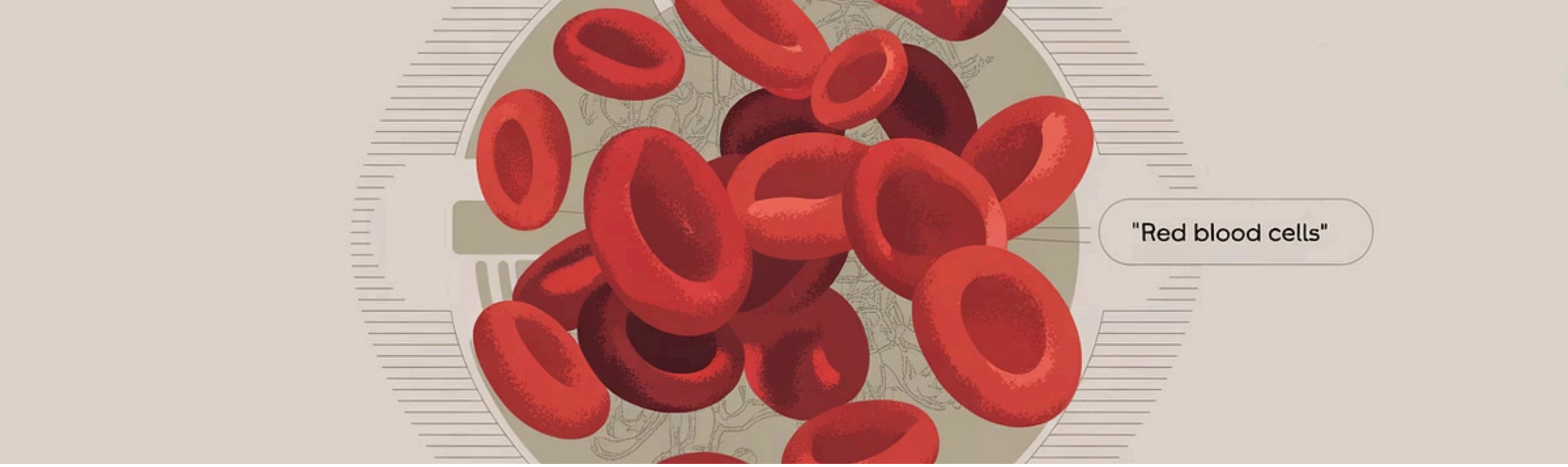

Splenic Sequestration Crisis in Sickle Cell Anaemia

Evidence-Based Clinical Practice Guide

Updated: January 2025 | For Medical Education and Clinical Practice
Based on ASH, NHLBI, and International Pediatric Guidelines

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Executive Summary & Key Clinical Points

Medical Emergency

Splenic sequestration crisis is a life-threatening emergency requiring immediate recognition and treatment

Definition

Rapid enlargement of spleen + hemoglobin drop ≥2 g/dL below baseline + reticulocytosis

High-Risk Population

Infants and children under 5 years with HbSS/HbSβ^0; adolescents with HbSC

Mortality Risk

Can progress rapidly to hypovolemic shock and death without prompt intervention

Immediate Management

IV fluids + cautious RBC transfusion in **3-5 mL/kg aliquots**

Recurrence Rate

Up to 78% requires long-term preventive strategy

Definition and Pathophysiology

Clinical Definition

Acute splenic sequestration crisis is characterized by:

- Acute enlargement of the spleen beyond baseline
- Sudden drop in hemoglobin concentration ≥2 g/dL below patient's baseline
- Reticulocytosis (equal to or greater than baseline)
- Often accompanied by thrombocytopenia

Pathophysiology

The spleen's unique anatomy and function make it particularly vulnerable in sickle cell disease. Blood flow through the red pulp is slow and hypoxic, promoting sickling of red blood cells. In sequestration crisis, sickled cells become trapped in the splenic sinusoids, creating a mechanical obstruction that propagates rapidly. This leads to:

01

Massive pooling of blood within the spleen

(up to 50% of circulating blood volume)

02

Acute reduction in circulating blood volume

03

Risk of hypovolemic shock

04

Potential cardiovascular collapse

Epidemiology and High-Risk Groups

Risk Factor	Details	Clinical Significance
Age	Peak incidence: 6 months - 5 years Median age: 1.4 years 75% occur before age 2	Risk decreases after age 5 due to autosplenectomy
Genotype	HbSS and HbSβ ^{0}-thalassemia (highest risk) HbSC (may occur in adolescence)	HbSC patients retain splenic function longer
Recurrence	50-78% recurrence rate Higher risk if first episode <1 year	Necessitates preventive strategy discussion
Prevalence	7-30% of children with SCD Can be presenting complaint in 20%	Important differential in pediatric SCD patients

Clinical Presentation

Signs and Symptoms

Classic Triad: Acute pallor + Splenomegaly + Hemodynamic instability

Presenting Features:

General

Sudden onset pallor, weakness, fatigue, irritability

Cardiovascular

Tachycardia, hypotension, poor perfusion

Abdominal

Left upper quadrant pain, abdominal distention, palpable splenomegaly

Respiratory

Tachypnea (due to anemia and hypovolemia)

Neurological

Altered mental status in severe cases

Severity Indicators

- Rapid decline in hemoglobin level from baseline.
- Pronounced splenomegaly (spleen extending beyond midclavicular line or rapidly increasing in size).
- Signs of shock: severe hypotension, marked tachycardia, prolonged capillary refill, cool extremities.
- Altered mental status, lethargy, or unresponsiveness.
- Development of acute chest syndrome concurrently.

Physical Examination & Clinical Pearls

Physical Examination Findings

Pallor: Most prominent on mucous membranes (conjunctivae) and palmar creases.

Splenomegaly: Spleen often firm and tender, extending several centimeters below the costal margin. Careful palpation is crucial to assess size and changes over time.

Cardiovascular: Tachycardia, faint heart sounds, weak peripheral pulses, hypotension.

Respiratory: Tachypnea, increased work of breathing.

Neurological: May range from irritability to coma in severe cases.

Clinical Pearls for Recognition

- Acute Splenic Sequestration (ASS) should always be considered in any child with Sickle Cell Disease (SCD) presenting with sudden pallor and splenomegaly.
- The rapid onset and progression of symptoms are characteristic; a seemingly stable child can decompensate quickly.
- Knowledge of the patient's baseline hemoglobin level is critical for interpreting the severity of anemia.
- ASS can sometimes be mistaken for an infection or other painful crisis. Persistent, significant splenomegaly with severe anemia usually points towards ASS.

Diagnostic Evaluation

Diagnostic Criteria

- Hemoglobin drop ≥2 g/dL from baseline (most important criterion)
- Acute splenomegaly beyond baseline size
- Reticulocytosis ≥ baseline levels
- Often accompanied by thrombocytopenia

Essential Laboratory Studies

- **Complete Blood Count with Differential:** Documents degree of anemia and thrombocytopenia
- **Reticulocyte Count:** Typically elevated (distinguishes from aplastic crisis)
- **Type and Crossmatch:** For potential transfusion needs
- **Baseline Labs:** Comparison with patient's steady-state values crucial
- **LDH:** May be elevated due to hemolysis

Laboratory Interpretation Guidelines

- Compare ALL values to patient's baseline, not normal ranges
- Reticulocyte count helps differentiate from aplastic crisis (low in aplastic)
- Platelet count often decreased due to splenic pooling
- Hemoglobin may drop rapidly (monitor closely)

Clinical Decision Points

- **Baseline Hemoglobin Knowledge:** Baseline hemoglobin knowledge is absolutely crucial
- **Differential Diagnosis:** Consider differential diagnosis (aplastic crisis, hypersplenism, vaso-occlusive crisis)
- **Urgent Consultation:** Urgent hematology consultation recommended
- **Time-Sensitive Diagnosis:** Time-sensitive diagnosis - don't delay treatment for extensive workup

Acute Management Protocols

The acute management of splenic sequestration crisis (SSC) is a medical emergency requiring swift and coordinated action. The primary goals are to stabilize the patient's hemodynamics, restore circulating blood volume, and correct severe anemia. Delays in diagnosis and treatment can lead to rapid deterioration and potentially fatal outcomes.

Immediate life-saving interventions are paramount. Do not delay treatment for extensive diagnostic workup.

Initial Stabilization and Resuscitation

01

Rapid Assessment

Immediately assess airway, breathing, and circulation (ABCs). Monitor vital signs, level of consciousness, and perfusion status (capillary refill, skin color).

02

Oxygen Therapy

Administer supplemental oxygen to all patients, especially if signs of hypoxemia or respiratory distress are present, to optimize tissue oxygenation.

03

Fluid Resuscitation

Initiate rapid intravenous fluid administration (e.g., normal saline or Ringer's lactate) to combat hypovolemic shock. Administer 10-20 mL/kg boluses, reassessing frequently.

04

Pain Management

Provide appropriate analgesia for abdominal pain, typically with opioids, to ensure patient comfort and reduce stress.

Transfusion Strategy for Pediatric Patients

Managing acute anemia in pediatric patients, particularly those with conditions like sickle cell disease experiencing a sequestration crisis, requires a carefully planned transfusion strategy. The goal is to restore adequate oxygen-carrying capacity without inducing complications such as hyperviscosity or circulatory overload.

Initial Dose

Initiate with a conservative dose of **3-5 mL/kg of packed RBCs**. This measured approach is crucial to prevent rapid overtransfusion, especially since splenic sequestration can release a significant volume of red blood cells back into circulation. Overtansfusion can lead to hyperviscosity, exacerbating vaso-occlusive crises, and circulatory overload, potentially causing cardiac strain.

Target Hemoglobin

Aim for a target hemoglobin (Hb) level of **8-10 g/dL**, strictly avoiding levels exceeding 10 g/dL. This range optimizes oxygen delivery to tissues while minimizing the risk of hyperviscosity. High Hb levels increase blood viscosity, which can worsen sickling and trigger new vaso-occlusive events or stroke, particularly in patients with sickle cell disease.

Reassessment Protocol

Perform frequent clinical reassessments and check Hb levels after **each aliquot of transfusion**. This close monitoring is vital to track the patient's response and prevent inadvertent hyperviscosity or overcorrection as sequestered red blood cells may be re-mobilized. Look for clinical improvement (e.g., improved perfusion, decreased respiratory distress) alongside laboratory values.

Exchange Transfusion

Consider exchange transfusion if simple transfusion fails to achieve clinical improvement or if the patient presents with severe complications like acute chest syndrome, stroke, or multi-organ failure refractory to conventional management. Exchange transfusion removes sickled cells and replaces them with healthy red blood cells, effectively reducing the percentage of HbS and viscosity.

Long-Term Management & Prevention

Prevention of Recurrence

Given the high recurrence rate (50-78%), all patients require discussion of long-term preventive strategies:

Strategy	Advantages	Disadvantages	Considerations
Splenectomy	• Eliminates recurrence risk • Definitive solution • Improves baseline Hb	• Increased infection risk • Surgical risks • Loss of immune function	Recommended after recurrent or life-threatening episodes
Chronic Transfusion	• Preserves splenic function • Reduces sickling • Reversible	• Iron overload • Alloimmunization • Vascular access issues	Temporary bridge to splenectomy or until autosplenectomy
Expectant Management	• Avoids surgery/transfusion risks • Natural autosplenectomy	• High recurrence risk • Requires excellent education • Emergency access essential	Only with exceptional family education and access

Parental Education is Critical

Early recognition and prompt medical attention can be lifesaving.

Warning Signs Requiring Emergency Evaluation

- Sudden onset of pallor or weakness
- Abdominal pain, especially left-sided
- Enlarged spleen from baseline
- Difficulty breathing or rapid heart rate
- Unusual fatigue or sleepiness
- Decreased activity level

Key Takeaways & Call to Action

Splenic Sequestration Crisis in Sickle Cell Anaemia demands rapid identification and immediate intervention. Early recognition of warning signs and prompt medical attention are crucial for saving lives and preventing long-term complications.

- **Recognize Early:** Be aware of sudden pallor, weakness, or increased spleen size.
- **Act Fast:** Seek emergency care immediately for suspected SSC.
- **Educate & Prevent:** Understand recurrence risks and long-term management options.

Share this post to raise awareness and help save lives!

References

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1. **Yates AM, Aygun B, Nuss R, Rogers ZR.** Health Supervision for Children and Adolescents With Sickle Cell Disease. *Pediatrics.* 2024 Jul 31. **PubMed PMID: 39034826**
2. **National Heart, Lung, and Blood Institute.** Evidence-Based Management of Sickle Cell Disease: Expert Panel Report, 2014. **Official PDF**
3. **Brousse V, Elie C, Benkerrou M, et al.** Acute splenic sequestration crisis in sickle cell disease: cohort study of 190 pediatric patients. *Br J Haematol.* 2012 Mar 12;156(5):433-8. **PubMed PMID: 22224796**
4. **Kavanagh PL, Fasipe TA, Wun T.** Sickle cell disease: a review. *JAMA.* 2022;328(1):57-68. **PDF**
5. **American Society of Hematology.** ASH Pocket Guides: Sickle Cell Disease (2020 update; originally 2014). **ASH Pocket Guides Site**
6. **Ben Khaled M, Ouederni M, Mankai Y, et al.** Prevalence and predictive factors of splenic sequestration crisis among 423 pediatric patients with sickle cell disease in Tunisia: a retrospective cohort study. *Blood Cells Mol Dis.* 2020 Jan;80:102374. **Wiley PDF**
7. **Serjeant GR, Mason KP, Hambleton IR, Serjeant BE.** Acute splenic sequestration in HbSS: observations from the Jamaican birth cohort. *Arch Dis Child.* 2024 Jan 21. **PubMed PMID: 37918896**
8. **George A, Conneely SE, Mangum R, et al.** Splenic Complications in Pediatric Sickle Cell Disease: A Retrospective Chart Review. *Preprint* (2024). **Preprint Full Text**
9. **Owusu-Ofori S, Remmington T.** Interventions for preventing splenic sequestration in sickle cell disease. *Cochrane Database Syst Rev.* 2017;7:CD011365. (*Not directly found in this search, but this is the standard citation for the review.*)