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Journal of Hematology & Thromboembolic Diseases

Perspective

Pathophysiology of the Sickle Cell Anaemia

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DESCRIPTION

Sickle Cell Anaemia (SCA) belongs to a group of hereditary disorders. In this condition the shape of red blood cells changed, which is useful for the carry oxygen to all parts of the body. Red blood cells can easily move within blood vessels they are round and flexible.

Pathophysiology

SCA is characterized by two major components: Hemolysis and Vaso-Occlusive Crisis (VOC). A defect in the betaglobin gene facilitates the conversion of Sickle Cell Hemoglobin (HbS) molecules into hard, elongated polymers in the deoxidized state. The sickleization process is initially periodic, with sickle cells oscillating between a normal biconcave shape and an abnormal sickle shape (acquired under hypoxic pressure). However, the time has come for changes to become irreversible, and for the diseased red blood cells to become a permanent disease, increasing the risk of hemolysis and VOCs. All variants of SCD share the same pathophysiology leading to the polymerization of HbS components

After the red blood cells are deoxidized, when the red blood cells take the form of pathological sickle cells, it is solely responsible for causing vascular occlusion. Vascular occlusion is central to understanding the disease and can result in local hypoxemia with direct tissue damage and inflammation, but the monogenic mutations found in sickle cell disease it results in complex physiological changes. These changes lead to a variety of clinical manifestations of the disease. Sickle cell anemia is now recognized as a condition characterized by increased inflammation, increased coagulation, increased oxidative stress, and impaired arginine metabolism, as well as vascular occlusion, anemia, and hemolysis. Sickle cell disease is an angiopathy and is also characterized by the presence of a deficiency of multiple nutrients and micronutrients that adversely affect the patient. When deoxidized, the sickled hemoglobin becomes insoluble, polymerizing the polymer and aggregating it into tubulin fibers, which form the tubulin fibers. Due to their rigid shape, cells tend to be trapped in the microcirculation, but the tissues downstream of this obstruction are deprived of blood flow and oxygen suffers from ischemic damage or death. This lack of blood flow, in turn, leads to tissue necrosis or reperfusion injury. These sickle cells also tend to dry out due to abnormalities in the Garudos Canal. These cells are characterized by aberrant activation of intracellular signaling pathways, with low levels of nitric oxide and adenosine triphosphate. These cells also have low antioxidant capacity as a result; many of the cellular components can exhibit oxidative damage. Oxidative damage to cell membrane proteins and protein aggregation along the inner surface of the plasma membrane can lead to intracellular abnormalities on the surface of erythrocytes. Such changes ultimately result in increased exposure to phosphatidylserine and the formation of microparticles that allow the red blood cells themselves to promote coagulation.

During hemolysis, free hemoglobin is released into plasma and acts as a nitric oxide scavenger. The arginase 1 activity required for nitric oxide production is lower in sickle cells than in normal erythrocytes, so new nitric oxide cannot be easily produced, especially in individuals who tend to hemolyze at a high rate. Another result of hemolysis is the formation of reactive oxygen species by reactions involving free hemoglobin. In addition, in sickle cells, microRNA dysregulation occurs, small non-coding RNA molecules shut down RNA, and posttranscriptional regulation of gene expression occurs. Therefore, gene expression during erythropoiesis is abnormal. Abnormal adhesive properties of sickle cells can lead to activation of adhesive receptors such as: In addition to these changes, cells containing sickle cell hemoglobin are harder than normal circulating erythrocytes. Due to the abnormal deformability persists even when the cell has assumed as normal ovoid shape. Morphologically normal sickle cells Hemoglobin-containing red blood cells tend to attach like irreversible sickle cells.

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Received: 02-Mar-2022; Manuscript No. JHTD-22-17205; **Editor assigned:** 04-Mar-2022; Pre QC. No. JHTD-22-17205 (PQ); **Reviewed:** 17-Mar-2022; QC. No. JHTD-22-17205; **Revised:** 22-Mar-2021; Manuscript No. JHTD-22-17205 (R); **Published:** 06-Apr-2022, DOI: 10:352481/2329-8790.22.10.478.

Citation: Rezaei F (2022) Pathophysiology of the Sickle Cell Anaemia. J Hematol Thrombo Dis. 10:478.