

Table 4: Evidence Supporting Anticipatory Guidance for Splenic Complications in Children with Sickle Cell Disease

TYPE OF EVIDENCE	KEY FINDINGS	LEVEL OF EVIDENCE (USPSTF RANKING*)	CITATION(S)
Clinical guidelines	<p>The NHLBI suggests the following regarding awareness of splenic complications:</p> <ul style="list-style-type: none"> • Infants with Hb FS should be started on prophylactic penicillin by 2 months of age, and parents should be educated about the importance of urgent medical evaluation and treatment for febrile illness and for signs and symptoms indicative of splenic sequestration. • Early education should be provided to parents of infants with SCD regarding palpation of the spleen, symptoms of progressive anemia, and appropriate action for obtaining rapid evaluation and treatment. • Patients who have a life-threatening episode of acute splenic sequestration that requires transfusion support should have a splenectomy shortly after the event or be placed on a chronic transfusion program. • Alternatively, patients younger than 2 years of age who have a severe episode of acute splenic sequestration should be placed in a chronic transfusion program to keep Hb S levels below 30% until a splenectomy can be considered after 2 years of age. • Patients with chronic hypersplenism (an overactive spleen) also should be considered for splenectomy. 	III	National Heart, Lung and Blood Institute. The Management of Sickle Cell Disease. National Institutes of Health. Bethesda, MD, 2002.
Clinical guidelines	<p>The AAP sections on Hematology/Oncology and the Committee on Genetics suggest the following:</p> <ul style="list-style-type: none"> • Education about the need for urgent medical evaluation for and treatment of febrile illness and acute splenic sequestration is critical. • The goal of ensuring timely medical treatment for acute illness also is facilitated by providing anticipatory guidance to patients and families about early recognition, appropriate medical evaluation, and treatment of common acute complications. • Prompt recognition and treatment with red blood cell transfusions may be lifesaving in 	III	American Academy of Pediatrics Section on Hematology/Oncology and Committee on Genetics. Health supervision for children with sickle cell disease. <i>Pediatrics</i> 2002; 109(3):526-535.

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	<p>cases of acute splenic sequestration. Surgical splenectomy to prevent recurrence is often recommended after recovery from life-threatening or recurrent episodes of sequestration.</p> <ul style="list-style-type: none"> • Health supervision for infants and young children should include ongoing discussion about the signs and symptoms of splenic sequestration, awareness of changes in spleen size, and the importance of evaluating febrile illnesses to uncover infection. 		
Clinical guidelines	<ul style="list-style-type: none"> • All patients with SCD should have regularly scheduled comprehensive medical evaluations, which also offer an ideal setting for providing age-appropriate family and patient education. • Acute splenic sequestration is a severe illness associated with hemoglobin that is 2 g/dL or more below the patient's baseline value with an acutely enlarged spleen. Hospitalization and transfusions are part of care. Surgical splenectomy and or/chronic transfusions should be considered for severe or recurrent events. 	III	Lane PA, Buchanan GR, Hutter JJ, et al. Sickle cell disease in children and adolescents: diagnosis, guidelines for comprehensive care, and care paths and protocols for management of acute and chronic complications. 2001; Annual Meeting of the Sickle Cell Disease Care Consortium, Sedona, AZ
Clinical guidelines	<ul style="list-style-type: none"> • Early in life, when the risk of infection is highest, counseling parents about the importance of immunizations and antibiotic prophylaxis, the detection of an enlarging spleen, and the dangers of fever and increasing pallor may be lifesaving. • Splenic atrophy and dysfunction underlie the childhood propensity for infection with encapsulated bacteria. • Urgent replacement of blood is often required for sudden severe anemia occurring in children when blood is sequestered in an enlarged spleen. 	III	Steinberg MH. Management of sickle cell disease. <i>N Engl J Med</i> 1999; 340(13):1021-1030.
Prospective cohort study	<ul style="list-style-type: none"> • In a cohort of children studied in Jamaica, an education program designed to enlist the skills and cooperation of parents with instruction in splenic palpation was followed by an increased incidence of reported episodes of splenic sequestration and a decrease in mortality. • The greater number of reported cases likely resulted from an increased awareness of the diagnosis; the decrease in the number of 	III	Emond AM, Collis R, Darvill D, Higgs DR, Maude GH, Serjeant GR. Acute splenic sequestration in homozygous sickle cell disease: Natural history and management. <i>J Pediatr</i> 1985; 107:201-206.

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	deaths was consistent with both improved management and earlier diagnosis, allowing more time for		

Note: USPSTF criteria for assessing evidence at the individual study level are as follows: I) Properly powered and conducted randomized controlled trial (RCT); well-conducted systematic review or meta-analysis of homogeneous RCTs. II) Well-designed cohort or case-control analytic study. III) Opinions of respected authorities, based on clinical experience; descriptive studies or case reports; reports of expert committees.