Sickle Cell Anemia Interventions

Sickle cell anemia (SCA) is an autosomal recessive genetic disorder that causes normal hemoglobin A to be partially or completely replaced by hemoglobin S. Interventions to treat sickle cell anemia, or to lessen symptoms associated with the disease include: maintaining adequate hydration and oxygenation, analgesics and warm compresses for pain, hydroxyurea, and hematopoietic stem cell transplantation. Patients with this disease should avoid high altitudes, dehydration, and strenuous exercise, as these may induce sickling of RBCs, causing a crisis. Prophylactic antibiotics and pneumococcal and meningococcal vaccines are also strongly encouraged in these patients to reduce the risk of infection.



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Interventions

Increase Hydration

Up-arrow Water-bottle

Patients should be educated on the importance of avoiding dehydration, as this can induce a sickle cell crisis. Maintaining adequate hydration is important to decrease the viscosity of blood and to ensure proper functioning of the kidneys. In a sickle cell crisis, fluids are administered intravenously.

Analgesics

A-nail-Jay-Z

Pain management is an essential element of treatment. Pain associated with a sickle cell crisis is severe and is often treated with opioid medications such as hydromorphone, or methadone, administered via a patient controlled analgesia (PCA) pump.

Oxygen

02-tank

Oxygen can be used to treat hypoxia and prevent further sickling of RBCs. Oxygen therapy does not reverse the sickling that has already occurred, nor does it decrease pain due to sickling.

Warm Compress

Warm Compress

Warm compresses encourage blood vessels to dilate, and can be used to alleviate pain. Cold compresses or ice packs should never be used, as they can decrease circulation increasing the risk of a sickle cell crisis.

Bone Marrow Transplant

Bone Train-plant

Bone marrow transplant, also called Hematopoietic Stem Cell Transplantation (HSCT), is a popular therapy for sickle cell anemia. Hematopoietic cells give rise to all other types of blood cells in the body. HSCT involves transplanting hematopoietic cells into a patient with SCA, in an effort to restore production of non-sickled hemoglobin A.

Hydroxyurea

Hydra-U-Rainbow

Among individuals with SCA, those who have higher levels of Hgb F (fetal hemoglobin) typically have fewer complications of the disease. Hydroxyurea is a chemotherapy drug that has been clinically effective in increasing production of hemoglobin F, decreasing the reactive neutrophil count, increasing RBC volume and hydration, and altering the adhesion of sickle RBCs to the endothelium, thereby reducing complications.

Considerations

Avoid High Altitudes

Avoid-sign at High Altitudes

Patients with SCA should avoid high altitudes, as the difference in availability of oxygen may cause the patient to experience a sickling crisis.

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Prophylactic Antibiotics

Purple-axes and ABX-guy

Prophylactic administration of penicillin is recommended, starting at two months of age to reduce a child’s risk of infection. Prophylactic antibiotics can be discontinued when the child reaches five years of age.

Avoid Strenuous Exercise

Avoid-sign at Strenuous Exercise

Strenuous exercise should also be avoided. Dehydration or lack of adequate oxygenation associated with this type of exercise could induce a sickling crisis.

Vaccines

Syringe

Vaccines, such as the pneumococcal and meningococcal vaccines, are recommended for children to reduce the risk of infection. Additionally, Haemophilus influenzae, influenza, and hepatitis immunizations should be administered. Remember, individuals with SCA are at an increased risk for infection due to splenic damage.

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