VASCULAR DISORDERS

SILENT INFARCTS WITH SICKLE CELL ANEMIA

The effect of transfusion therapy on the risk for new silent infarct or stroke in children with sickle cell anemia and abnormal transcranial Doppler (TCD) ultrasonography was determined at the University of Miami, FL, and other centers in the STOP trial (Stroke Prevention in Sickle Cell Anemia). One hundred thirty subjects with elevated TCD ultrasonographic velocity, including 47 (37%) with previous silent infarcts, were randomized to receive long-term transfusion therapy or standard care. MRIs were performed initially, annually, and when indicated by clinical symptoms. Among children with previous silent infarcts, standard care was associated with a significantly increased risk of new silent infarcts or stroke when compared to those receiving transfusion therapy. MRI lesions at baseline carried an increased risk of stroke or new silent lesions in standard care patients. Transfusion therapy lowers the risk for new silent infarct or stroke in children with both abnormal cerebral artery velocity and silent infarct. Subjects with both abnormalities who are not transfused are at increased risk of new silent infarct or stroke when compared to those with normal initial MRI. An abnormal initial MRI indicates the need for TCD screening and probable transfusion therapy. Elevated TCD velocity indicates the need for MRI to exclude silent infarcts. (Pegelow CH, Wang W, Granger S et al. Silent infarcts in children with sickle cell anemia and abnormal cerebral artery velocity. Arch Neurol Dec 2001;58:2017-2021). (Reprints: Charles H Pegelow MD, Department of Pediatrics (R-131), University of Miami School of Mediicne, PO Box 016960, Miami, FL 33101).

COMMENT. Children with sickle cell anemia who have elevated TCD ultrasonographic velocities account for only 10% of patients. However, most strokes in these patients are associated with an abnormal TCD, and the test is important particularly in those with MRI evidence of previous silent infarct. Patients with both abnormal TCD and MRI are candidates for transfusion therapy. Studies are recommended to determine the need for transfusion therapy in children with normal TCD and abnormal baseline MRI.

THALAMIC INFARCTION AND RUBRAL TREMOR

Two children, ages 14 months and 9 years, with a combined resting-postural-kinetic tremor following bilateral thalamic infarction are reported from Ataturk University, and Hacettepe University, Ankara, Turkey. The 14 month-old child had been treated with surgery and chemotherapy for Wilms' tumor. One week after completion of chemotherapy, he was admitted with high fever, and 3 days later, he developed a resting tremor in all limbs. This was complicated after 24 hours by 3-Hz clonic movements on the right side including the tongue. The movements tended to lessen in sleep. Two subsequent afebrile generalized seizures were associated with generalized spike and wave discharges on EEG, maximal amplitude over the left frontal area, and confirmed by video-EEG. Diazepam obliterated the EEG discharges, but the resting tremor persisted. The paroxysmal activity and tremor were thought to be independent. MRI of both patients showed bilateral, ischemic lesions in the medial thalami. Seizure and tremor responded to valproate and benztropine. The second patient had only resting tremor and the EEG was normal. (Tan H, Turanli G, Ay H, Saatci I. Rubral tremor after thalamic infarction in childhood. Pediatr Neurol Nov 2001;25:409-412). (Respond: Dr H Tan, Hacettepe Universitesi, Pediatrik Noroloji Bolumu, 06100 Sihhiye, Ankara, Turkey).