ABSOLUTE TOTAL CARE TOTAL™

Guidelines for Preventive Health Maintenance of Sickle Cell Patients

Health Maintenance Recommendations

Patient/Parent Education

- Physical assessment skills (e.g. palpation of spleen)
- How to avoid vaso-occlusive complications and how to treat pain
- When to administer prophylactic antibiotics
- Importance of taking prompt action at the first sign of fever and other signs of infection

Routine Clinical Laboratory Evaluations in children

Test	Age	Frequency
CBC w/ WBC differential, reticulocyte count	• 3 mo—24 mo • >24 mo	• every 3 months • every 6 months
Percent Hb F	• 6 mo —24 mo • >24 mo	• every 6 months • annually (may vary based on clinical course)
Renal function (creatinine, BUN, urinalysis)	• ≥ 12 mos	• annually
Hepatobiliary function (ALT, fractionated bili)	• ≥ 12 mos	• annually
Pulmonary function (transcutaneous 0_2 saturation)	• ≥ 12 mos	• every 6 mo (may vary based on clinical course)

Pneumococcal Immunization in children

Previously Unvaccinated Children w/ Sickle Cell Disease

Product	1st Dose	Primary Series	Additional doses
PCV7 (Prevnar)	2-6 mo	3 doses 6-8 wk apart	1 dose at 12 to <16 mo
	7-11 mo	2 doses 6-8 wk apart	1 dose at 12 to <16 mo
	≥ 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	1 dose, 3-5 yr after 1st PPV23 dose

Previously Vaccinated Children w/ Sickle Cell Disease

Age	Previous Dose	Recommended
12-23 mo	Incomplete primary PCV7	2 doses PCV7, 6-8 wk apart
≥ 24 mo	4 doses PCV7	1st dose PPV23, 6-8 wk after PCV7 2nd PPV23 dose, 3-5 yr after 1st PPV23
	1-3 doses PPV7 (before 24 mo of age)	1 dose PPV7 1st dose PPV23, 6-8 wk after PCV7 2nd PPV23 dose, 3-5 yr after 1st PPV23
	1 dose PPV23	2 doses PCV7, 6-8 wk apart, 1st dose given at least 8 wk after PPV23 dose; 2nd PPV23 dose, 3-5 yr after 1st PPV23

Penicillin Prophylaxis in children

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

- Penicillin VK: 125 mg by mouth twice daily for those under 3 years of age
- Penicillin VK: 250 mg twice daily for those 3 and older (up to age 5 years old)
- Alternative to oral is an injection: 1.2 million units of long-acting Bicillin™ every 3 weeks
- For children allergic to penicillin, erythromycin ethyl succinate (20mg/kg) divided into 2 daily doses can provide adequate prophylaxis

Evaluation of Fever in children

- All children with SCD who have a fever (>38.5C or 101F) and other signs of infection (chills, lethargy, irritability, poor feeding, vomiting) should be evaluated promptly.
- 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 s/s of infection

Anemia Prevention- in children and adults

- 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.
- Transfusions to maintain a hematocrit of more than 36 percent do not reduce complications of pregnancy
- 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)].

Crisis Prevention in Adults

- Hydroxyurea decreases crisis in patients with severe sickle cell disease [evidence from the Multicenter Study of Hydroxyurea in Sickle Cell Anemia (MSH) trial)].

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.
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