VASCULAR DISORDERS

SICKLE CELL DISEASE AND ASSOCIATED SEIZURES

The prevalence of cerebral vasculopathy in 76 patients with sickle cell disease (SCD), with and without seizures, and of magnetic resonance (MR) perfusion abnormality in those with recent seizures, was determined in a study at Great Ormond Street Hospital for Children, London, UK. Neurologic complications of SCD in 47 patients (23 male; median age 12 years; range 1.7-27 years) included recurrent seizures in 6, stroke in 9, transient ischemic attack (8), headaches (9), behavior and/or learning difficulties (9), or abnormal transcranial Doppler, more frequent in the seizure (4/6) and nonseizure (26/41; 63%) groups than in the asymptomatic (10/29; 34%) group. All seizure patients had relative decreased cerebral perfusion, and perfusion abnormalities were ipsilateral to electroencephalographic abnormalities. The development of seizures in patients with SCD is related to vasculopathy and focal hypoperfusion. (Prengler M, Pavlakis SG, Boyd S et al. Sickle cell disease: ischemia and seizures. Ann Neurol August 2005;58:290-302). (Respond: Dr Prengler, Neurosciences Unit, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, UK).

COMMENT. Seizures are reported in 12 to 14% of patients with sickle cell disease (SCD), and the prevalence in children with SCD is 10 times the general population (Liu JE et al, 1994; cited by authors). Vasculopathy and ischemia are factors in the occurrence of seizures in SCD. MR perfusion studies showing relative hypoperfusion in gray and white matter may be indicative of the cause of seizures when other neuroimaging methods are negative.

COAGULATION ABNORMALITIES ASSOCIATED WITH STROKE OR PORENCEPHALY

The prevalence of genetic and functional coagulation abnormalities in 59 children (age 0-18 years) with arterial ischemic stroke or porencephaly was compared with previously published population frequencies in a study at the National Institute for Neurological Disorders and Stroke, Bethesda, MD. Two thirds had at least 1 prothrombotic risk factor, and 5 had 3 or more. A family history of early thrombosis was found in one third of the children with coagulopathies and stroke. Abnormal factors included plasminogen activator inhibitor-1, methenyltetrahydrofolate reductase (involved in homocysteine metabolism), elevated lipoprotein (a), activated PC resistance, and factor V. Prothrombotic abnormalities are common in cerebrovascular disorders in children. (Lynch JK, Han CJ, Nee LE, Nelson KB. Prothrombotic factors in children with stroke or porencephaly. Pediatrics August 2005;116:447-453). (Reprints: Dr John Kylan Lynch, National Institute of Neurological Disorders and Stroke, Building 10, Rm 5S220, 10 Center Dr, MSC 1447, Bethesda, MD 20892).

COMMENT. Previous studies of coagulation abnormalities in children with stroke have shown variable results. The above findings support prothrombotic screening. Common risk factors for stroke have included cerebral arterial abnormalities, previous varicella zoster