

Sickle is important to stabilize the patient to



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Sickle cell disease (SCD) is a genetic condition that is inherited when a child receives two sickle cell genes - one from each parent. Sickle cell occurs when the red blood cells are rigid and sickle-shaped, which causes them to stick together and block oxygen to the body. A large number of infants are born with SCD annually; today every newborn is screened for the sickle cell gene.

In this article the author covers the following topics: pain, acute chest syndrome (ACS), infection, stroke, and available treatment options. As stated by the author, adults and children with SCD must be continuously checked for pain with the use of the appropriate pain scale. There are several natural pain treatment approaches such as, acupuncture, physical therapy, massage, hydration, behavior modification, deep breathing, imagery, and distraction. Medicated treatment includes administration of IV fluids, opioids or nonsteroidal anti-inflammatory drugs and oxygen if hypoxia is present. Acute Chest Syndrome (ACS) is a rapid deterioration in respiratory function caused by vaso-occlusion in the vessels of the lungs. Due to severe pain and the use of narcotics as opioids, there is a potential for decreased respiratory effort, which may lead to ACS. Patients are encouraged to use an incentive spirometer to promote deep breathing, and blowing efforts for younger ages.

ACS is the most common cause of death amongst young adults. Infection is the most common condition that causes death in SCD patients called *Streptococcus pneumoniae* sepsis. Patients who experience fever should be treated as an emergency and require intervention as soon as possible, including blood cultures, complete blood count, chest radiographs, IV fluids, and IV antibiotics.

One of the great imaging tools to determine whether a child with SCD is at risk of a stroke is Transcranial Doppler, this tool is used regularly and is only to be used for children. If the stroke is suspected it is important to stabilize the patient to prevent further brain damage, and an exchange transfusion should be performed immediately. A chemotherapeutic agent that has been shown to decrease vaso-occlusive episodes and incidence of ACS is called Hydroxyurea. Hydroxyurea therapy is one of the available pain treatments, which results in less need for blood transfusion and is not used in the acute state. The bone-marrow transplant is rarely used; due to high death rate from complications in children who receive a bone marrow transplant. Finding a match through the Bone Marrow Registry is extremely difficult. Prior to surgeries patient needs to be checked for any signs of fever, infection, dehydration, and pain, which may trigger or cause a sickling crisis.

Those receiving blood transfusion may be in the risk of suffering from complications. Because SCD is a hematologic disease, it is important to track blood loss and to watch the patient's Hgb closely in case he or she requires an intraoperative or postoperative blood transfusion. Postoperative care includes controlling pain, administering supplemental oxygen if needed, and maintaining fluids. The nurse should continually monitor the patient after surgery for ACS, pain crisis, and infection. Surgical patients should be encouraged to use incentive spirometry, deep breathing, and ambulation when it is allowed.