

## Guidelines for Preventive Health Maintenance of Sickle Cell Patients

Health Maintenance Recommendations			
<b>Patient/Parent Education</b>	<ul style="list-style-type: none"> <li>Physical assessment skills (e.g. palpation of spleen)</li> <li>How to avoid vaso-occlusive complications and how to treat pain</li> <li>When to administer prophylactic antibiotics</li> <li>Importance of taking prompt action at the first sign of fever and other signs of infection</li> </ul>		
<b>Routine Clinical Laboratory Evaluations in children</b>	<b>Test</b>	<b>Age</b>	<b>Frequency</b>
	CBC w/ WBC differential, reticulocyte count	<ul style="list-style-type: none"> <li>3 mo—24 mo</li> <li>&gt;24 mo</li> </ul>	<ul style="list-style-type: none"> <li>every 3 months</li> <li>every 6 months</li> </ul>
	Percent Hb F	<ul style="list-style-type: none"> <li>6 mo—24 mo</li> <li>&gt;24 mo</li> </ul>	<ul style="list-style-type: none"> <li>every 6 months</li> <li>annually (may vary based on clinical course)</li> </ul>
	Renal function (creatinine, BUN, urinalysis)	≥ 12 mos	annually
	Hepatobiliary function (ALT, fractionated bili)	≥ 12 mos	annually
	Pulmonary function (transcutaneous O <sub>2</sub> saturation)	≥ 12 mos	every 6 mo (may vary based on clinical course)
<b>Pneumococcal Immunization in children</b>	<b>Previously Unvaccinated Children w/ Sickle Cell Disease</b>		
	<b>Product</b>	<b>1<sup>st</sup> Dose</b>	<b>Primary Series</b>
	PCV7 (Prevnar)	2-6 mo	3 doses 6-8 wk apart
		7-11 mo	2 doses 6-8 wk apart
		≥ 12 mos	2 doses 6-8 wk apart
	PPV23 (Pneumovax)	≥ 24 mo	1 dose at least 6-8 wk after last dose of PCV7 dose
			Additional doses
			1 dose at 12 to <16 mo
			1 dose at 12 to <16 mo
			1 dose, 3-5 yr after 1 <sup>st</sup> PPV23 dose
<b>Previously Vaccinated Children w/ Sickle Cell Disease</b>			
<b>Age</b>	<b>Previous Dose</b>	<b>Recommended</b>	
12-23 mo	Incomplete primary PCV7	2 doses PCV7, 6-8 wk apart	
≥ 24 mo	4 doses PCV7	1 <sup>st</sup> dose PPV23, 6-8 wk after PCV7 2 <sup>nd</sup> PPV23 dose, 3-5 yr after 1 <sup>st</sup> PPV23	
	1-3 doses PPV7 (before 24 mo of age)	1 dose PPV7 1 <sup>st</sup> dose PPV23, 6-8 wk after PCV7 2 <sup>nd</sup> PPV23 dose, 3-5 yr after 1 <sup>st</sup> PPV23	
	1 dose PPV23	2 doses PCV7, 6-8 wk apart, 1 <sup>st</sup> dose given at least 8 wk after PPV23 dose; 2 <sup>nd</sup> PPV23 dose, 3-5 yr after 1 <sup>st</sup> PPV23	
<b>Penicillin Prophylaxis in children</b>	Penicillin is given twice daily from as early as 2 months of age, a treatment supported by the hallmark Penicillin Prophylaxis Studies of the 1980s: <ul style="list-style-type: none"> <li>Penicillin VK: 125 mg by mouth twice daily for those under 3 years of age</li> <li>Penicillin VK: 250 mg twice daily for those 3 and older (up to age 5 years old)</li> <li>Alternative to oral is an injection: 1.2 million units of long-acting Bicillin<sup>TM</sup> every 3 weeks</li> <li>For children allergic to penicillin, erythromycin ethyl succinate (20mg/kg) divided into 2 daily doses can provide adequate prophylaxis</li> </ul>		
<b>Evaluation of Fever in children</b>	<ul style="list-style-type: none"> <li>All children with SCD who have a fever (&gt;38.5C or 101F) and other signs of infection (chills, lethargy, irritability, poor feeding, vomiting) should be evaluated promptly.</li> <li>Instruct parents not to treat with antipyretics at home and wait for recurrence or persistence of fever. Child should be evaluated immediately upon presentation of 2/3 of infection</li> </ul>		
<b>Anemia Prevention in children and adults</b>	<ul style="list-style-type: none"> <li>In surgical settings, simple transfusions to increase hemoglobin levels to 10g/dl are as good as or safer than aggressive transfusions to reduce sickle cell hemoglobin (Hb S) levels to below 30 percent.</li> <li>Transfusions to maintain a hematocrit of more than 36 percent do not reduce complications of pregnancy</li> <li>Transfusions to reduce Hb S levels to below 30 percent prevent strokes in children with high central nervous system blood flow [evidence from the Stroke Prevention Trial in Sickle Cell Anemia (STOP I)].</li> </ul>		
<b>Crisis Prevention in Adults</b>	<ul style="list-style-type: none"> <li>Hydroxyurea decreases crisis in patients with severe sickle cell disease [evidence from the Multicenter Study of Hydroxyurea in Sickle Cell Anemia (MSH trial)].</li> </ul>		

References: National Institutes of Health, National Heart, Lung, and Blood Institute. The Management of Sickle Cell Disease. NIH Publication No. 02-2117. Revised June 2002. Fourth Edition. Accessed on 06/01/2015 from [http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc\\_mm.pdf](http://www.nhlbi.nih.gov/health/prof/blood/sickle/sc_mm.pdf)