

8. SPLENIC SEQUESTRATION

Principles

- To ensure the early detection of acute splenic sequestration in patients with SCD.
- To institute urgent therapy for patients with acute splenic sequestration crises to restore intravascular volume and improve tissue oxygenation.
- To prevent recurrent episodes of splenic sequestration crises.

Recommendations

Education and Monitoring

- All parents and caregivers of children with SCD should be taught how to palpate the spleen at their first clinic visit. Teaching should be reinforced at subsequent follow-up visits.
- At clinic visits, steady state splenic size should be accurately and consistently documented by physicians or nurses.
- Palpating for the spleen must be done in the clinical assessment of any patient with SCD who presents to an emergency department.
- Patients with newly enlarged or enlarging spleen size should be assessed emergently in a health facility; urgent bloodwork for complete blood count (CBC), reticulocyte count, blood type and cross match must be performed.

Acute Management

- Immediate transfusion of packed red blood cells must be performed using guidelines for blood transfusions in patients with SCD ([see Part I, section 2 on Transfusion - Recommendations C1-2](#)).
- Exchange transfusions aimed at reducing the concentration of sickle hemoglobin (HbS) to below 30% have not been shown to be superior to simple red cell transfusions, and are not routinely recommended.
- Emergency splenectomy should be performed in patients who do not respond to red cell transfusions. Lack of response may include inability to maintain hemoglobin level despite blood transfusions, increasing splenic size, and persistent hypovolemia.

Long-term Management

- Recurrent splenic sequestration crises occur in up to 50% of patients. Following one episode of acute splenic sequestration, treatment options to prevent recurrence should be discussed with the patient and family.
- Elective splenectomy and chronic transfusion therapy are the two most common approaches to preventing recurrent episodes of splenic sequestration.¹¹⁻¹⁴ To date, there are no randomized control trials comparing each of these options. It is strongly recommended that patients consult with a hematologist specialized in the care of patients with SCD.
- Acute splenic sequestration should be considered in the differential diagnoses of any patient with SCD who presents with hypovolemic shock and exacerbated anemia.

Background

Splenic sequestration is an acute and life-threatening complication of sickle cell disease (SCD), caused by the trapping of blood within the spleen. Acute splenic sequestration may result in hypovolemic shock and death, if not rapidly recognized and treated. It is characterized by the presence of an enlarging spleen and a sudden drop in hemoglobin concentration. The reticulocyte count is usually increased, and thrombocytopenia may be present. Although most episodes occur between the ages of 6 months and 5 years, cases of acute splenic sequestration have been described in infants younger than 6 months as well as in adults. Splenic sequestration is more commonly associated with sickle cell anemia (HbSS) SCD. Patients with compound heterozygous genotypes such as hemoglobin SC disease (HbSC) and HbS-beta-thalassemia may remain at risk of this complication during adolescence and into adulthood, however, due to persistent splenic enlargement.¹⁻⁸ Patients with acute splenic

sequestration often present with sudden weakness, pallor, tachycardia, tachypnea, abdominal fullness, left upper quadrant pain, and palpable splenomegaly. Atypical symptoms such as back pain, left flank pain, chest pain, or mental obtundation may also occur.^{2,7} Acute splenic sequestration in the absence of a palpably enlarged spleen has also been reported.⁹ During an acute splenic sequestration crisis, the mean decline in hemoglobin is usually ≥ 3 g/dl from baseline.^{7,10} Patients who have had one episode of splenic sequestration are more likely to have recurrent episodes, and to have an increased risk of mortality with subsequent episodes.¹¹

References

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