

Suzie A. Noronha, MD¹ Sophie Lanzkron, MD² ¹ University of Rochester Medical Center ² Johns Hopkins University School of Medicine

November 2014

Health Maintenance and Management of of Sickle Cell Disease



Screening and Follow-Up for Common Complications of Sickle Cell Disease (SCD), cont'd

Complication	Screening	Follow-Up
Pulmonary complica- tions	Assess for signs and symptoms of respiratory problems (such as asthma, chronic obstructive pulmonary disease, restrictive lung disease, or obstructive sleep apnea) by history and physical examination <i>(Consensus–Expert Panel)</i> .	If signs or symptoms are present, further assess including with pulmonary function tests <i>(Consensus–Expert Panel)</i> . In individuals with symptoms or signs suggestive of pulmonary hypertension (eg, dyspnea on exertion, lower extremity edema), refer for echocardiography <i>(Strong recommendation, moderate-quality evidence)</i> . If tricuspid regurgitant velocity is elevated 2.5 m/sec by echocardiography, consult a provider with expertise in pulmonary hypertension to guide further assessment and management, including right heart catheterization and consideration of pulmonary hypertension therapy <i>(Consensus–Expert Panet)</i> .
Renal complications	 Beginning at age 10, screen annually for microalbuminuria and proteinuria with spot urine to estimate protein/creatinine ratio (Consensus–Expert Panel). Hyposthenuria, or inability to concentrate urine, is common, resulting in increased risk of dehydration and enuresis. Between 4 and 18% of people with SCD will develop chronic kidney disease (CKD). Early identification is important. Significant impairment may be masked because of hypersecreted creatinine. Once the serum creatinine rises, it often reflects significant renal dysfunction. Microalbuminuria is most often the first manifestation of CKD in SCD. 	Panel). If microalbuminuria or macroalbuminuria is identified, order a 24-hour urine test for protein (Consensus-Expert Panel). Refer to or consult a nephrologist: • Adults or children with proteinuria (>300 mg/24 hours) (Strong recommendation, low-quality evidence) • Children with microalbuminuria (Consensus-Expert Panel)



Care Coordination