3.2.8

Polycythemia

Polycythemia is an abnormally high total red blood cell count with a hematocrit greater than 50%. It is split into two categories, relative and absolute, based on the cause of the polycythemia. Please watch the video Polycythemia. This video will have more detail than you need. Use it to understand the level of material taught below and on the study guide.

Relative Polycythemia

The increased hematocrit of relative polycythemia is caused by a loss of plasma volume without an increase in red blood cells. Some possible examples of this could be excess use of diuretics, diarrhea, and dehydration.

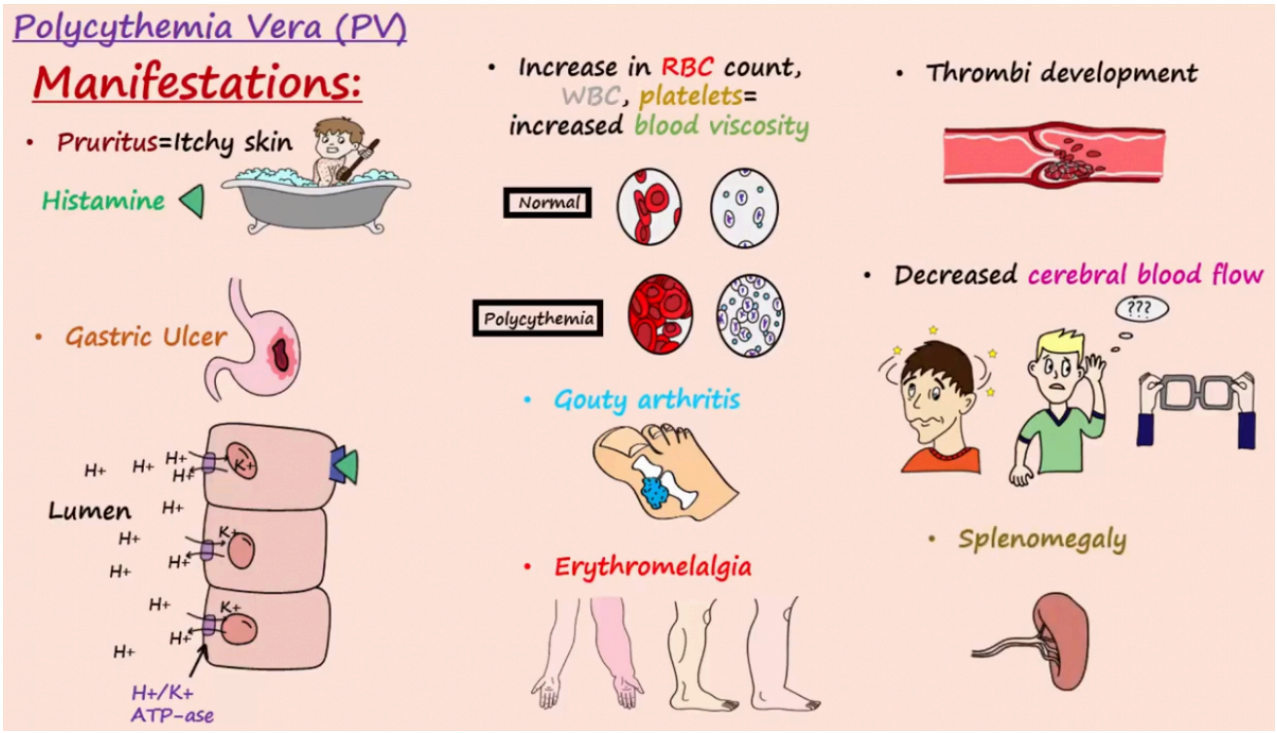
Absolute Polycythemia

Absolute polycythemia is characterized by an increase in red blood cell count and hematocrit due to an overall increase in the total mass of red blood cells. Absolute polycythemia can be broken down into two categories: primary and secondary.

Primary polycythemia is a genetic condition characterized by elevated levels of red blood cells, white blood cells, and platelets. Hematocrit, hemoglobin levels, blood volume and blood viscosity are all increased. An increase in viscosity and hematocrit will affect cardiac output and blood flow. Patients often experience splenomegaly because of the increased number of RBCs and decreased cerebral blood flow which causes difficulty with hearing and vision. Venous stasis can cause the skin to appear bluish, especially in the lips and fingernails. Treatment to reduce the risk of thromboembolism is done through phlebotomy and reduces the hematocrit and blood viscosity. Hydroxyurea is commonly used in the treatment of primary polycythemia and suppresses bone marrow function and controls the elevated blood cell count. JAK inhibitors are another class of drugs that can help block the second messenger systems stimulated by EPO.

Primary Familial & Congenital Polycythemia (PFCP) is a specific example of primary polycythemia. It is an inherited mutation to the EPO receptor that can make it overactive. This increases erythropoiesis and leads to a congenital rise in a person's hematocrit.

Polycythemia Vera (PV) is another example of primary polycythemia, however this condition is not congenital. PV results from a genetic mutation in hematopoietic stem cells that causes excessive production of RBCs, WBCs, and/or platelets. It is unknown what causes this mutation to occur, but it is most common between the ages of 50-75. The image below details some aspects of PV.



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Secondary polycythemia is an acquired condition that often results from increased levels of erythropoietin as a compensatory mechanism to hypoxia. Hypoxia can be caused by smoking, chronic heart and lung disease, living at higher altitudes, sleep apnea, neoplasms, and chronic carbon monoxide exposure. Another possibility for secondary polycythemia is pathology in the kidney that results in excessive EPO release, even in the absence of hypoxia. Treatment for secondary polycythemia generally focuses on relieving hypoxia (for example supplemental oxygen may help in many instances).