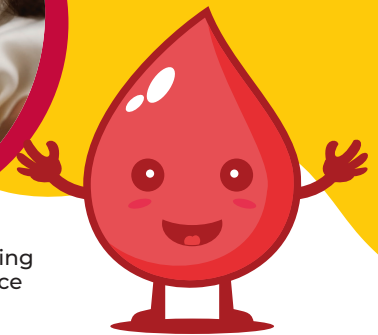


Health Maintenance Guidelines for Families with a Child with Sickle Cell Disease

Sickle Cell Disease, or SCD, is a blood condition that is inherited, or passed down, where the red blood cells look like a sickle, a "C-shape". This causes them to stick together. Red blood cells carry hemoglobin (Hb), a protein that attaches to oxygen. Children with SCD have problems with getting enough blood flow throughout their body. It is important for families (and in time the child) to know what kind of SCD they have. (See the back for details.)



It is very important that you keep your child safe and healthy by following your doctor's orders and the guidelines below. Keep this chart in a place that will remind you of what is due at each doctor visit.

Age	Primary Care Visits	Hematology Visits	Labs	Penicillin	Immunizations	Screenings
0 - 6 months 	Every 2 months	Baby's 1st visit should be before 2 months old Then every 3 months after that	Blood work every 3 months	Start penicillin twice a day	Start all vaccines (shots) recommended by your baby's doctor	None
7 months - 12 months 	Every 3 months	Every 3 months	Blood work every 3 months (every 6 months for Hb SC or Hb S-β+)	Continue penicillin twice a day	Continue to get all vaccines (shots) recommended by the doctor	Neurodevelopmental screening
12 months - 24 months 	Every 3 months	Every 3 - 6 months	Blood work every 3-6 months Urine test every year	Continue penicillin twice a day	Continue to get all vaccines (shots) recommended by the doctor PLUS: – Flu, yearly – PPSV @ 2 years old – MCV4 <i>if not already given</i> <i>(If your doctor doesn't have these you may need to get them somewhere else.)</i>	Oxygen level testing (with a pulse oximeter) Neurodevelopmental Screening Dental Exam every 6 months
25 months - 5 years 	Every 6 months	Every 6 months	Blood work every 6 months (more frequently as needed) Urine test every year	Continue penicillin twice a day At 3 years, the Penicillin dose should be increased to 250 mg by your doctor	Continue to get all vaccines (shots) recommended by the doctor PLUS: – Flu yearly – PPSV – MCV4 <i>if not already given</i>	Oxygen level testing (with a pulse oximeter) Neurodevelopmental Screening Dental Exam every 6 months PLUS: Transcranial Doppler (TCD) every year for those with certain types of Sickle Cell (Hb SS & Hb S-β0) Confirm with the hematologist if this screening is needed or not.
Over 5 years 	Every 6-12 months	At least every year	Blood work: Every 6 months for HbSS and HbS-β0 OR Every 12 months for Hb SC or HbS-β+ (more frequently as needed) Urine test every year	Consult with hematology	Continue to get all vaccines (shots) recommended by the doctor PLUS: – Flu yearly – PPSV booster 5 years after first dose – MCV4 booster 5 years after 2 dose series complete	Oxygen level testing (with a pulse oximeter) Neurodevelopmental Screening Dental Exam every 6 months PLUS: TCD at least yearly from 2 to 16 years of age to prevent stroke for HbSS and HbS-β0 Screening MRI/MRA for school age children Yearly dilated eye exam starting at age 10
Adolescence (11 - 21 years) 	Every 6-12 months Develop a transition plan	At least every year	Blood work: Every 6 months for HbSS and HbS-β0 OR Every 12 months for Hb SC or HbS-β+ (more frequently as needed) Urine test every year	Consult with hematology	Continue to get all vaccines (shots) recommended by the doctor PLUS: – Flu yearly – MCV4 booster every 5 years	Oxygen level testing (with a pulse oximeter) Neurodevelopmental Screening Dental Exam every 6 months PLUS: TCD at least yearly from 2 to 16 years of age to prevent stroke for HbSS and HbS-β0 Screening MRI/MRA if not previously done Yearly dilated eye exam

SICKLE CELL DISEASE: DEFINITIONS AND DETAILS

What is Sickle Cell Disease?

Sickle cell disease (SCD) is a group of inherited (passed down) red blood cell disorders. The type of SCD a person has depends on the genes they inherited from their parents. It is important for families and, in time, the child to learn what type of SCD they have. SCD is a lifelong illness, but it is very treatable.

What are the most common types of Sickle Cell Disease (SCD)?

The most common types of SCD include hemoglobin SS (HbSS), hemoglobin SC (HbSC), hemoglobin SBeta0 thalassemia (HbSB⁰), and hemoglobin SBeta+ thalassemia (HbSB⁺).

HbSS

People who have this type of SCD inherit two "S" genes, one from each parent. Hemoglobin S is an abnormal type of hemoglobin that causes the red cells to become rigid, and sickle shaped. This is commonly called sickle cell anemia and is usually the most severe type of the disease.

HbSC

People who have this form of SCD inherit a hemoglobin "S" gene from one parent and a hemoglobin "C" gene from the other parent. This abnormal type of hemoglobin usually causes a milder form of SCD.

HbS beta thalassemia

People who have this form of SCD inherit a hemoglobin "S" gene from one parent and beta thalassemia from the other parent. Beta thalassemia also causes another type of hemoglobin abnormality. There are two types of beta thalassemia: "zero" (HbS beta0 or HbSB⁰) and "plus" (HbS beta+ or HbSB⁺). Those with HbS beta0-thalassemia usually have a severe form of SCD. People with HbS beta+-thalassemia tend to have a milder form of SCD.

What is a hematologist?

A hematologist is a doctor for blood problems such as sickle cell disease (SCD).

Why does my child need penicillin every day?

Children with SCD are more likely to get infections. Penicillin every day helps to lower the chances of infection. Infections for children with SCD can cause serious problems. Penicillin helps protect children until they are fully immunized.

Why does my child need immunizations (vaccines) or shots?

Shots help prevent many serious infections. Children with SCD should get all shots including a flu shot on time.

What are PPSV and MCV4?

PPSV is Pneumococcal Polysaccharide Vaccine, a vaccine that helps protect against 23 types of pneumococcal infections (pneumonia) and MCV4 is Meningococcal Conjugate Vaccine, a vaccine that helps prevent 4 types of meningococcal infections (meningitis.) Both vaccines need to be given after the age of 2.

Why is my child on a different shot schedule than other children?

People with SCD are at "high risk" for certain infections and should follow a special shot schedule. This may not be the same as other children in your family.

What is neurodevelopmental screening?

This screening shows how your child is growing and learning over time. During this screening you will be asked questions about your child's milestones including activity, sounds/words and behaviors.

What is a pulse oximeter?

A pulse oximeter is a painless clip-like device usually placed on the finger to see if there is enough oxygen in the blood.

What is a dental exam?

A dental exam is when a doctor of teeth, also known as a dentist, looks at the teeth and mouth.

Why does my child need a dental exam every 6 months?

Children with SCD may have teeth that come in later, softer (more likely to decay), not straight, and may have gums and mouth bones that are not healthy.

Why does my child need to have a urine test every year?

A urine test can help find kidney problems which may lead to treating them early and lessen harm.

What is a Transcranial Doppler (TCD) study?

Transcranial Doppler (TCD) is a safe, painless, non-invasive (nothing enters the body) test that uses sound waves (similar to ultrasound during pregnancy) to see the blood flow in the brain. It helps find children at higher risk for stroke.

Why does my child need a Transcranial Doppler study?

Children with sickle cell disease are at risk for stroke. Transcranial Doppler (TCD) ultrasounds help predict that risk and allow treatment to prevent strokes.

What is an MRI/MRA?

MRI/MRA (Magnetic Resonance Imaging/Angiogram) is a way of taking pictures of the brain that helps your doctor know whether your child is having sickle cell-related problems like stroke that can affect their ability to think and learn. It is a safe, non-invasive procedure, but some people become scared due to having their head in a small space and the noise the machine makes. It usually cannot be done without sedation (medicine that can make someone calm, and sometimes sleepy.) MRI/MRA usually is not done until a child is 5-6 years of age.

Why does my child need a dilated eye exam every year after age 10?

Dilated eye exams help with early screening and treatment which can lower the chance of problems that cause loss of sight.

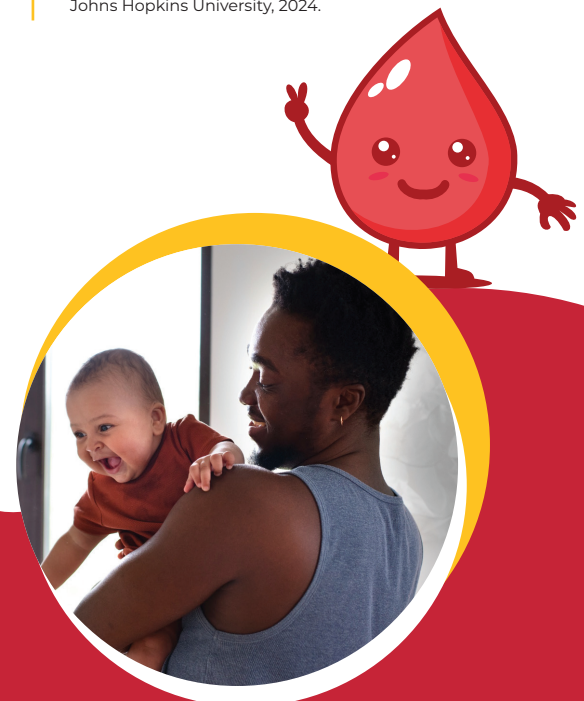
What is a transition plan?

A transition plan is a plan for your child to move from care for children to care for adults. Talking about transition usually starts around age 12-14 but is different for everyone.

Why does my child need a transition plan?

As children become adults, they need to learn to handle their own health and healthcare.

Reference: Sickle Cell Advisory Committee of the Genetic Network of New York, Puerto Rico and the Virgin Islands. **Guidelines for the Treatment of People with Sickle Cell Disease.** 2015; and personal communication from Dr. James Casella, Pediatrics and Oncology, Chief, Division of Pediatric Hematology, Johns Hopkins University, 2024.



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