Adult Sickle Cell Health Maintenance Recommendations

In general, both specialist and PCP visits at least twice a year. All elements of routine health maintenance, including routinely recommended vaccines, in addition to sickle cell specific care. All elements of care could be done by either specialist or PCP with communication of results.

	Care Element	Suggested Frequency, Guidance
Utilization	ED Visits and Hospital Admissions (Provider Portal)	Each visit
	Specialist or other outpatient visits (Provider Portal)	Each visit
Pain	Acute or chronic pain	Each visit, Increasing frequency or severity or chronic pain, See Pain algorithm
	Priapism	Each visit, Painful erection or lasting >2 hr, Pain algorithm
	Pain medications used (CSRS, Provider Portal)	Each visit, Assess for multiple unexpected fills from multiple providers, Pain algorithm
	Pain Agreement Plan	Each visit, Recommended for patients with chronic pain, Pain algorithm
	If long acting pain meds, pain provider?	Each visit, one main pain provider preferred, Pain algorithm
History	Hydroxyurea	Each visit, Typically prescribed by specialist. Generally recommended for Hg SS and SBthal0 with 3+ painful crises a year, ACS, severe anemia, etc. CBC every 4-12 weeks if on hydroxyurea.
	Receiving chronic transfusion	Each visit, Typically managed by specialists. Marker of severe disease. Risk of liver and heart failure with significant long standing iron overload (Hydroxyurea algorithm)
	Vision	Risk of retinal infarction. Refer to ophthalmologist for concerns
	Shortness of breath	Each visit, Danger of comorbidities of asthma and sickle cell disease. Hypoxia can lead to sickle cell crises. Good asthma control important in sickle cell disease patients
	Depression screen: PHQ-2/PHQ-9	Each visit, Manage depression or link to behavioral health specialists in your area
Physical	Vision	Vision screen each visit, Comprehensive eye exam annually, Risk of retinal infarction. Refer to ophthalmologist for concerns
	Spleen	Each visit, If palpable and enlarged, consider splenic sequestration, requiring hospitalization and transfusion
	Liver and gall bladder	Each visit, if painful or enlarged, consider gall stones which are associated with increased risk of multi-organ failure. Refer to Sickle Cell specialists
	Hip/knee/shoulder pain	Each visit, See Pain algorithm
	Leg ulcers	Each visit, possible etiology venous insufficiency, Initial standard therapy (debridement, wet to dry dressings, topical agents), risk of osteomyelitis with chronic recalcitrant deep ulcers, wound culture if infection is suspected or if ulcers deteriorate
	ENT exam, enlarged tonsils with s/sxs of OSA	Each visit, OSA associated with night time hypoxia, ACS, progression of CNS and peripheral vascular disease. Refer to ENT and Sickle Cell specialist
	Dentition	Each visit, Dental caries has risk of chronic seeding and inflammation that can worsen chronic pain and trigger acute pain. Refer to dentist.
	EKG to assess QTc if on methadone	At least annually, If QTc > 400ms, wean down Methadone
Labs	CBC with retic*	Every 6 months, Typically done by specialist. Red flag Hg < 2 g/dl or < 6g/dl than baseline. Risk of acute splenic sequestration, aplastic episode, ACS, etc. Refer for emergency care.
	Ferritin	Typically done by specialist annually, red flag >1000 ng/m, Ensure sickle cell specialists has results
	T. Bili, LDH	Typically done by specialist every 6 months. Red flag significantly elevated above baseline. Ensure specialist has results.
	AST, ALT, Cr	Typically done by specialist every 6 months, Red flag elevated above normal. Ensure specialist has results.
	UA. First morning void spot micro-albumin/Cr ratio, if UA+	Typically done by specialist annually. Red flag >30mg albumin/g Cr. Ensure specialist has results.
Health Maintenance	Transition to adult care	Each visit for patients age 18-22 years, Counseling about school, college, working
	PCN	If hx of surgical splenectomy or pneumococcal sepsis, at least to age 21, consider longer
	Pneumococcal vaccine (PPSV23)	If confirmed that 2 doses were given during childhood, give another dose after age 65 yrs. If confirmation of earlier doses unavailable, give one dose and then repeat after age 65 years.
	Meningococcal vaccine (MCV4, Menactra)	Every 5 years

* To establish baseline, consider doing Hg levels monthly for 3 months

Developed by the NC Division of Public Health; the comprehensive sickle cell centers at Carolinas Health Care, Duke University, East Carolina University, University of North Carolina at Chapel Hill, Mission, and Wake Forest University; and primary care physicians from across North Carolina. Adapted from the 2014 NIHLBI guidelines for Evidence-Based Management of Sickle Cell Disease.