Sickle Cell Anemia

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Pathophysiological Process

- SCA is a genetic disorder inherited from a mother, or both parents that are present from birth. SCA is as a result of a single mutation on a globin beta gene which causes the synthesis of an abnormal hemoglobin S (HbS) (Connes et al., 2018). Hemoglobin (Hb) is a molecule in RBC responsible for the transportation of oxygen and carbon dioxide. RBC polymers and cell fragmentation which also leads to a pathological cascade that results in injury to the red cell sickle, hemolysis, and vaso-occlusive crisis of RBC (Connes et al., 2018).

- The sickled RBC becomes a blockage that slows or stops the flow of blood (Conn et al., 2018). From this happen, oxygen cannot reach target tissues. Dehydration can cause atonies of sudden and severe pain called “pain crises” (Conn et al., 2018). Due to the deformed shape, cells can also get stuck with walls (Waltz, 2014). RBC membrane damage leading to an imbalance of calcium, potassium, and water exchange into and out of the cell (Waltz, 2014). Accumulation of Calcium within the cell allows for the efflux of water which shrinks the cell and can result in a Vaso-occlusive crisis (Waltz, 2014). The vaso-occlusion causes an obstruction and reduces blood flow to the vital organs leading to ischemia and, necrosis. Repeated episodes of dehydration and oxygenation weaken the RBC cell membranes (Smith, 2014). The average life span of a healthy RBC is 120 days. However, due to the formation of the sickled cell, the results in abnormal HbS present in the person’s red blood cells (Waltz, 2014). Pain crises have a hereditary influence on individuals with a sickle trait. The swelling and inflammation of the hands and other areas of the body can occur when a sickle cell crisis occurs. The sickle cell crisis is associated with the patient’s quality of life and life span (Waltz, 2018).