



# Pediatric Clips

**NURSING**

## *Sickle cell disease*

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Pediatric Nursing Clips by Pediatric Advanced Practice Nurses at Dayton Children's provides quick reviews of common pediatric conditions.

The Children's Medical Center of Dayton is the region's pediatric referral center for a 20-county area. As the only facility in the region with a full-time commitment to pediatrics, Dayton Children's offers a wide range of services in general pediatrics as well as in 35 subspecialty areas for infants, children and teens. We welcome your inquiries about services available – call 937-641-3666 or e-mail marketing@childrensdayton.org.

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### CASE STUDY

Gretchen, a 3-year-old girl with sickle cell anemia (HbSS), presented to the emergency department with a seizure and right-sided weakness. Gretchen was admitted emergently to the intensive care unit (ICU) and had an exchange transfusion. A MRI/MRA was obtained

after she was stabilized showing an acute infarct in the left parietal hemisphere along with periventricular white matter changes bilaterally. The MRA showed narrowing of the cavernous portion of the left internal carotid artery with changes suggestive

of moyamoya. Moyamoya is a rare, progressive cerebrovascular disorder caused by blocked arteries at the base of the brain. Moyamoya means *puff of smoke* in Japanese and describes the look of the tangle of tiny vessels formed to compensate for the blockage.

### CASE DISCUSSION

Sickle cell disease (SCD) is identified by a newborn screen performed at birth for hemoglobinopathies and confirmed by a hemoglobin electrophoresis (HGEL). Hemoglobins (Hb) identified by neonatal screening are generally reported in order of quantity. Because more fetal hemoglobin (HbF) than adult hemoglobin (HbA) is present at birth, most normal infants show HbFA. Infants with hemoglobinopathies also show a predominance of HbF at birth. Sickle cell disease shows HbS in absence of HbA. Sickle cell anemia (HbSS) is caused by inheriting two HbS genes; one from each parent. HbSS is the most common and severe type of sickle cell disease, primarily seen in African heritage. The second most common type is HbSC followed by HbSBeta-thalassemia. In the US over 80,000 people have SCD, making it one of the most common genetic diseases in the country. Each year approximately 1,000 infants are born in the US with the disease.

#### DIAGNOSIS

Sickle cell disease is one of the few conditions associated with childhood stroke. This occurs in approximately 12% of children with certain types of sickle cell disease (HbSS and HbS $\beta^0$  thalassemia). Symptoms of brain ischemia/infarct include hemiparesis, difficulty with speech or vision, seizures - especially focal and sudden, severe headaches with or without alteration of consciousness. As soon as a stroke is suspected a thorough evaluation is recommended.

After initial stabilization and evaluation, patients should receive an urgent non-contrast CT scan of the brain to rule out hemorrhage or other nonischemic etiologies including CNS infection, trauma or intoxication. In early ischemia episodes (less than three hours) the cranial CT scan may be negative or show only subtle signs. MRI provides better details of the areas of ischemia and MRA may show large vessels occlusive disease. An evaluation within the first hours to days with an MRI/ MRA is recommended. 13% of children with SCD have "silent" brain lesions on their MRI, predominately the frontal lobe. These lesions are associated with poor performance on neuropsychological testing.

#### TREATMENT

The treatment for a sickle cell patient who had a stroke is placement on a chronic transfusion program. It has been estimated that as many as two-thirds of children with SCD who had a stroke will have additional strokes unless treated with chronic blood transfusions. The transfusion is given every three to four weeks. The goal of treatment is to maintain the HbS level or sickle hemoglobin less than 30%. Current recommendations are: transfusions should be continued for at least five years or until the child reaches the age of 18. With sickle cell anemia, the stroke is a constant threat after age 2, but the threat increases in the middle of a child's first 10 years of life.

After the age of 2, a Transcranial Doppler (TCD) is obtained yearly for all patients with HbSS and HbS $\beta^0$ . Any abnormalities on the TCD, velocity greater than 200cu/sec, is an indication for an MRI/ MRA. Depending on the results, a child may be placed on chronic transfusions prophylactically.

Studies have shown that a drug called Hydroxyurea has benefited children with SCD. Treatment with hydroxyurea in children has roughly the same effect on acute events as in adults and may prevent splenic dysfunction, cerebral-artery damage and secondary stroke. It raises hemoglobin F (fetal hemoglobin) by stimulating the production of protective fetal Hgb within the red cells. Having a higher percentage of circulating hemoglobin as hemoglobin F (15-20%) protects against sickling and its many complications. Some sickle cell centers are using chronic transfusions and hydroxyurea therapy to prevent strokes.

The main side effect of chronic transfusion therapy is iron overload and alloimmunization, and is treated with an iron chelator. Deferoxamine is one type of iron chelator given as a subcutaneous infusion usually eight hours, five days a week. Compliance is often poor, especially with the adolescent population. When deferisirox, another type of iron chelator that can be taken orally, became available most patients were placed on it. Sickle cell patients are usually placed on deferisirox when their ferritin reaches

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1,000 or greater (Normal ferritin level is 7-140).

## CONCLUSION

Gretchen has done well over the last several years. She remains on chronic transfusions, and her neurological deficit has resolved almost completely. Her annual MRI/MRA has been stable with no further progression of vascular disease. She will remain on chronic transfusions and hydroxyurea. She will continue on deferasirox until transfusions are stopped and her ferritin level drops below 1,000ng/ml, preferably 500 ng/ml.

Children with sickle cell disease have many challenges, stroke being just one of them. They deal with painful vasocclusive crisis

affecting the body in addition to other complications such as anemia, gallstones, delayed growth and development, proteinuria and other kidney problems, priapism, leg ulcers and increase risk for bacterial infection due to decrease or lack of splenic function. Being a chronic condition, patients have to adjust to being on medication all their life and be aware of their care. They must avoid extreme temperatures, drink plenty of fluids, eat a healthy diet and have regular checkups with their hematologist in addition to their pediatrician and dentist. As per standard care, patients are put on penicillin prophylaxis at 2 months of age and continue through their 5th birthday for children with HbSS and 2 years of age for all other sickle cell disease. In addition pa-

tients receive all the appropriate vaccinations including the pneumovax, meningococcal and yearly flu vaccine.

## REFERENCES

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## FEATURED NURSE SPECIALIST



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### THE WEST CENTRAL OHIO COMPREHENSIVE SICKLE CELL CENTER AT DAYTON CHILDREN'S

The West Central Ohio Comprehensive Sickle Cell Center is located in the hematology/oncology department at The

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