

Pediatric Sickle Cell Health Maintenance Recommendations

PCP: Visits on routine periodicity until 2 years, then at least 2 times a year. **Specialist: Visits every 3 months until 12 months of age and then at least annually.**
 All elements of well child visits. All routinely recommended vaccines, including Prevnar and Influenza. Each element can be performed in conjunction with the specialist.

When to Start	Care Element to Assess	Guidance
2 months	Painful crises	See acute and chronic pain algorithm
	Any medication used for pain-OTC/Rx	Assess proper dose and frequency
	Vision problems by history	Risk of retinal infarction. Refer to ophthalmologist for concerns.
	Breathing problems/wheezing	Asthma control important in sickle cell disease patients. Hypoxia can lead to sickle cell crises.
	Spleen	If palpable and enlarged, consider splenic sequestration requiring hospitalization and transfusion
	Neurological exam	If deficits noted, see neurological algorithm
	PCN	All: 2 mos-3yrs at 125 mg bid. SS, Sβthal: Continue to 5yrs at 250 mg bid. If hx of surgical splenectomy or pneumococcal sepsis, 250mg bid up to at least age 21, consider longer.
Meningococcal Vaccine	MenHibRix or Menveo recommended 2, 4, 6, and 12 months. MCV4/Menactra at age 2 and then every 5 years. See CDC/ACIP recommendations at http://www.cdc.gov/vaccines/hcp/acip-recs/vacc-specific/mening.html	
12 months	Dentition/Dental Caries	Risk of chronic seeding and inflammation, can worsen chronic pain and trigger acute pain. Refer to dentist.
	Hydroxyurea	Typically prescribed and monitored by specialist. Generally recommended for all Hg SS and Sβthal0 starting at 9 months. CBC typically every 4-6 weeks if on hydroxyurea. Can be done in conjunction with specialist. (Hydroxyurea algorithm)
	CBC with retic ^o	Can be done in conjunction with specialist annually. Red flag Hg < 2 g/dl or < 6g/dl than baseline. Risk of acute splenic sequestration, aplastic episode, ACS, etc. Refer for emergency care.
	T. Bili, LDH	Can be done in conjunction with specialist annually. Red flag, significantly elevated above baseline.
	AST, ALT, Cr	Can be done in conjunction with specialist annually. Red flag, elevated above normal.
	Receiving Chronic transfusions	Typically done by specialist. Marker of severe disease. Risk of liver and heart failure with significant long standing iron overload. Red Cell antibody screen if within 4-12 weeks of a transfusion.
2 yrs	Priapism	Painful erection or lasting >2 hr, needs immediate emergency care
	ENT exam/Enlarged tonsils with sx's of OSA	OSA associated with night time hypoxia, ACS, progression of CNS and peripheral vascular disease. Refer to ENT and Sickle Cell specialist
	Pneumococcal Vaccine (PPSV23)	Often done by specialist. Age 2 and age 7.
	Trans-cranial Doppler	Done by specialist. Annually until 16 yrs for Hg SS, Sβthal. Per specialist's recommendations for HgSC, Sβthal.
3 yrs	Vision screening	Risk of retinal infarction. Refer to ophthalmologist for concerns.
	Ferritin	Can be done in conjunction with specialist annually. Red flag >1000 ng/mL. Ensure specialists has results.
6-9 yrs	Hip/knee/shoulder pain	See pain algorithm
	Behavioral Health Screen - PSC/PSC-Y	If positive, consider secondary screens and link to behavioral health specialists in your area
10 yrs	Comprehensive eye exam	Done by ophthalmologist annually. Risk of retinal infarction.
	Any medication used for pain-OTC/Rx, including narcotics	Chronic narcotics have risk of estrogenization/delayed puberty in males
	Growth/pubertal delays	Chronic disease and chronic narcotics can contribute. Consider referral to endocrinologists if significant delays
	UA. First morning void spot micro-albumin/Cr ratio, if UA+	Can be done in conjunction with specialist annually. >30mg albumin/g Cr. Ensure specialist has results.
13 yrs	Reproductive Health	Hydroxyurea is a teratogen. Reliable birth control (e.g. LARC) important if sexually active while on hydroxyurea. Progesterone-only contraception may be preferable. Importance of knowing partners' hemoglobin genotype for genetic counseling.
	Transition to adult care	Counseling about school, college, work, finding adult care.