Sickle cell anemia is an inherited blood disorder in which there are not enough healthy red blood cells to bring oxygen throughout the body. Normally, red blood cells are circular shaped, making it easy for them to travel throughout the blood vessels; however, in patients with sickle cell anemia, those cells transform and become long and thin (sickle-shaped) and are unable to move as freely within the arteries and veins.

Due to this altered shape, the cells tend to stick together, which can block small blood vessels and prevent oxygen delivery to those organs. When blockage of vessels occurs, this can cause severe pain and dysfunction, known as a sickle cell crisis. In the case of bone, this results in bone cell death, known as osteonecrosis.

Osteonecrosis (also known as avascular necrosis) of the hip joint develops when the bone in the ball of the hip joint doesn’t get enough blood flow and oxygen. Without oxygen, bone cells will die and harm the hip joint. This can happen in other joints like the knee or the ankle, but the hip joint is most commonly affected. Osteonecrosis of the hip can cause the ball (femoral head) to collapse, or cave in, which can cause severe joint pain and the early development of arthritis. Osteonecrosis is the most common reason for total hip replacement in young patients with sickle cell anemia. However, before someone has a hip replacement, treatment with medications and physical therapy are usually done to try and help with pain before considering surgery.
Advances in modern medicine have improved the management of sickle cell anemia. Prevention of further sickle cell events is the most important treatment your doctor will prescribe. This can include medications such as hydroxyurea (prevents formation of sickle-shaped red blood cells), oxygen therapy, and special blood transfusions. One such transfusion is known as apheresis, where a special machine replaces your red blood cells with healthy donor red blood cells. New emerging treatments may include bone marrow transplants, in which a matched donor’s bone marrow is transplanted into the patient; however, this option is typically limited to children. Decreasing the number and severity of sickle cell crises a person can lower the risk of osteonecrosis of the hip joint or other complications associated with the disease. The pain and functional limitation associated with osteonecrosis of the hip can be severe, which can lead patients to pursue earlier surgical intervention with hip replacement. If surgery is recommended, your doctor may ask you to obtain a complete blood count (CBC) prior to surgery to make sure that you are not anemic (low blood count) and may recommend blood transfusions before surgery to make sure that you have an adequate supply of well-functioning red blood cells to carry oxygen to your tissues during your recovery period after surgery.

Patients with sickle cell anemia who develop osteonecrosis of their hip can benefit from joint replacement as it can relieve pain and improve joint function. The need for hip replacement may come at a young age for a person with sickle cell anemia, and this raises concern that the hip replacement may eventually wear out with further surgery needed later in life (https://hipknee.aahks.org/dont-take-your-new-joint-for-granted-follow-up-care/). While modern day implants are expected to last 15-20 years, we do not fully know the longevity limits of hip replacement components in young patients (https://hipknee.aahks.org/what-is-my-hip-replacement-made-of/). It is possible that a future surgery will be necessary when the procedure is performed at a young age. It is important to discuss these topics with your orthopedic surgeon if you find yourself in need of hip replacement due to osteonecrosis from sickle cell anemia.

Image Citation:
NIH – National heart, Lung, and Blood Institute:

*Image is widely posted at various websites.