Sickle Cell Anemia (Management)

Sickle cell disease management must be understood to be able to treat the many severe complications of the disease. Management can be broken into long term treatment, acute treatment, and prophylaxis. Long term includes hydroxyurea, which has been proven to decrease crisis complications and improve survival. Bone marrow transplants are the only effective cure for sickle cell disease, and folic acid administration is necessary for effective erythropoiesis. Acute management is described by an exchange transfusion, to replace sickled cells with normal red blood cells. Penicillin is used for prophylaxis in patients who are asplenic or functionally asplenic, as well as children under the age of five with this affliction.



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Long Term Treatment

Hydroxyurea
Hydra-U-Rainbow
Hydroxyurea is normally a chemotherapy medication. It is used as a causative agent to reduce the amount of painful crisis episodes and acute chest syndrome. It works by increasing fetal hemoglobin (HbF) production to decrease the relative amount of hemoglobin S (which causes sickle cell).

Bone Marrow Transplant
Bone Train-plant
Bone marrow transplants are the only known cure for sickle cell disease, and have been proven to be effective in children.

Folate
Foliage-tree
Folate administration is important for maintaining effective erythropoiesis in patients, and children born with the disease typically take 1mg of folic acid daily for life. It also helps prevent aplastic crisis.

Acute Treatment

Exchange Transfusion
Exchange Transfusion
Exchange transfusion requires that the patient’s blood be removed and replaced. This increases the proportion of normal RBCs to sickled RBCs and is considered in the management severe complications of sickle cell anemia that do not respond to fluids, analgesics and cardiac decompensation. These severe complications include acute chest syndrome, stroke and priapism.

Prophylaxis

Penicillin Prophylaxis
Pencil-villain Purple-axes
Children who have sickle cell disease and are under the age of five years are at increased risk of life-threatening pneumococcal infection due to absent or non-functional spleens and a decreased immune response against encapsulated organisms. To prevent pneumococcal infection, penicillin prophylaxis is recommended in children with sickle cell disease under the age of five or in children who have had a previous infection or have functional/surgical asplenia.

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