

A 12-year-old African American male with known sickle cell disease presents with a 2 hour history of right-sided arm weakness and slurred speech. He has been hospitalized before for pain crises and pneumonia. He takes hydroxyurea, oxycodone as needed, and folic acid. His temperature is 98.0°F (36.6°C), blood pressure is 153/83 mmHg and heart rate is 112/min with regular rhythm. On physical examination, he has right arm weakness and mild dysarthria. His laboratory values are the following:

Hemoglobin	8.2 mg/dL
WBC count	14,000/mm ³
Platelet count	210,000/mm ³
Creatinine	0.9 mg/dL

CT of the head shows no evidence of intracranial bleeding. Which of the following is the best initial management for this patient?

- ☐ A. Beta-blockers and aspirin
- ☐ B. Exchange transfusion
- ☐ C. Fibrinolytic therapy
- ☐ D. Heparin and warfarin
- ☐ E. Plasmapheresis

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- ☐ A. Beta-blockers and aspirin [7%]
- ☒ B. **Exchange transfusion** [46%]
- ☐ C. Fibrinolytic therapy [34%]
- ☐ D. Heparin and warfarin [6%]
- ☐ E. Plasmapheresis [7%]

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Explanation:

User Id: [REDACTED]

This patient's slurred speech may represent a Broca's aphasia. In combination with his right-sided arm weakness, these findings are suggestive of a focal abnormality in the left frontal lobe, most likely an acute stroke given his history of sickle cell disease. There are a variety of factors which can lead to stroke in a young sickle cell patient, most prominently sludging and occlusion of the cerebral arterial vasculature by the malformed red blood cells. While the head CT is reportedly negative, this should not dissuade one from considering stroke as it can be difficult to detect changes of stroke in the first few hours on a CT.

In the acute setting, one of the primary treatments for stroke in a sickle cell patient is

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In the acute setting, one of the primary treatments for stroke in a sickle cell patient is exchange transfusion. While this may not reverse changes from the initial vascular event, it helps to decrease the percentage of sickle cells in the bloodstream and makes an additional stroke less likely. Continuing the hydroxyurea should also help to decrease the percentage of sickle cells by increasing the proportion of Hgb F.

(Choice A) It is unlikely that this patient has true underlying vascular disease given his young age. Treatment should be directed towards the sickle cell disease itself.

(Choice C) Fibrinolytic therapy is unlikely to be effective since this patient's symptoms are likely secondary to sludging of sickle cells as opposed to a true thrombus.

(Choice D) Heparin and warfarin will likely not be effective as this patient probably does not have a true thrombus.

(Choice E) Plasmapheresis is not a common treatment used in sickle cell disease.

Educational objective:

Stroke is a common complication of sickle cell disease secondary to sludging and occlusion in the cerebral vasculature. Exchange transfusion is the recommended treatment acutely since it helps to decrease the percentage of sickle cells and prevent a second infarct from occurring.

References:

1. [Exchange blood transfusion compared with simple transfusion for first overt stroke is associated with a lower risk of subsequent stroke: a retrospective cohort study of 137 children with sickle cell anemia.](#)