Inpatient Management of Sickle Cell Disease

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KEYWORDS

• Sickle cell disease • Inpatient management • Vasoocclusive crisis
• Hemoglobinopathy • Acute chest syndrome • Bone marrow transplantation

HOSPITAL MEDICINE CLINICS CHECKLIST

1. Sickle cell disease is a group of hereditary hemoglobinopathies characterized by the presence of hemoglobin S, which is caused by a valine to glutamate substitution in the gene coding for the beta globin chain.
2. The most common reason for hospitalization of patients with sickle cell disease is vasoocclusive crisis, which results from the blockage of small blood vessels by sickled cells, with pain resulting from localized ischemia, vascular endothelial damage, and inflammation.
3. Intravenous hydration and pain control with nonsteroidal antiinflammatory drugs and narcotics are the mainstays of inpatient therapy for vasoocclusive crisis.
4. Complete blood count, reticulocyte count, and basic metabolic panel should be routinely ordered on admission for vasoocclusive crisis, with urine analysis, chest radiograph, and cultures as needed for urinary, respiratory, or infectious symptoms.
5. Blood transfusions do not help with the alleviation of pain crises but are warranted in patients having end-organ damage or symptomatic anemia; the posttransfusion goal should *not* exceed hemoglobin 10% or hematocrit 30 because this can increase viscosity and worsen sickling.
6. Exchange transfusion (removing a few units of native blood and replacing them with donor blood) is helpful for patients with severe end-organ damage, such as acute chest, stroke, or multisystem organ failure.

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7. Clinically significant complications seen in inpatients with sickle cell disease include acute chest syndrome, acute stroke, leg ulcers, osteomyelitis, aplastic crisis, and hyperhemolysis.
8. Perioperative management of patients with sickle cell disease includes adequate hydration and aggressive DVT prophylaxis; preoperative transfusions are not routinely recommended but may be considered on a case-by-case basis. Common operations include cholecystectomy, tonsillectomy, and splenectomy.
9. New modalities include bone marrow transplantation in patients who suffer strokes; life-threatening complications of vasoocclusive crisis, such as large pulmonary emboli; or severely compromised quality of life.

DEFINITIONS

1. What is the definition of sickle cell disease?

Sickle cell disease (SCD) is a group of hereditary hemoglobinopathies characterized by the presence of hemoglobin S (Hgb S) caused by a single amino acid substitution in the beta globin chain. The substitution confers reduced solubility, particularly in the deoxygenated state, resulting in polymerization and deformation of the red blood cell (RBC) (sickling). SCD includes homozygous patients with hemoglobin (Hgb) SS; and compound heterozyges with other hemoglobinopathies, including Hg SC and Hgb Sβ thalassemia, which tend to have more mild disease manifestations.

2. What is a vasoocclusive crisis?

Vasoocclusive crisis (VOC), also termed sickle cell pain crisis, results from the blockage of small blood vessels by sickled cells, with pain resulting from localized ischemia. Additional contributing factors likely include associated vascular endothelial damage and inflammation. Over time, recurrent VOC with intermittent ischemia can result in chronic end-organ damage, which is also characteristic of SCD.

EPIDEMIOLOGY OF INPATIENT TREATMENT OF SCD[4]

1. What is the prevalence and life expectancy of SCD?

With advances in medical care, including early identification by newborn screening and use of prophylactic penicillin, pneumococcal vaccination and hydroxyurea, life expectancy in SCD has improved since the 1980s; 93% to 99% of patients are now expected to live beyond 20 years of age.[1] Life expectancy varies based on type, with the previously noted median age of death in Hgb SS disease of mid-40s and near-normal ranges in Hgb SCD; these numbers predate the medical advances noted earlier and likely now represent underestimates.[2] Adjusting for early mortality, US prevalence of SCD is estimated to be around 70 000 to 100 000 persons.[3,4]

2. How often are patients with SCD admitted to the hospital?

Emergency department (ED) visits and hospital admission are significantly more common in patients with SCD compared with age- and race-matched populations, with the risk of hospitalization increasing as much as 7 to 30 times baseline.[5] The