

Common Tests for Patients with Sickle Cell Disease

Throughout your child's life, there will be certain tests and investigations your hematology team will do to monitor for damage from sickle cells in your child's body. These include:

Audiology Exam



What is it?

An audiology exam is a hearing test that checks for hearing loss.

Why does my child need it?

Sickle cells can damage small structures in your child's ears and cause hearing loss. Audiology exams help ensure hearing is normal and monitor hearing if there is damage from sickle cells.

When is it done?

Your child will have his/her first test by 5 years of age. It will be done every five years after that, or sooner if your child has signs of has hearing loss or delayed speech development.

Ophthalmology Exam



What is it?

An ophthalmology exam is an eye exam done by an ophthalmologist (a doctor that specializes in eye health.) This is different from an optometrist, who measure eyes for prescribing glasses or fitting contacts. An ophthalmology exam can find eye problems that an optometry exam cannot.

Why does my child need it?

Sickle cells can block the small blood vessels in the eyes and prevent the eyes from getting oxygen. More blood vessels grow to help increase oxygen flow, but these blood vessels are fragile and can cause bleeding, retinal detachment, and blindness.

When is it done?

Sickle cell eye problems usually show up in adulthood. However, your child will have testing between 5 and 10 years of age, and then every 5 years. This helps the hematology team to detect and treat problems early.

Transcranial Doppler (TCD)



What is it?

A TCD is an ultrasound of the large blood vessels leading to the brain. It uses sound waves to show the blood flow to the brain.

Why does my child need it?

Children with sickle cell have a higher risk of stroke than other children. Blood vessels leading to the brain can narrow, decreasing the amount of oxygen the brain tissue receives. A TCD is used to monitor this blood flow and helps us determine your child's risk for stroke.

When is it done?

Your child will have a TCD annually starting at 2 years of age. If a result is abnormal, testing may be done more frequently.

Magnetic Resonance Imaging (MRI)



What is it?

An MRI uses powerful magnets and radio waves to create computerized pictures of the brain.

Why does my child need it?

Sickle cells can block small blood vessels leading to the brain, decreasing the amount of oxygen his/her brain receives. This can cause brain cells to die (called an *infarct*). Large infarcts can have noticeable symptoms, but small infarcts might go unnoticed and increase your child's risk of having a stroke. An MRI can show if your child has had infarcts or a stroke.

When is it done?

MRIs will be done if your child shows signs of a stroke, or if there are signs of mental or physical delay.

Echocardiogram



What is it?

An echocardiogram uses high frequency sound waves to create a picture of the heart as it beats. An echocardiogram can show problems with both the anatomy and function of the heart.

Why does my child need it?

Your child's heart has to work harder to supply the body's tissues and organs with oxygen. This can make the heart grow bigger and can lead to problems like hypertension later in life. Also, if sickle cells block blood flow to the heart muscle they can cause some of the heart tissue to die (called a myocardial infarction, or MI). An echocardiogram shows doctors how healthy the heart is and how hard it is working.

When is it done?

Your child will have an echocardiogram at about 5 years of age. It will be repeated every 5 years or so to detect changes in heart function, and more frequently if needed.



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Chest X-Ray



What is it?

An x-ray is a uses electromagnetic energy to create pictures of bones and organs in the body.

Why does my child need it?

Your child is at risk for a serious complication of sickle cell disease called *acute chest syndrome*. Sickle cells can clump in the small blood vessels in the lungs, causing shortness of breath, chest pain, faster breathing, fever and coughing. Acute chest syndrome is an emergency. An x-ray can show if there is acute chest syndrome present. It can also be used to see if the heart is enlarged (as described above.)

When is it done?

Your child will have a baseline x-ray around 5 years old, and again every 2 or 3 years. If your child develops chest pain or breathing difficulties, x-rays will be done more frequently.

<u>Pulmonary Function Tests (PFTs)</u> What are they?



PFTs are tests that determine how much air the lungs take in and push out, and how well the lungs oxygenate the blood.

Why does my child need them?

Sickle cell disease can cause long term lung damage. Your child might not transfer oxygen from his/her lungs into the bloodstream very well. PFTs will help the hematology team monitor the health of your child's lungs. PFTs also monitor lung function after an episode of acute chest syndrome.

When are they done?

Your child will have PFTs done once he/she is about age 6. After that, they will be done every 4 to 5 years. They will be done more often if your child has had poor results or an episode of acute chest syndrome.

Abdominal Ultrasound



What is it?

An ultrasound is uses high frequency sound waves to create a two dimensional picture of the inside of the abdomen.

Organs in the abdomen (i.e. the gallbladder and spleen) can then be easily analyzed for size and problems.

Why does my child need it?

Your child's body makes and destroys red blood cells very quickly. When red blood cells are destroyed they release *bilirubin* into the blood. When bilirubin levels are high, painful crystals can form in the gallbladder called *gallstones*. Ultrasounds can show whether your child has gallstones.

Your child might also have an enlarged spleen. The spleen is an organ that cleans certain bacteria out of the bloodstream. Sickle cells can become trapped in the spleen as they flow through, causing the spleen to enlarge. Although this is common, it can lead to more serious complications. Ultrasounds can help monitor the size of the spleen over time.

When is it done?

Your child will likely have an abdominal ultrasound by about 5 years of age. It will be repeated every 2 to 3 years. Testing will be done more often if your child develops signs of gallstones (ex. abdominal pain, nausea, vomiting and fullness after small meals) or an enlarged spleen (pain, paleness, fatigue, and increased heart rate).

For more information contact:

Heather McCartney, BSN RN

Hemoglobinopathies Nurse Clinician, BC Children's Hospital

Office: (604) 875-2345 ext 7103

Pager: (604) 707-3895 Email: <u>hmccartney@cw.bc.ca</u>