Routine Health Care Maintenance of Pediatric Patients with Sickle Cell Diseases

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This document is intended to identify routine health maintenance issues important in the care of children with sickle cell hemoglobinopathies and to aid primary care physicians and hematologists who together provide the medical home for these patients. This is meant as a supplement to, not a substitute for, age-appropriate routine health maintenance for children and adolescents.

I. Visit Frequency with Comprehensive Hematology Program:

First 24 months of life	q 2-4 months
≥ 2 years – 12 years	q 6 months
> 12 years	q 6-12 month

** More frequent visits may be required for patients with increased educational needs, accumulated complications, and therapeutic monitoring (e.g. hydroxyurea and chronic transfusion therapy).*

II. Elements of Comprehensive Visits

Should include, but not be limited to:

- Medication Review: including prophylactic medication and home pain plan
- Interval History: Inquire about fever, painful episodes, respiratory symptoms, priapism, neurological symptoms, splenic sequestration, nocturnal enuresis, snoring, ED visits, admissions, transfusions and missed school
- Physical Examination:
- Educational Review: Should begin from infancy and be reinforced at each visit. Document topics covered and remaining educational needs. As child matures, begin similar curriculum with them with goal of adolescent understanding all topics at age of transition.

Education Topics

General Information	Health Maintenance	Acute Episodes	Treatments	Psychosocial
o Introduction o Genetics o Growth & Development o Prognosis o Role of Primary and Specialty Care	o Penicillin o Immunizations o Nutrition o TCD Screening o Contraception o Hydration o Nocturnal Enuresis o Smoking o Pain Prevention o Anemia o Dental Care o Vision exams	o Access to Care o Fever o VOC and Home Management o Acute Chest o Splenic Sequestration o Aplastic Crisis o Stroke o Priapism o AVN o Gallstones o Leg Ulcers	o Blood Transfusions o Hydroxyurea o Chronic Transfusion o Bone Marrow Transplant o Iron Chelation	o Parenting a Child with a Chronic Illness o Child Care o Education and Educational Advocacy o Transition to Adult Care o Vocational Issues o Fears of Addiction o Chronic Pain o Drug and Alcohol Use o Depression and Anxiety

III. Laboratory Monitoring

CBC with reticulocyte count	Within first year of life and q year thereafter
Quantitative electrophoresis	Repeat between 1 and 2 years of age Family studies and/or DNA-based testing if needed for clarifying diagnosis or genetic counseling
RBC antigen testing	Between 1 and 2 years of age, or before first transfusion
LFTs/Bili/Renal	Annually
Urinalysis	Annually in older children and adolescents

IV. Medications

Penicillin	Birth - 36months	125mg PO BID
Penicillin	3 y.o. - 5 y.o.	250mg PO BID
Penicillin	>5 y.o.	If surgically splenectomized continue 250 mg PO BID indefinitely, otherwise may discontinue.
Erythromycin ethyl succinate	For patients with penicillin allergy	~20 mg/kg divided into BID dosing
Folic Acid	Not necessary for all patients*	400 mcg – 1 mg PO QD
Hydroxyurea	For select patients	To be prescribed by hematologist only.

** Prescribe for adolescent females, pregnant patients, those with elevated reticuocytosis (retic > 10%), and those with diets low in folic acid*

V. Screening

Pulmonary Function Tests (PFTs)	SaO2 q visit Baseline PFT when adolescent, earlier if clinical concern • Consider repeating after severe or recurrent ACS • Repeat annually if persistent RAD or if previous year abnormal Second baseline when young adult
EKG and echocardiogram	CXR, EKG and echo only if clinical concern including unusual murmur, hx of fluid intolerance or significant pulmonary disease. Lower threshold for cardiac evaluation in older adolescents.
Transcranial Doppler Ultrasound (TCD)	q 6-12 months from ages 3 to 16 years. Not indicated in patients with HbSC or HbS-β^+ thalassemia, or those on chronic transfusion programs
Dilated ophthalmology with retinal exam	Annually once 10 years old
Neuropsychometric testing	Consider for school or developmental concerns
Audiology	Only if clinical concern, including the prolonged use of ototoxic antibiotics or meningitis
Abdominal ultrasound	Not appropriate as screening. Only if clinical concern

VI. Immunizations

Pneumococcal Conjugate Vaccine (PCV7) (Prevnar™)	Per routine childhood schedule For patients of all ages	At least 2 doses, 6-8 weeks apart if over age 2 years
Pneumococcal Polysaccharide Vaccine (PPV23) (Pneumovax™, Pnu-immune™)	Starting at 24 months Given after PCV7 series	One booster in 3 years (after 5 years if over age 10)
Haemophilus influenza b (Hib)	Per routine childhood schedule	For ages over 5 and unimmunized, give 1-2 doses at least 1 month apart.
Meningococcal (Meningovax A&C™)	Starting at 24 months Not standard in all programs	Must give before splenectomy or dorm living
Influenza	Starting at age 6 months	Household contacts should also be immunized Live-virus currently not recommended

Selected references:

National Heart, Lung, and Blood Institute. *Management of Sickle Cell Disease*. (July 2002). http://www.nhlbi.nih.gov/health/prof/blood/sickle/index.htm.

AAP Section on Hematology/Oncology. Health supervision for children with sickle cell disease. *Pediatrics*. 2002: 109(3): 526 – 535. http://www.aap.org/policy/re1011.html.