IWK Health	Document Scope: Hospital-wide Patient Care	
	Document Type: Clinical Practice Guideline Approved on: January 2024 Next Review Date: January 2027	
	Acute Splenic Sequestration: Guidelines for Management in Children with Sickle Cell Disease (Adapted from the Hospital for Sick Children)	Version: 1

1.0 Background

Although, the spleen does not properly perform its filtration function in patients with sickle cell disease, its reservoir function is overactive: sequestration of large quantities of blood (often half or more of a child's blood volume) can occur rapidly. This complication, termed acute splenic sequestration, is characterized by pooling of large quantities of sickled RBCs in the splenic red pulp, sudden enlargement of the spleen (within hours), and a precipitous decline in hemoglobin (Hb) and platelets, and an increase in reticulocytes.

Presentation is often (60%) associated with episodes of fever, suggesting an underlying viral etiology. Most commonly occurs in infants and young children between 6 months and 5 years of age with sickle cell anemia. It may also occur in older patients with any sickle cell phenotype with or without chronic splenomegaly. Often there is no obvious triggering event.

2.0 Clinical/Laboratory Features

A child with an acute splenic sequestration presents with symptoms of:

- acute anemia (pallor, tachycardia, frank cardiovascular collapse);
- splenomegaly/abdominal pain (pain in the left upper quadrant); and
- evidence of an active bone marrow response (increased reticulocytes) plus or minus thrombocytopenia.

Retrospective reviews have shown a first-episode mortality of as high as 14%. On physical examination, patients show signs of anemia, hypovolemia, and an enlarged spleen (larger than baseline), sometimes massively so. Mild cases may be asymptomatic.

Hemoglobin concentration is at least 20g/L below the baseline steady state. In severe cases, hemoglobin may decline to life-threatening levels. Reticulocyte counts are usually elevated, which distinguishes this condition from aplastic crisis. The platelet count often declines to <50 X 10^9/L.

The mainstay of management is transfusion to provide circulating erythrocytes and volume. Risk of recurrence is approximately 40–50%, usually within 3 years. Because it is not possible to predict which children will have recurrent attacks, most experts recommend splenectomy after the first major attack (for patients >2 years old), or chronic transfusion to maintain a hemoglobin S level under 50% until the patient can get to surgery once all relevant immunizations have been completed.

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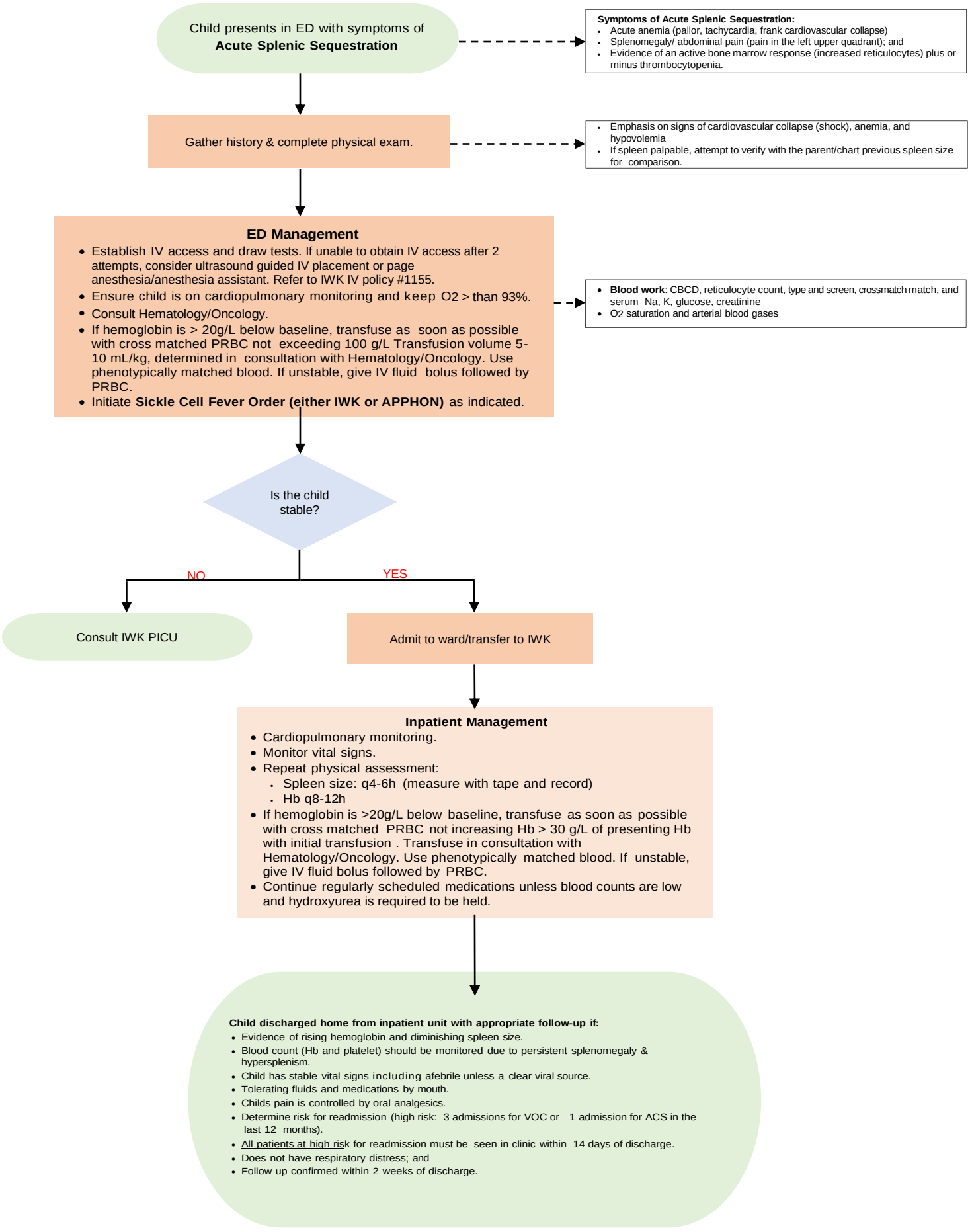

4.0 References

1. Emond AM, Collis F, Darvill D, et al. Acute splenic sequestration in homozygous sickle cell disease: natural history and management. *J Pediatr*. 1985;107(2):201–06.
2. National Heart, Lung, and Blood Institute. Evidence-based management of sickle cell disease: Expert panel report, 2014.
3. Powell RW, Levine GL, Yang YM, et al. Acute splenic sequestration crisis in sickle cell disease: early detection and treatment. *J Pediatr Surg*. 1992;27(2):215–19.