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This medical industry feature, titled "Impact of Hemoglobin S Polymerization on Sickle Cell Disease" is sponsored by Global Blood Therapeutics.

Speaker:
Just a single point mutation in the beta-globin gene leads to the debilitating damage of sickle cell disease. This inherited change drives a complex, unrelenting condition characterized by vaso-occlusion, chronic hemolysis, and chronic anemia. Hemoglobin S polymerization is the root cause of sickle cell disease pathology and its long-term sequelae. In low oxygen environments, hemoglobin S molecules coalesce and begin to polymerize. The polymers coalesce into long fibers that distort red blood cells into the characteristic sickle shape. Hemoglobin S polymerization is the key event that leads to drastic changes in the integrity and function of red blood cells. The polymers deform red blood cell membrane structure, making the cells much more rigid and adhesive. This slows or obstructs blood flow, resulting in vaso-occlusion and diminished oxygen delivery. Lower local oxygen levels induce further sickling, vaso-occlusion, reperfusion injuries, and inflammatory responses. Additionally, membrane changes caused by hemoglobin S polymers lead to cellular dehydration, chronic hemolysis, and early cell death. Sickle cells only survive for about 10 to 20 days versus 120 days for healthy red blood cells, which stresses the bone marrow and increases reticulocyte production. When red blood cells become fragile and lyse, they release hemoglobin and other cellular contents that contribute to vasculopathy and further inflammation. Free hemoglobin is broken down, decreasing the amount of active hemoglobin and circulation and leading to chronic anemia and its clinical complications. Left unchecked, the pathologic effects of vaso-occlusion, chronic hemolysis, and chronic anemia can lead to progressive tissue damage and end organ damage. Organs that may be affected by long-term chronic damage include the brain, eyes, lungs, heart, kidneys, and gallbladder. In summary, a single nucleic acid substitution in the beta-globin gene causes hemoglobin S polymerization that initiates extensive pathological changes leading to vaso-occlusion, chronic hemolysis, and chronic anemia. There is an ongoing need for improved management of patients with sickle cell disease and related end organ damage.

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