Pediatric Neurology Part III: Chapter 199. Cerebrovascular complications in children with sickle cell disease (Handbook of Clinical Neurology)

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Cerebrovascular accidents were until recently responsible for much mortality and morbidity in children with sickle cell disease; the likelihood of a child with HbSS having a stroke was 11% before age 20 years, with a peak incidence of ischemic stroke between 2 and 5 years of age, and of hemorrhagic strokes between 20 and 29 years of age. Vessels occlusion is likely initiated by intimal proliferation and amplified by inflammation, excessive adhesion of cells to activated endothelium, hypercoagulable state, and vascular tone dysregulation. Silent infarcts may occur and are associated with decreased cognitive functions. Transcranial Doppler ultrasonography (TCD) was more recently demonstrated able to achieve early detection of the children at high risk for clinical strokes. A randomized study demonstrated that a first stroke may be prevented by monthly transfusion in children with abnormal TCD, leading to a recommendation for annual TCD screening of children aged between 2 and 16 years and monthly transfusion for those with abnormal results. In children who have had a first stroke, the risk of recurrence is more than 50% and is greatly reduced by chronic transfusion, although not completely abolished. Hematopoietic stem cell transplant is indicated in children with cerebral vasculopathy who have an HLA-identical sibling.

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