Hemolytic Anemia

Anemia is an abnormally low level of red blood cells. Hemolytic anemia occurs when red blood cells are destroyed too quickly.

In people with hemolytic anemia, red blood cells have an abnormally short life span. There can be something wrong with the red blood cells. Or the red blood cells are normal but are destroyed by an external process.

Many different problems can cause hemolytic anemia. These include:

1. **Inherited abnormalities in red blood cell membranes :**

   A red blood cell is like a little balloon filled with fluid. Defects in the membrane can cause the cells to change shape. Abnormally shaped red blood cells are identified by the spleen as abnormal, and destroyed.

   e.g. : - 1- Hereditary Spherocytosis.
   2- Hereditary Eliptocytosis .
   3- Hereditary Stomatocytosis.

2. **Inherited enzyme deficiencies inside red blood cells :**

   Enzymes are proteins. Certain abnormal enzyme levels cause red blood cells to become fragile.

3. **Hemoglobin disorders :**

   Some people inherit a gene that makes an abnormal kind of hemoglobin. Hemoglobin disorders include :

   1- Sickle cell anemia (defective hemoglobin)
   2- Thalassemias (defective synthesis) : Hemoglobin disorders can cause red blood cells to be destroyed easily.

4. **(Acquired) Physical damage to red blood cells:**

   Hemolytic anemia can occur when red blood cells are damaged:

   a. During heart-lung surgery .
   b. Artificial heart valves .
   c. Exposed to excessive heat, as in a patient with severe burns.

5. **(Acquired) Autoimmune hemolytic anemia :**

   This occurs when the body's immune system mistakenly destroys its own red blood cells. Autoimmune hemolytic anemia can be caused by:

   a. autoimmune conditions such as lupus .
   b. certain types of infections .
   c. certain medications .(drugs)

6. **Hypersplenism :** The spleen is enlarged and overactive circulating red blood cells and destroys them before they are old.
7. **Miscellaneous :**
    a. Haemolytic anaemia , due to direct action of drugs or Chemicals .
    b. Haemolytic anaemia , due to infection (***Clostridium walchii***) .
    c. Haemolytic anaemia , due to burns .
    d. Lead poisoning .

Symptoms :

Symptoms vary widely. Mild cases may not cause any symptoms. The condition may only be discovered during a routine blood test.

In other people, clear symptoms of anemia can develop. These include:

1. Pale skin
2. Fatigue
3. Weakness
4. Dizziness
5. Lightheadedness
6. Breathlessness
7. Yellowing of the skin and eyes
8. Dark tea-colored urine .

I) Hereditary Haemolytic Anaemia :

A – Defects of membrane :

1 – Hereditary Spherocytosis :

The commonest hereditary haemolytic anaemia in north European , due to defect in a structural protein of the red cell membrane . The cell becomes more spherecal and unable to pass through the splenic microcirculation , where the spherocyte die prematurely . the reason for loss of the normal biconcave shape of the red cells .

Laboratory Diagnosis of Hereditary Spherocytosis :

☒ **Blood picture :**

1 – Hb. = 9 – 12 g / dl.
2 – Osmotic Fragility = up to 0.7 – 0.8 .
3 – Reticulocyte count are raised to 5 – 7 % .
4 – Coombs test = negative .
5 – Auto-haemolysis is increased .
6 – Platelets count decreased .
7 – Blood smear == spherocyte
8 – MCV normal Or reduced .
9 – MCH normal .
10 – MCHC often increased (34 – 40 %).

☒ **Biochemistry :**

1 – Bilirubin slightly raised .

2 – Urine bilinogen raised .

3 – Haptoglobin decreased .

2 – Hereditary Elliptocytosis :

This has similar clinical and laboratory features to hereditary spherocytosis except for the appearance of the blood film .

B – Defect of Haemoglobin :

1 – Abnormal of Haemoglobin :

These are divided into main groups :

a – Synthesis of an Abnormal Haemoglobin :

These contain an amino acid substitution in either alpha or Bata globin chain .

The most important of these diseases in Sickle cell anaemia .

b – Reduced Synthesis Of Abnormal Globin Chains :

This includes the alpha and Beta – thalassaemia In which synthesis of one or other globin chain is reduced .

2 – Sickle Cell Anaemia :

This disease is due to the synthesis of an abnormal Haemoglobin , which only differ from the normal adult Hb. In that the amino acid in the sixth position from the amino end of one of the two peptide chain types is Valine instead of the glutamic acid in beta chain .

This abnormality represents a different only one amino acid in zoo .

Laboratory Diagnosis : -

1- Hb. Is usually 6 – 9 gm. / dl.

2- Sickle cell and target cell occur in the blood .

3- Features of splenic atrophy (e.g. Howelly-Jolly bodies may also be present).

3 – Thalassaemia :

There are 2 types : 1 – Alpha , affecting mainly Alpha chain

2 – Beta, affecting mainly Beta chain.

☒ **Beta – Thalassaemia : (major)**

There is reduced Beta chain production hence reduced Hb. A Leading to microcytic , hypochromic anaemia ,

Increase in Hb. A2 and Hb. F , because the lack of Bate chain , alpha chains accumulate in cells forming aggregates in

erythroblast and mature red cells causing infective erythropoiesis and haemolysis .

Laboratory Diagnosis of Beta-thalassaemia Major :

☒ **Blood Picture :**

Resemble iron deficiency anaemia .

1 – Microcytosis with hypochromia .

2 – Target cells are prominent .

3 – All MCV, MCH, MCHC are decrease .

4 – Reticulocytosis 10 % .

5 – Osmotic Fragility decreased .

❖ **Beta-thalassaemia (minor) :**

Usually symptomless abnormality , characterized by marked , Hypochromic , microccytic blood picture .

MCV , MCH , MCHC , all very low , but mild or no Anaemia (Hb. = 11 – 15 gm./dl) .

A raised Hb. A2 confirms the diagnosis

II) Acquired Haemolytic Anaemia :

❖ **Auto- Immune Haemolytic Anaemia (AIHA) :**

These anaemia are due to antibody production by the body against its own red cell .

They are characterized by a positive direct Coombs test , and divided into **warm** , and **cold** types according to whether .

The antibody reacts better with red cells at 37°c or 4°c .

- **Warm AIHA :**

  The red cells are usually coated with IgG either alone Or with complement . but a minority of cases show IgA or IgM coating alone or combined with IgG antibody .

- **Cold AIHA :**

  In these syndromes the auto-antibody attaches to red cells mainly in the peripheral circulation , where the blood temperature is cold , and.

  the antibody is usually IgM , and bind to the red cells best at 4 °c .