗	✗	✗	✓	✗	✗	✗
B+	✓	✓	✗	✗	✓	✓	✗	✗
AB-	✓	✗	✓	✗	✓	✗	✓	✗
AB+	✓	✓	✓	✓	✓	✓	✓	✓

Plasma compatibility

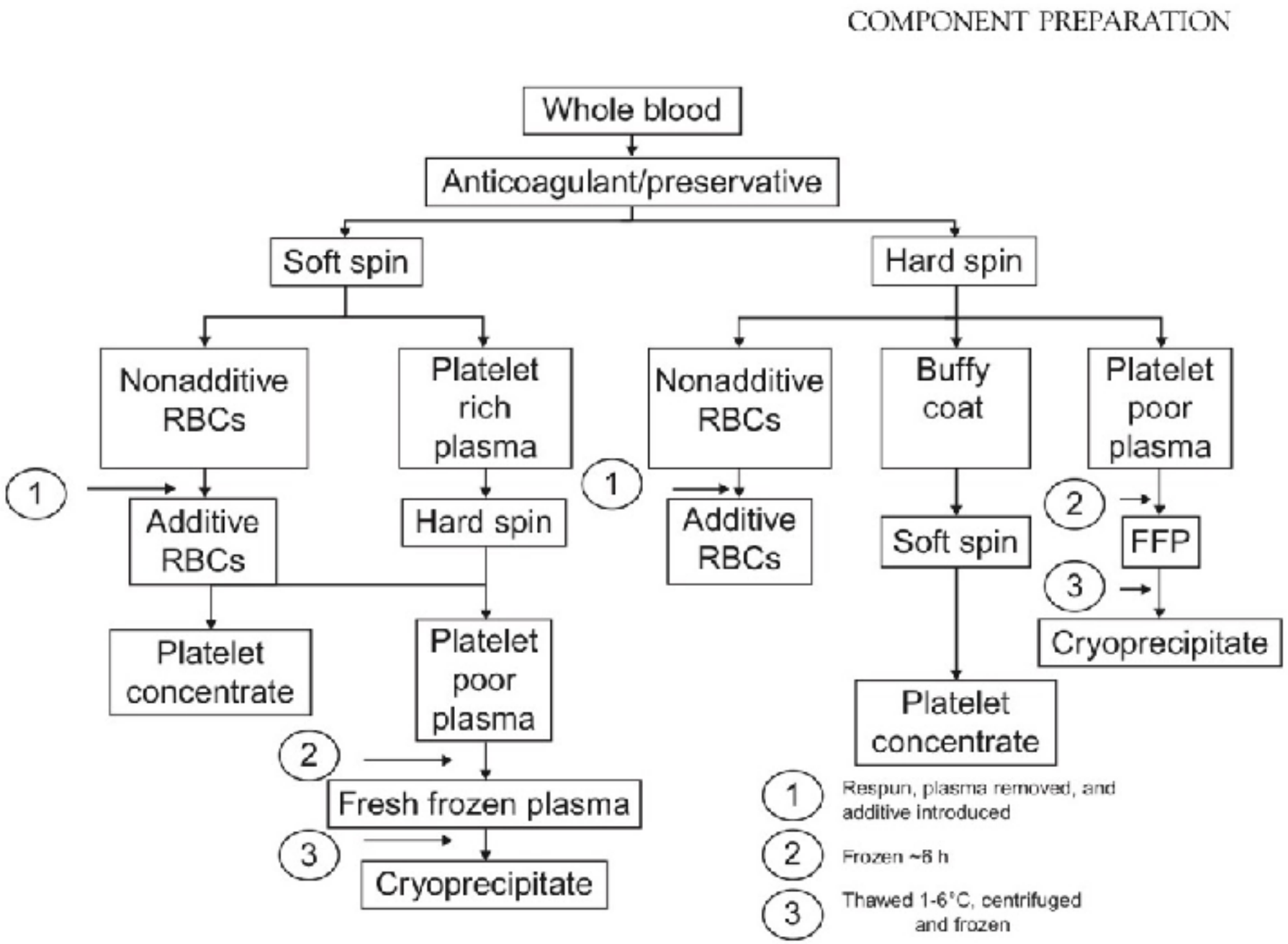


Plasma compatibility table

Recipient	Donor[1]			
	O	A	B	AB
O	✓	✓	✓	✓
A		✓		✓
B			✓	✓
AB				✓

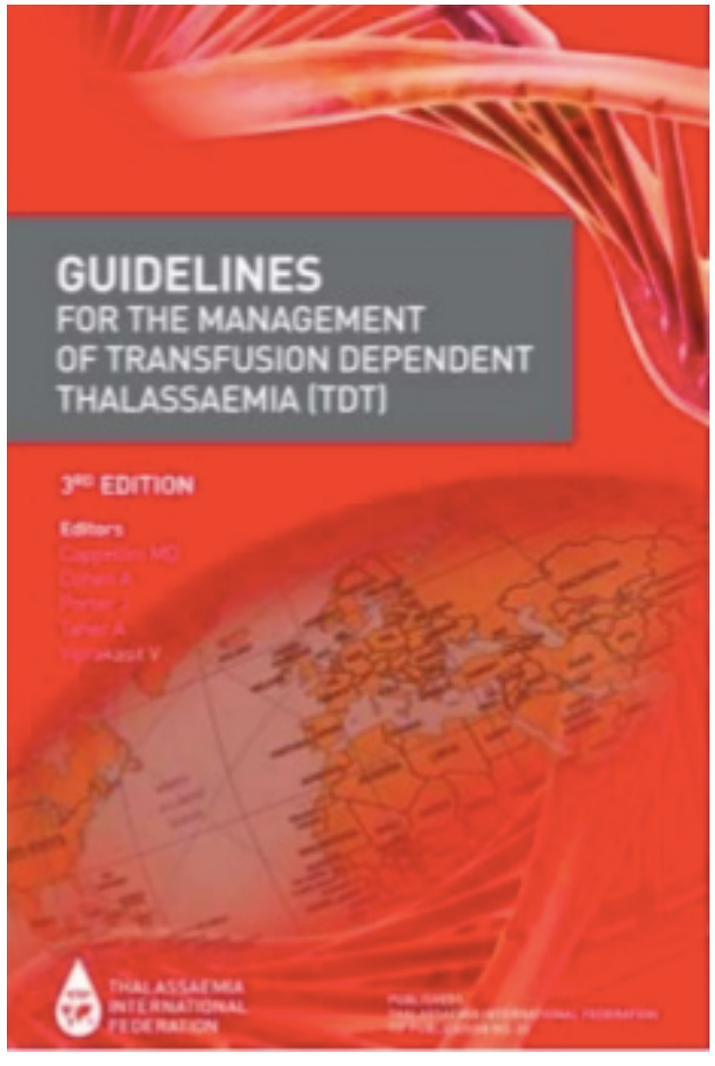
	You Can Give Red Cells To:	You Can Give Platelets To:	You Can Give Plasma To:
O–	O– O+ A– A+ B– B+ AB– AB+	O– O+	O– O+
O+	O+ A+ B+ AB+	O+	O– O+
A–	A– A+ AB– AB+	O– A– O+ A+	O– A– O+ A+
A+	A+ AB+	O+ A+	O– A– O+ A+
B–	B– B+ AB– AB+	O– B– O+ B+	O– B– O+ B+
B+	B+ AB+	O+ B+	O– B– O+ B+
AB–	AB– AB+	O– A– B– AB– O+ A+ B+ AB+	O– A– B– AB– O+ A+ B+ AB+
AB+	AB+	O+ A+ B+ AB+	O– A– B– AB– O+ A+ B+ AB+

Blood collection



Reference


แนวทางการวินิจฉัยและการรักษา
โรคโลหิตจางธาลัสซีเมีย
Clinical practice guidelines for
diagnosis and management of thalassaemia syndromes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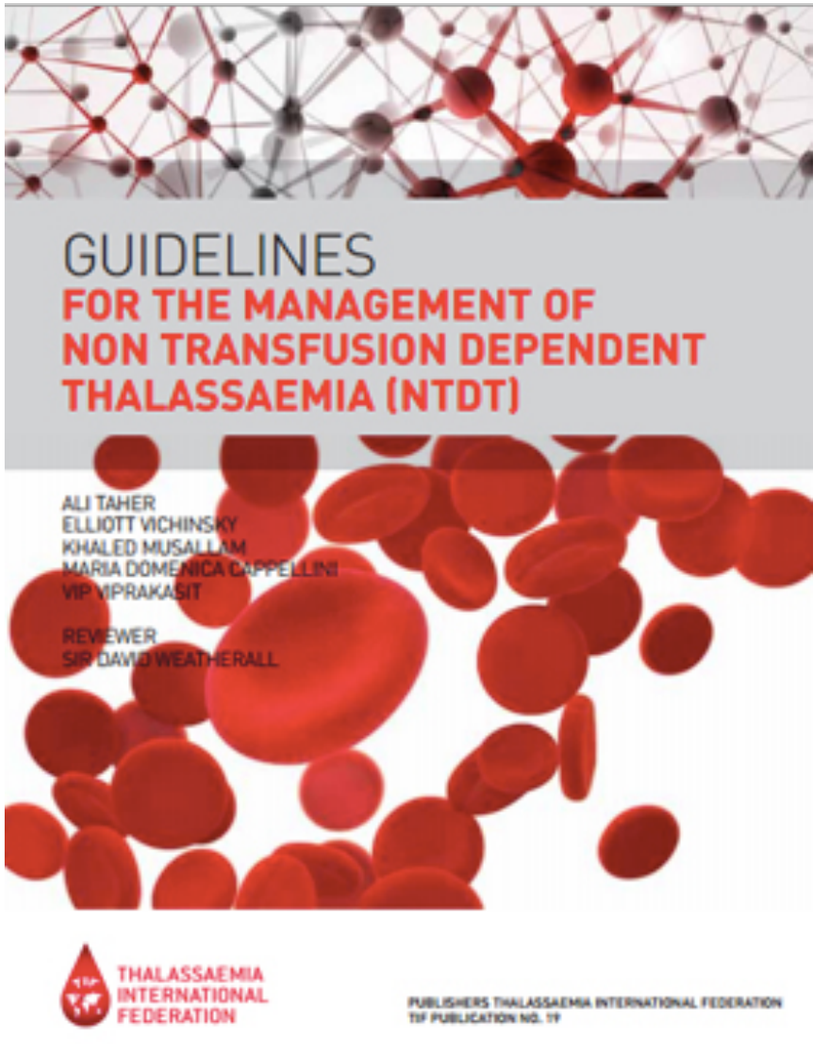
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