



Stroke in a child with sickle cell anemia

A 5-year-old's life hinges on the nurse's knowledge of stroke risk.

By Shannon Hudson, PhD, RN

ADRIENNE JOHNSON, age 5, is admitted to the general pediatric unit. She has sickle cell anemia (hemoglobin SS subtype) and recently was discharged after being hospitalized for acute chest syndrome. According to her mother, Adrienne has been complaining of severe persistent headaches for the past 24 hours. Her primary care provider admitted her for further evaluation.

History and assessment hints

When you enter her room, you find Adrienne awake and alert. She says she has a headache, which she rates as a 4 (“hurts a whole lot”) on the **FACES Pain Rating Scale**. You notice her speech is slightly slurred. Her mother confirms her speech is abnormal and tells you this is the first time she has noticed a change in her daughter's speech.

You quickly assess Adrienne's vital signs: blood pressure, 102/64 mm Hg; heart rate, 110 beats/minute; respiratory rate, 24 breaths/minute; and oxygen saturation, 98% on 2 L/minute oxygen by nasal cannula. Given her sickle cell anemia, headache, slurred speech, and recent history of acute chest syndrome, you suspect she's having a stroke. You immediately activate the pediatric rapid response team (RRT) and ask the unit's nurse manager to call Adrienne's primary care provider.

On the scene

When Jenna, the RRT nurse arrives, you report Adrienne's history and assessment findings. She conducts a rapid head-to-toe assessment; the neurologic exam reveals substantial hemiparesis. Jenna quickly calls the pediatric intensivist and hematologist and initiates Adrienne's transfer to the pediatric intensive care unit (PICU) for closer monitoring. As ordered, she obtains samples for a complete blood count with reticulocyte count and for blood typing and crossmatching. She also prepares Adrienne for a STAT computed tomography (CT) scan of the head without contrast. The CT scan confirms ischemic stroke.

Outcome

In the PICU, the pediatric intensivist establishes two-way venous access, initiates a partial-volume exchange

transfusion, orders maintenance I.V. fluids, and continues supplemental oxygen administration. After the exchange transfusion, magnetic resonance imaging confirms Adrienne has had an acute ischemic cerebral infarct, and it reveals multiple lesions from earlier clinically silent ischemic events.

Over the next 48 hours, Adrienne's neurologic status improves and she is transferred back to the general pediatric unit. Twenty-four hours later, she is discharged. You instruct her parents to follow up with their daughter's primary care provider and specialty providers.

Education and follow-up

Approximately 11% of children with sickle cell disease have a stroke by age 20. Typically, stroke in children with this disease presents with hemiparesis, altered speech or aphasia, visual deficits, headache, and seizures. These signs and symptoms can occur with or without an altered level of consciousness. Children with hemoglobin SS, significant anemia, or recent acute chest syndrome and those ages 2 to 10 are at highest risk of stroke. To prevent future strokes, chronic transfusion therapy is recommended. Without treatment, more than half of children with an initial stroke experience repeat events.

Throughout the child's hospitalization, support the family and assess family-level needs. At discharge, provide general instructions on caring for a child with sickle cell disease, including the importance of avoiding vaso-occlusive crises, managing pain, and administering prophylactic medications. Teach the family about signs and symptoms of stroke and instruct them to seek immediate care if the child experiences one or more. Urge them to keep the child's follow-up medical appointments. Inform the parents that having their child evaluated by a hematologist with transcranial Doppler ultrasonography monitoring and chronic transfusion therapy may prevent future strokes. ★

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