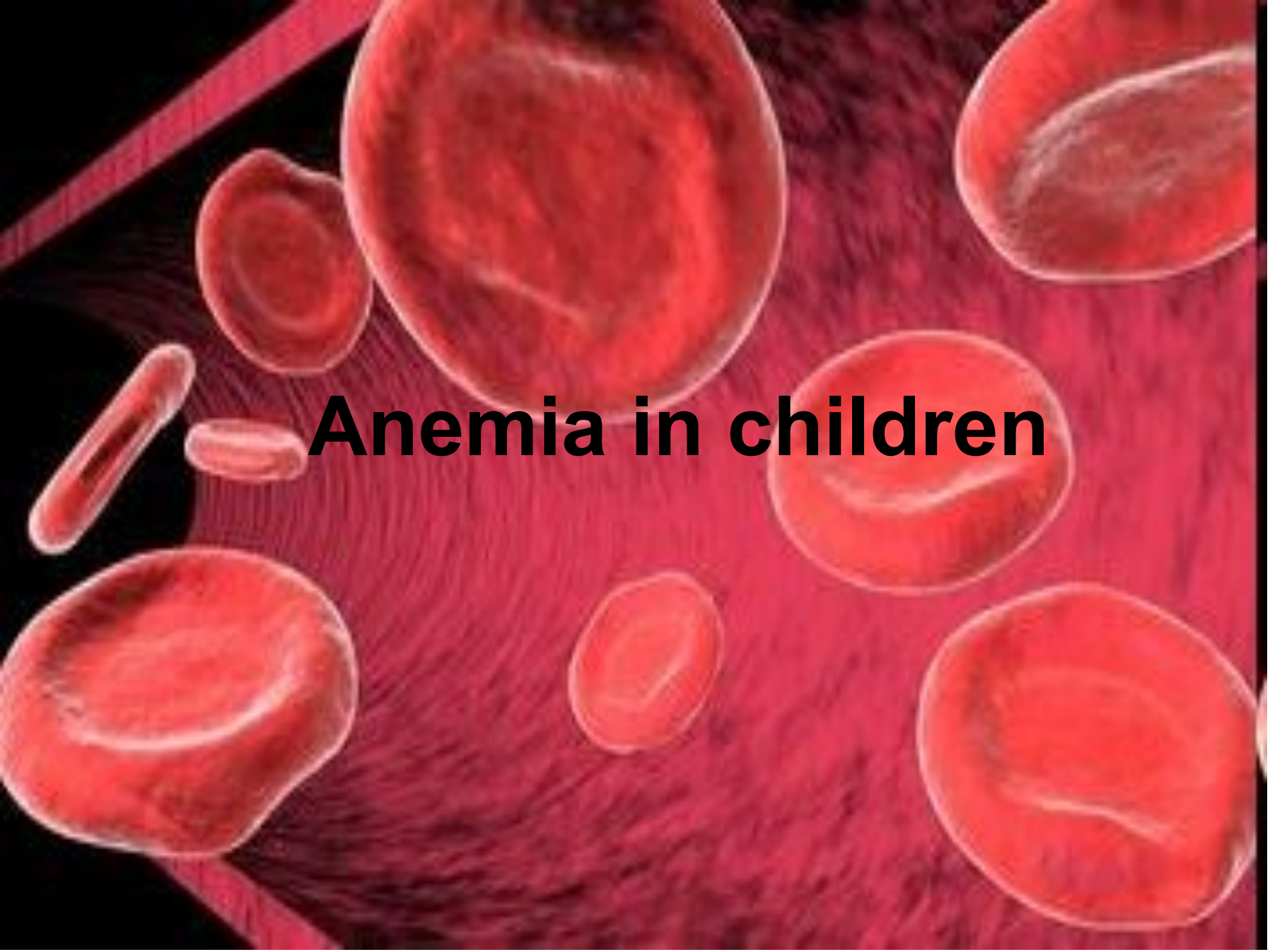
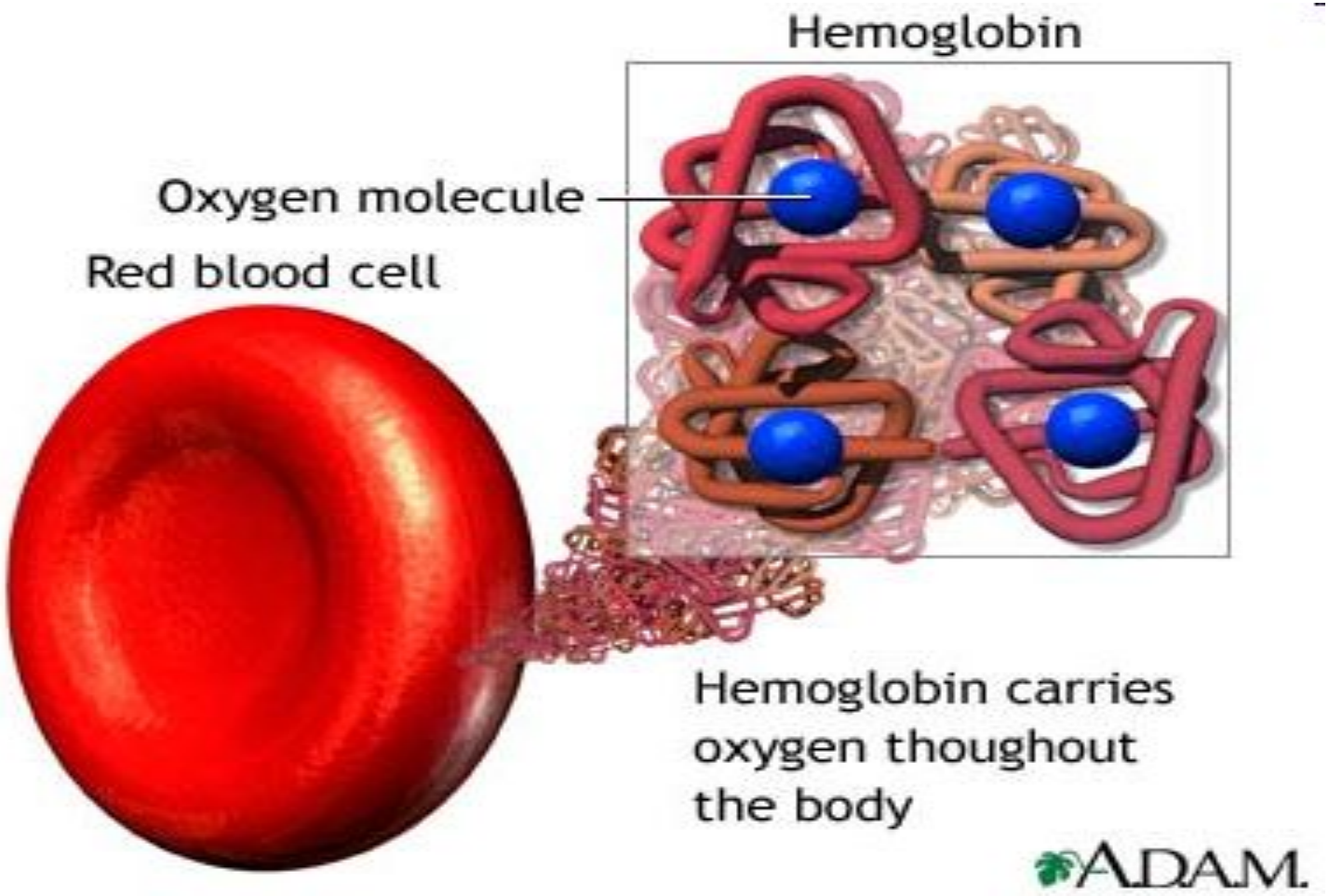
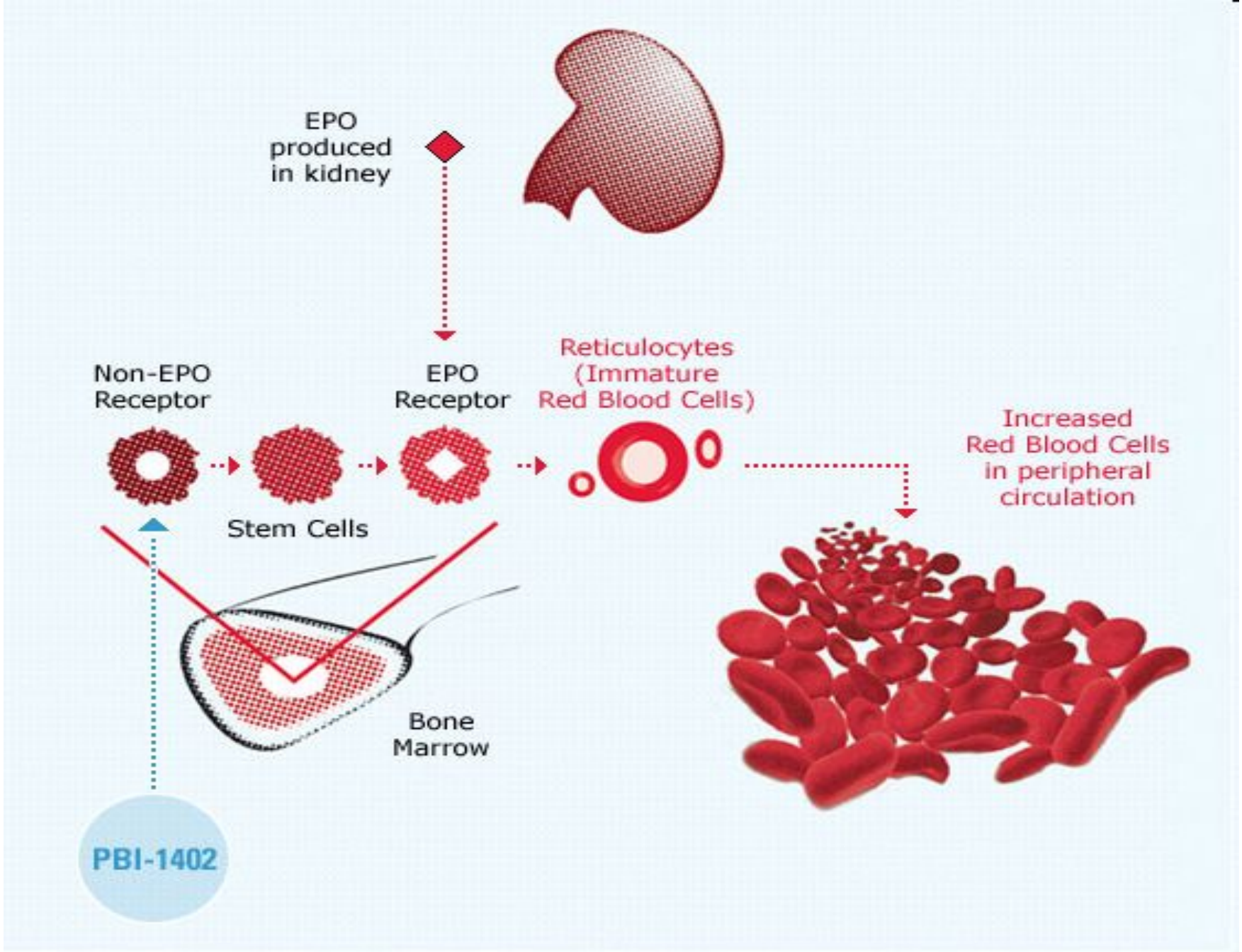
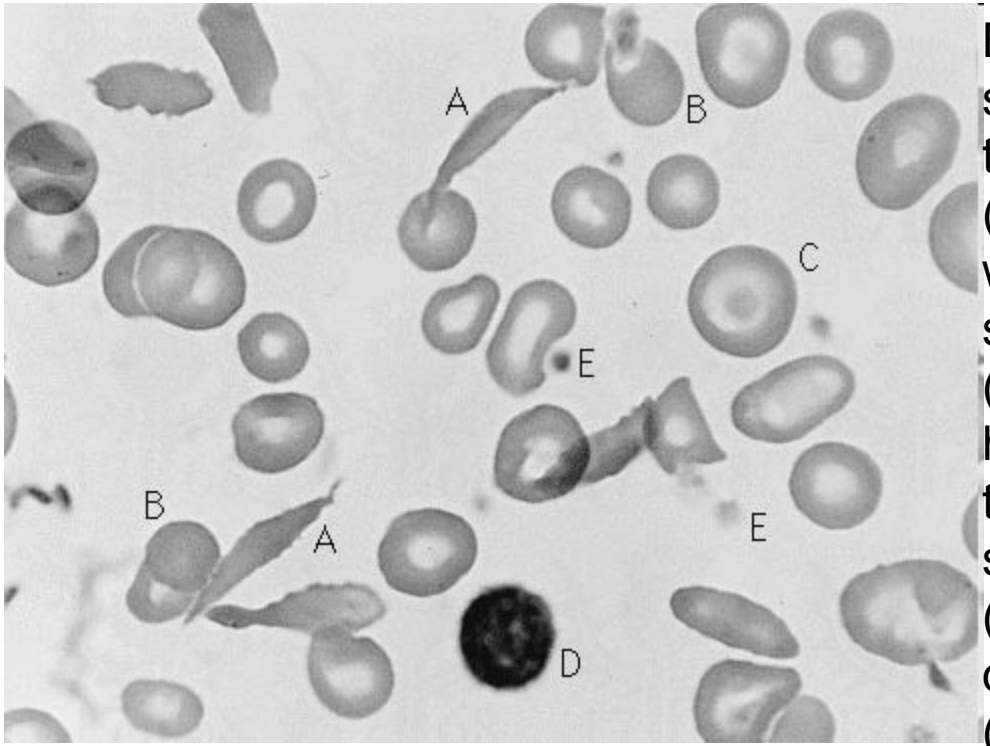

Anemia in children


Hemoglobin
Oxygen molecule
Red blood cell
Hemoglobin carries oxygen thoughout the body
A.D.A.M.


EPO
produced
in kidney
Non-EPO
Receptor
EPO
Receptor
Reticulocytes
(Immature
Red Blood Cells)
Increased
Red Blood Cells
in peripheral
circulation
Stem Cells
Bone
Marrow
PBI-1402



Blood smear in which the red cells show variation in size and shape typical of sickle-cell anemia.
(A) Long, thin, deeply stained cells with pointed ends are irreversibly sickled.
(B) Small, round, dense cells are hyperchromic because a part of the membrane is lost during sickling.
(C) Target cell with a concentration of hemoglobin on its centre.
(D) Lymphocyte.
(E) Platelets.

AVERAGE NORMAL BLOOD VALUES AT DIFFERENT AGE GROUPS

AGE	Hb (gm %)	RBC (m/L)	HCT %	MCV (cu.mm)	MCH (pg)	MCHC %	Retic %
1 day	18.0	5.14	61	119	36.0	31.6	32
4 weeks	14.2	4.0	43	106	35.5	33.5	0.6
1 year	11.6	4.6	35	77	25.0	33.0	0.9
10-12 years	13.0	4.8	39	80	27.0	33.0	1.0
Adult-Men	16.0	5.4	47	87	29.0	34.0	1.0

CLASSIFICATION AND AETIOLOGY OF ANEMIA :

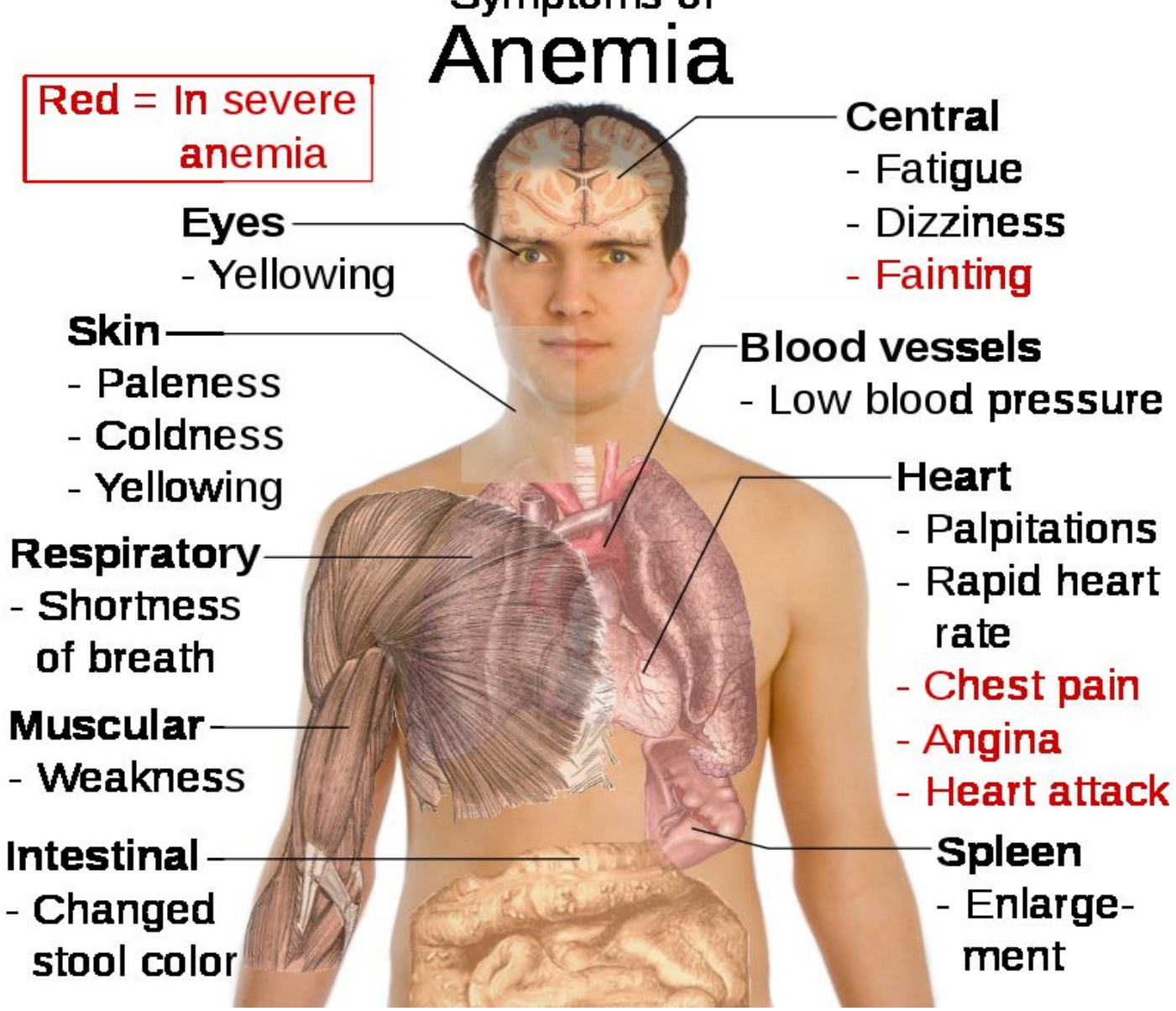
There are four basic causes of anemia - loss, destruction, sequestration and hypoproduction.

Anemia can be further classified by
RBC size - micro, normo, and macrocytic anemia.
RBC shape - e.g. Sickle cell.
Etiology

Blood loss : Acute Chronic

Decreased iron assimilation : Nutritional deficiency
Hypoplastic or aplastic anemia
Bone marrow infiltration like leukemia & other malignancies, myelodysplastic syndrome
Dyserythropoietic anemia

Increased physiologic requirement
Extracorpscular like alloimmune & isoimmune hemolytic anemia, microangiopathic anemias, infections, hypersplenism, Intracorpsular defect like :
Red cell membranopathy i.e. congenital spherocytosis, elliptocytosis
Hemoglobinopathy like HbS, C,D,E etc.
Thalassemia syndrome
RBC enzymopathies like G6PD deficiency, PK deficiency etc.


Symptoms of
Anemia
Red = In severe anemia
Central
- Fatigue
- Dizziness
- Fainting
Eyes
- Yellowing
Skin
- Paleness
- Coldness
- Yellowing
Blood vessels
- Low blood pressure
Heart
- Palpitations
- Rapid heart rate
- Chest pain
- Angina
- Heart attack
Respiratory
- Shortness of breath
Muscular
- Weakness
Intestinal
- Changed stool color
Spleen
- Enlarge-ment

Etiology

B Thalassemia

- Family Hz, mild anemia, Mediterranean, Asian, or African Heritage
- **TRAIT**: single defect in B-globin gene results in less normal B-globin chains
- Decrease HG A1
- Autosomal Recessive
- **B MAJOR**: little to no prod. of B chain of HG A1
- Noted > 6months when gamma chain and Hg F prod. usually fall

A Thalassemia

- Silent Carrier: 1 gene
- A Thal Trait: Asian Africans, 2 genes
- Hb H Disease: 3 genes which causes a lack of A chains
- Bart's Hemoglobinopathy: 4 genes leades to catastrophic anemia and Hydrops fetalis

HEREDITARY SPHEROCYTOSIS

RBC membrane defect with **SPECTRIN**
RBC destroyed prematurely in spleen
MOST COMMON HEREDITARY RED CELL DISORDER!!!!!!
Autosomal Dominant

Increased RBC turnover leads to **cholelithiasis** and **cholecystitis**
Susceptible to aplastic crisis from **PARVOvirus**
Physical shows pallor, jaundice, splenomegaly
Lab findings include reticulocytosis, increased MCHC (decreased in IDA), spherocytes in smear.

Antiglobulin test rules out an immunce cause for the HA
Osmotic fragility test
Tx includes careful management of situation, esp. aplastic crisis
Splenectomy: spherocytes remain but RBC destruction stops! **(only after 5 yrs old)**
Immunize prior to procedure with **Hib**, **Pneumovax**, and **N. meningitis vaccine** b/c increase rish for encapsulated organisms.
Also penicillin prophy

AREGENERATIVE ANEMIAS

- ***Parvo B19***
- Fifth's Disease with affinity for red cell precursors causing marrow aplasia
- Causes Hydrops Fetalis

Diamond-Blackfan Anemia
Relative insensitivity to EPO (idiopathic)
Develops **insidiously** in 1st year of life **and no recovery!**
Short stature, abnormal facies, abnormal thumbs
Macrocytosis (any anemic child with Macrocytosis is very serious)
Tx includes transfusions, steroids for life
Increase risk of myelogenous leukemia

Transient Erythroblastopenia of Childhood
2nd yr of life and is idiopathic
VERY low Hbn but no symptoms
Recover with no intervention at all!
Normochromic/normocytic anemia

SICKLE CELL DISEASE

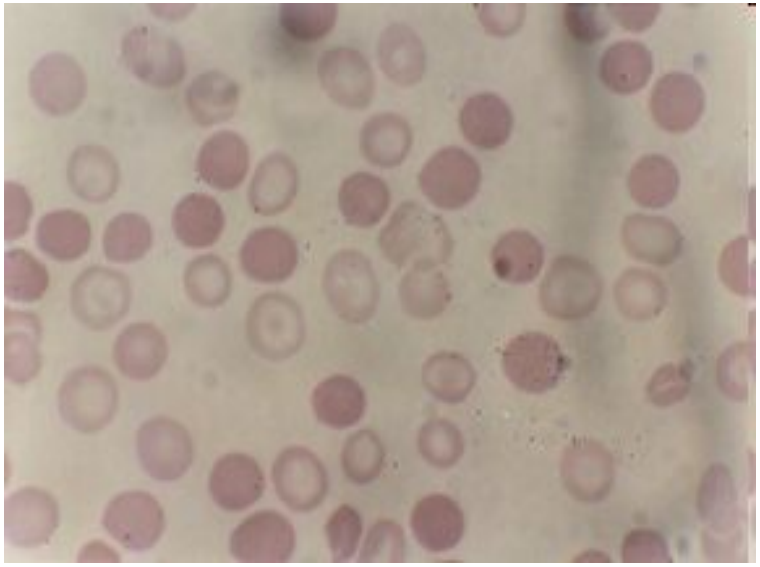
Etiology:

Valine for glutamic Acid in 6th position of Beta chain Hb

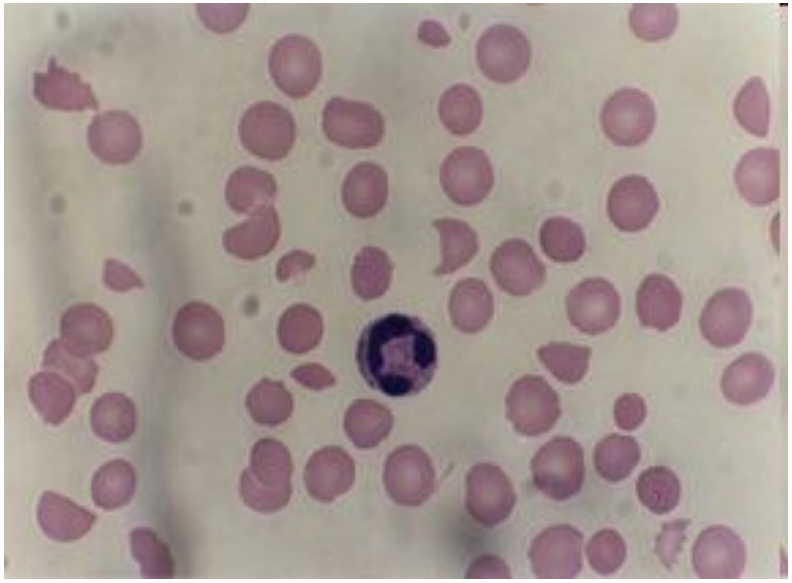
- **Most common in African descent**
- **Only appears after 6 months when B chains have fully developed into Hb A1.**
- **Defect on Chrom 11 Neonatal screening!!!**

G6PD DEFICIENCY

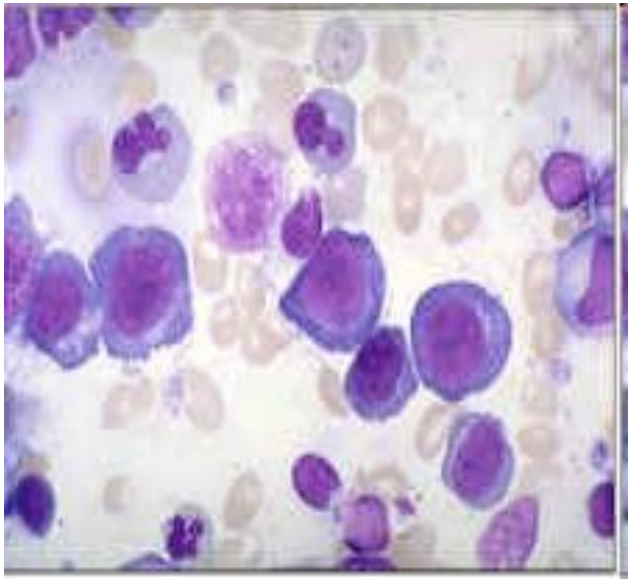
- Central enzyme in PPP pathway
- Makes NADPH which forms reduced Glutathione that removes radicals
- **X-Linked**
- **A form** is common in AA and mild
- **B form** in Meds and very serious
- **Canton form** in oriental and rare but most serious



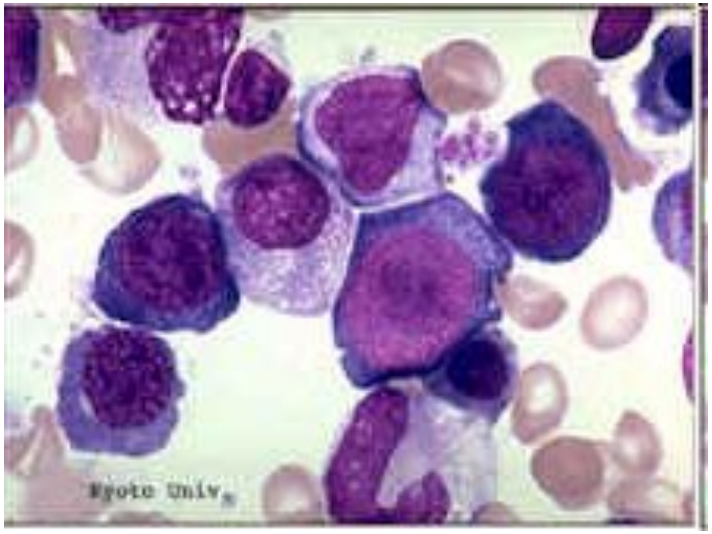
Microcytic Anemia - Hypochromia, Target Cells, Microcyte



Normocytic Normochromic - Hemolytic Anemia



Megaloblastic Anemia, Bone marrow smear, May-Giemsa stain, x1000



Pernicious Anemia, Bone marrow smear, May-Giemsa stain, x1000