VASCULAR DISORDERS

FACTOR V LEIDEN MUTATION AND STROKE

The association of the factor V Leiden mutation and cerebrovascular disorder (CVD) in children is reviewed and the clinical features of 8 children with cerebral palsy (CP) and the Leiden mutation are described from the National Institute of Neurological Disorders and Stroke, Bethesda, MD. Since 1995, 120 children with CVD or CP and factor V Leiden mutation have been reported in the literature. Cerebral infarction had occurred in 73 (typically left middle cerebral), cerebral venous thrombosis in 18, intraventricular or intracerebral hemorrhage in 14, CP in 9, and porencephaly in 6. Over half occurred in the perinatal period, and 69% in the first year of life. Seizures followed by acute hemiparesis were the most common neurologic symptoms. Additional exogenous risk factors for CVD in two thirds included birth complications, infection, cardiac disease, or malignancy, and 42% had additional acquired or genetic thrombotic risk factors. Maternal histories included pregnancy-related thrombosis, spontaneous abortion, toxemia, and placental abnormalities. At long-term follow-up, recurring seizures, hemiparesis, and developmental delay were common findings. In the 8 patients with CP followed by the authors, the translational product of the factor V Leiden mutation was elevated at 12-18 mcg/mL in all; protein C, protein S, and antithrombin III levels were also elevated in some; and the placenta was infarcted in 4 of 6 examined. (Lynch JK, Nelson KB, Curry CJ, Grether JK. Cerebrovascular disorders in children with the factor V Leiden mutation. J Child Neurol October 2001;16:735-744). (Respond: Dr John Kylan Lynch, Building 10, Room 5S220, 120 Center Drive, MSC 1447, Bethesda, MD 20892).

COMMENT. Although the risk of stroke in a child born to a mother with factor V Leiden factor is probably low, the incidence of factor V Leiden mutation in children with cerebral palsy or porencephaly is significant. Associated exogenous and endogenous risk factors are also important and deserve further study. Factor V Leiden mutation leads to thrombus formation in the heart, ductus arteriosus, deep veins, or placenta, which then result in emboli to the brain, infarction and stroke. An association with intraventricular hemorrhage and cerebral venous thrombosis is also demonstrated.

KAWASAKI DISEASE AND CNS INVOLVEMENT

A 4-year-old female hospitalized with encephalopathy developed classic signs of Kawasaki disease (KD) on the fifth day after admission, in a report from the Hopital Farhat-Hached, Sousse, Tunisia, and Saint-Luc University Hospital, Brussels, Belgium. Following a 2-day history of fever and progressive loss of consciousness, she had a left-sided hemiparesis, and a rash on the chest. EEG showed slowing, especially on the right side, and CT scan and CSF were normal. C-reactive protein was 4.8 gm/L (normal <0.6). Seizures recurred, coma persisted, the rash became generalized, palms and conjunctivae were injected, extremities swollen, and an echocardiogram revealed bilateral coronary artery aneurysms. KD was then diagnosed and she was treated with iv immunoglobulin and aspirin, with slow recovery of consciousness and motor function. At 3 month follow-up she could stand and walk, and at 12 months she displayed autistic behavior, communication dysfunction, and persistent seizures. MRI showed severe cerebral atrophy. (Tabarki B, Mahdhaoui A, Selmi H et al. Kawasaki disease with predominant central nervous system involvement. Pediatr Neurol Sept 2001;25:239-241) KD, an idiopathic acute vasculitis, may present with CNS signs.