

Hematology

Aplastic anemia and Polycythemia

LEC 9 &10

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- **Aplastic anemia** is a blood disorder that occurs when the bone marrow produces too few of all types of blood cells: red cells, white cells and platelets (**Pancytopenia**).
- ☐ A low number of **red blood cells** reduces the blood's ability to carry oxygen.
- ☐ A reduced number of **white blood cells** makes the child more susceptible to infection.
- ☐ A low number of **platelets** reduces the blood's ability to clot.

- **Causes of Aplastic anemia:**
- **A. Primary:**
- 1. Idiopathic: occurring with no known reason.
The disorder can be the result of a previous illness or presented problem.
- 2. Inherited genetic disorder.

- **B. Secondary : (Acquired causes)**
- 1. Specific infectious diseases, such as hepatitis, Epstein-Barr virus, or cytomegalovirus.
- 2. taking certain medications including some antibiotics and arthritis drugs.
- 3. Exposure to certain toxins, such as benzene, TNT, Chlordane, and DDT.
- 4. Exposure to radiation or chemotherapy.

- **Symptoms:**

- 1. lack of energy or tiring easily
- 2. pale skin, lips, and hands, or paleness under the eyelids
- 3. shortness of breath
- 4. fevers or infections
- 5. bleeding, such as bruising, bleeding gums, nosebleeds or blood in the stool
- 6. irregular heartbeat
- 7. Headache.

- **Laboratory Tests:**

- **1.** The initial test for anemia, the complete blood count (CBC), may reveal many abnormal results.
- ☐ Hemoglobin, RBC, WBC, and platelet count are low.
- ☐ Red blood cell indices are usually normal.
- ☐ The differential white blood count shows a decrease in most types of cells but not lymphocytes

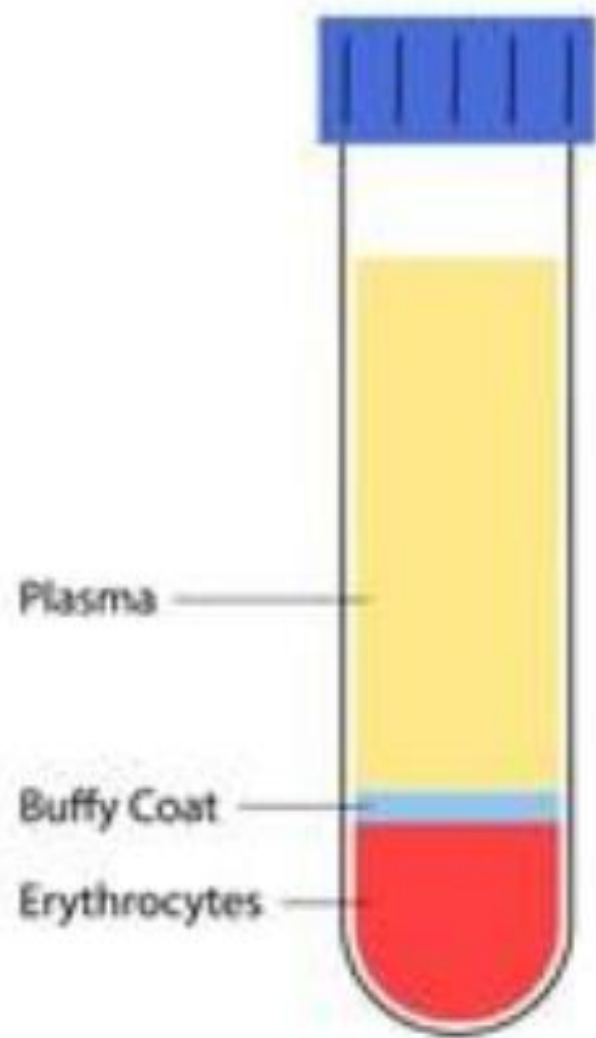
- 2. Some additional tests that may be performed to help determine the type and cause of anemia include:
 - 1. Reticulocyte count—low.
 - 2. Bleeding time —increased.
 - 3. Erythropoietin—increased.
 - 4. A bone marrow aspiration will show a decrease in the number of all types of cells.
 - 5. Tests for infections such as hepatitis, EBV, CMV help to determine the cause.
 - 6. Test for arsenic (a heavy metal) and other toxins.

Polycythemia

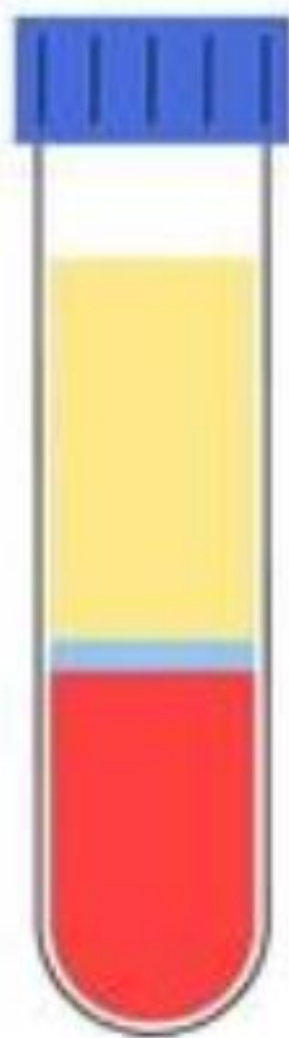
- Is defined as an elevation in packed cell volume (PCV), rather than a raised haemoglobin concentration

- **Classification of polycythemia**
- **1. Absolute polycythemia:** in which the red cell mass (volume) is raised, can then be subdivided into
 - **a. Primary polycythemia**
 - 1. Polycythemia vera
 - 2. Familial polycythemia

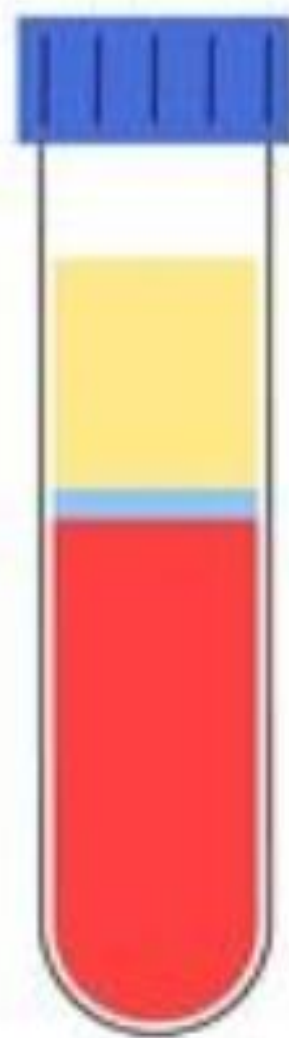




Anemia



Normal blood



Polycythemia

- **b. Secondary polycythemia**
- 1. Caused by **compensatory erythropoietin** increase in high altitudes, pulmonary disease, cardiovascular disease (especially congenital), heavy cigarette smoking.
- 2. Caused by **inappropriate erythropoietin** increase in renal diseases, tumours, and hepatocellular carcinoma

- **2. Relative or pseudopolycythaemia:** in which the red cell volume is normal but the plasma volume is reduced.
- ☐ Dehydration: water deprivation, vomiting
- ☐ Plasma loss: burns, enteropathy

- **Polycythemia vera**
- In Polycythemia vera , the increase in red cell volume is caused by a clonal malignancy of a marrow stem cell. The disease results from somatic mutation of a single **haemopoietic stem cell**. The JAK2 mutation is present in haemopoietic cells in almost 100% of patients JAK2 activation leads to cell survival and proliferation. Although the increase in red cells is the **diagnostic finding**, in many patients there is **also increase** in granulocytes and platelets.

- **Clinical features**

- This is a disease of older subjects with an equal sex incidence.
- 1. Headaches, blurred vision and night sweats.
- 2. Pruritus, characteristically after a hot bath.
- 3. Splenomegaly in 75% of patient.
- 4. Haemorrhage (e.g. gastrointestinal, uterine, cerebral) or thrombosis.
- 5. Hypertension in one-third of patients.
- 6. Gout (as a result of raised uric acid production).
- 7. Peptic ulceration occurs in 5-10% of patient

- **Laboratory findings of polycythemia**
- 1. The complete blood count (CBC):
 - ☐ Hemoglobin, RBC count and PCV are increased.
 - ☐ A **neutrophil increased** in over half of patients, and some have **increased** circulating **basophils**.
 - ☐ **Platelet** count **increased** in about half of patients.

- 2. The JAK2 mutation is present in nearly 100% of patients.
- 3. The neutrophil alkaline phosphatase (NAP) score is increased.
- 4. The bone marrow is hypercellular.
- 5. Serum erythropoietin usually low.
- 6. Blood viscosity is increased.
- 7. Circulating erythroid progenitors and erythroid colony forming unit (CFU-E) are increased compared to normal.

**Thank You
For
Listening**