HEMATOLOGY & HEMOSTASIS ACADEMY

Section 4

DISORDERS OF RED BLOOD CELLS

LEARNING OBJECTIVES

When you complete this section, you will be able to:

1. **Define anemia**
2. **Recognize terms used to describe RBC changes in anemia**
3. **Indicate types ofanemia**
4. **Describe polycythemia**

ANEMIA

Anemia indicates a deficiency of RBCs. It is a clinical sign, not a diagnosis. The terms that physicians may use to describe types of defective RBCs and anemias are listed and defined below.

- **Anisochromatic:** Unequally distributed hemoglobin in RBCs; the periphery is pigmented but the centers are almost colorless (aniso- means unequal, -chromic means color)
- **Anisocytosis:** A condition in which RBCs vary considerably in size hypochromic: Pale; less than normal hemoglobin
- **Macrocytic:** Larger than normal non-nucleated RBC; macrocytic hyperchromic cells contain large amounts of hemoglobin due to their size
- **Microcytic:** Smaller than normal non-nucleated RBC
- **Normochromic:** Normal color; normal hemoglobin
- **Normocytic:** Normal healthy cell
- **Poikilocytosis:** A condition in which RBCs have irregular shapes (poikilo- means irregular)

The following are some of the most common anemias. They are arbitrarily grouped according to RBC color and size; other sources may group them in different ways.

1. Anemia due to bleeding: hypochromic-microcytic anemia

A temporary anemia can occur after a rapid hemorrhage due to trauma such as a car accident, surgery, or childbirth. The body quickly replaces plasma, but it usually needs several weeks to replace RBCs. With chronic blood loss, as in undiagnosed internal hemorrhaging, anemia develops due to lack of sufficient functional iron and RBCs contain too little hemoglobin.

2. Anemia due to deficient rbc formation: hypochromic-microcytic anemia

Iron-deficient anemia is very common. It is often due to defective production of heme or globin. The iron deficiency may, in turn, be due to:

- Lack of dietary iron (strict vegetarianism or poor diet)
- Defects in iron utilization. Called sideroblastic anemia, the condition is characterized by ineffective RBC formation and the presence of ferritin in developing RBCs
- Defects in iron reutilization. This is the second most common form of anemia and is related to a variety of chronic diseases, infections, and cancers
- Defects in iron transport; a very rare condition called atransferrinemia

3. Anemias due to deficient rbc formation: normochromic-normocytic anemias

Anemia Due to Renal Failure. Chronic kidney failure shortens the life span of RBCs, probably due to an interaction with uremia (excess nitrogen waste in the blood). Replacing RBCs is hindered because failing kidneys release erythropoietin inefficiently.

Aplastic or Hypoplastic Anemia. Both aplastic and hypoplastic indicate incomplete or defective development. They describe an anemia that arises from partial failure of the bone marrow. Some physicians prefer the term pancytopenia, which means reduced numbers of all types of blood cells. Causes include x-rays, toxic chemicals (benzene, DDT), therapeutic drugs (chloramphenicol, phenylbutazone), or infections such as viral hepatitis.

4. Anemias due to excessive rbc destruction

Hemolytic Anemias. Hemolysis (lysis = break down) is a process in which red cells fall apart and spill out their hemoglobin. Some hemoglobin normally leaks out of cells and dissolves in plasma. Anemia arises when hemolysis occurs in response to a number of factors: spleen dysfunction, immunologic abnormalities, trauma, blood transfusion reactions, malaria, use of some therapeutic drugs, exposure to toxic chemicals, or infections.

Genetic hemolytic conditions include a deficiency in the enzyme G6PD (glucose 6-phosphate dehydrogenase). In some inherited conditions, RBC abnormalities cause the cells to rupture easily, especially in the spleen. In the resulting anemias, sufficient or even excessive RBCs are formed, but their life span is very short. Types of hemolytic anemias due to RBC defects include:

- Hereditary spherocytosis. Microcytic spherical RBCs rupture easily.
- Sickle cell anemia. Hemoglobin S, an abnormal hemoglobin, precipitates into crystals when exposed to oxygen, damaging the cell membrane and causing it to assume a sickle shape. Anisocytosis and poikilocytosis are present. This serious condition is present in about 1% of West African and American Blacks.
- Thalassemia. Faulty or incomplete formation of hemoglobin causes production of tremendous numbers of hypochromic, microcytic, fragile RBCs that often rupture before leaving the bone marrow. Anisocytosis and poikilocytosis are present.
- Erythroblastosis fetalis. RBCs in the fetus are attacked by antibodies from the mother, causing the child to be born with serious anemia.

5. Megaloblastic anemia (pernicious anemia)

These patients remain in constant need of RBCs due to production of oversized, strangely shaped, normochromic RBCs that rupture easily. Causes are inadequate vitamin B12 or folic acid, or both (**Table 4**). Pregnancy increases folic acid requirements and can lead to "megaloblastic anemia of pregnancy" in the last trimester and in pregnancies involving multiple fetuses.

UNDERLYING CAUSES OF VITAMIN B12 AND/OR FOLIC ACID DEFICIENCIES
Insufficient diet (due to poverty, strict vegetarianism)
Damage to the stomach lining
Gastrectomy; total removal of the stomach
Inflammatory conditions of intestines; such as sprue, a tropical disease
Intestinal parasites
Malabsorption syndromes
Alcoholism
Pregnancy and lactation

Table 4. Causes of B12 and/or folic acid deficiencies.

POLYCYTHEMIA

This is the opposite of anemia; the term literally means "many cells in the blood". The blood becomes highly viscous (thick) and flows sluggishly. The condition results when tissues become hypoxic. Examples include:

- **Secondary polycythemia.** People who live in high altitudes automatically produce large quantities of RBCs due to the sparse oxygen in the air. A patient with cardiac failure is also likely to develop this condition; the inefficient heart is not able to deliver enough oxygenated blood to tissues.
- **Polycythemia vera** (also erythremia). This tumorous condition of the blood-forming organs causes production of massive amounts of RBCs, WBCs, and platelets. The most common cell line elevated in polycythemia vera is the RBC often referred to as red cell dyscrasia.

COLD AGGLUTINATION

This is a disorder in which RBCs – in their own serum or in other serum – clump together (agglutinate) in response to slight cooling (below 86° F). The clumping is caused by a group of antibodies called agglutinins. Cold agglutinins may be present in elderly people or may be due to infections such as those causing atypical pneumonia or infectious mononucleosis. The agglutination may be mild and transient or may progress into a disease called "cold antibody disease" or "cold agglutinin disease". Hemolysis, hemoglobinuria, and hemosiderinuria may be present.

The presence of cold agglutinins in the blood is likely to cause discrepancies in the results of blood tests.