Sickle Cell Disease and Hidden Hearing Loss

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EXECUTIVE SUMMARY

Sickle cell disease (SCD) is an autosomal recessively inherited group of red blood cell disorders. It is more commonly found in African Americans and Hispanics in the United States. The sickle-shaped cells characteristic of the disease cause physiological problems, including hemolytic anemia and vaso-occlusion. Research has linked hearing loss to sickle cell disease. Studies have shown that children and adults with sickle cell disease may have higher rates of sensorineural and central auditory hearing impairment. This case study examines hidden hearing loss in an African American boy with SCD who presents with normal hearing thresholds. It also discusses the role of audiologists in the care of individuals with sickle cell disease and hearing loss.

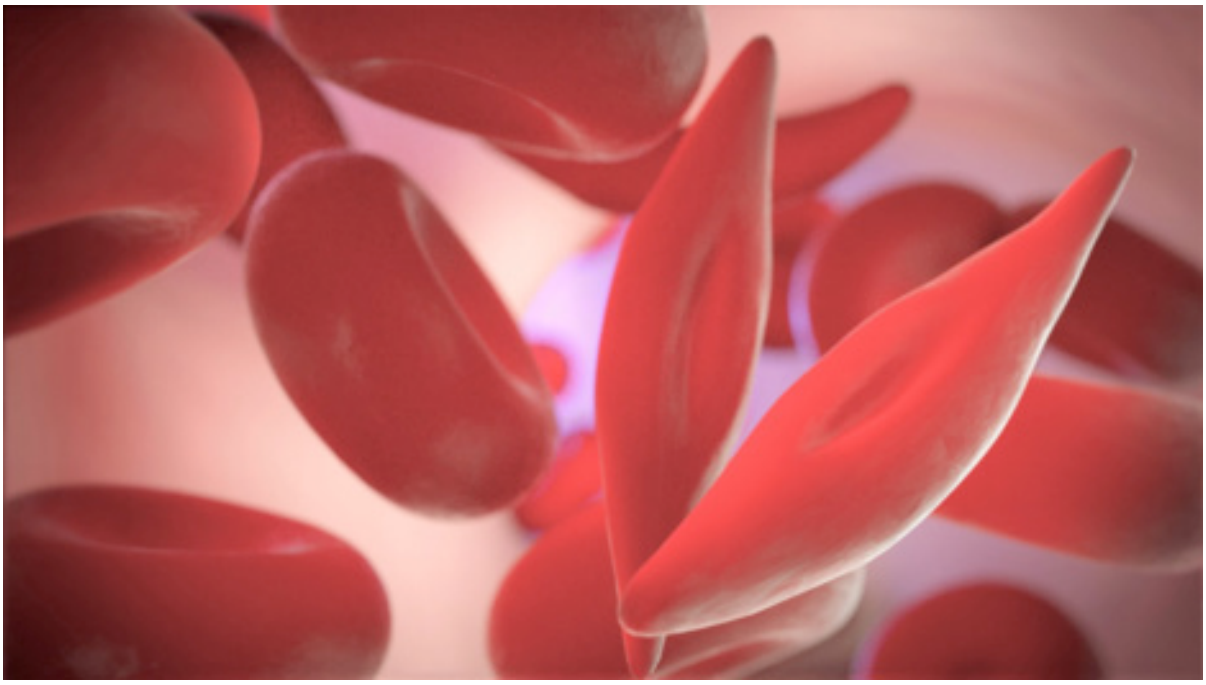
INTRODUCTION

Sickle Cell Disease

Sickle cell disease (SCD) is a group of autosomal recessively inherited red blood cell disorders. Red blood cells contain a protein called hemoglobin (Hb). Hemoglobin gives blood its characteristic red color, and its job is to transport and release oxygen throughout the body. People who have sickle cell disease have abnormal hemoglobin in their red blood cells. The abnormal hemoglobin, known as hemoglobin S (HbS), causes sickle cell disease.

Red blood cells that contain normal hemoglobin are disc-shaped and flexible so that they can move easily through large and small blood vessels of the body to deliver oxygen. Since red blood cells are soft and pliable, they can squeeze through blood vessels that are narrower than themselves. This flexibility (pliability) occurs because the normal hemoglobin remains dissolved in the presence of low levels of oxygen. Sickle hemoglobin is not like normal hemoglobin. Problems occur when oxygen levels in the

Figure 1. Normal and sickled red blood cells (U.S. National Library of Medicine)



blood are lower, which happens once the hemoglobin has delivered oxygen to cells in the body's tissues. With less oxygen present, the abnormal hemoglobin can cause rigid, nonliquid protein strands to form within the red blood cell. These rigid strands can then change the shape of the cell, producing the sickled red blood cell that gives the disease its name.

Because sickle-shaped cells are not flexible, they can stick to vessel walls, causing a blockage that slows or stops the flow of blood, referred to as vaso-occlusion. When this happens, oxygen cannot reach nearby tissues. The lack of oxygen in tissues can cause attacks of sudden severe pain, otherwise known as pain crises. Pain crises can occur without warning, and individuals who have them often need to go to the hospital for effective treatment.

Given that sickle cells cannot change shape easily, they tend to burst apart. Normal red blood cells live about 90 to 120 days in the body, but sickle cells last only 10 to 20 days. The body is always making new red blood cells to replace the old cells. In the case of sickle cell disease, the body may have trouble keeping up with replacing the old red blood cells given how quickly the old cells are being destroyed. Under these circumstances, the number of red blood cells circulating in the body is usually lower than normal. This condition is called hemolytic anemia, which can lead to fatigue and jaundice (National Heart, Lung, and Blood Institute, 2019).

Over time, SCD can lead to major complications such as infections, delayed growth, and more pain crises. Most children who have sickle cell disease are pain-free between crises, but adolescents and adults may also suffer with chronic, ongoing pain. Over a lifetime, SCD can harm an individual's spleen, brain, eyes, lungs, liver, heart, kidneys, penis, joints, bones, or skin (National Heart, Lung, and Brian Institute, 2019).