VASCULAR DISORDERS

PRETHROMBOTIC DISORDERS IN THROMBOSIS AND STROKE

The frequency of inherited and acquired prethrombotic disorders in 30 consecutive children with arterial ischemic stroke (AIS) and 10 with sinovenous thrombosis (SVT) was determined at the Hopital de Pediatria, Buenos Aires, Argentina. The median age for AIS was 7.3 years (range, 0.4-18); for SVT it was 6 years (range, 0.8-12.6). Risk factors and underlying disease included infection, dehydration, head trauma, Moyamoya, and lupus. Hemostatic assays included protein C, protein S, antithrombin, plasminogen, activated protein C resistance, factor V Leiden mutation, and antiphospholipid antibodies. Nine children (30%) with AIS had one or more prethrombotic disorders (inherited protein S deficiency (2), inherited protein C deficiency (1), acquired antithrombin deficiency (2), antiphospholipid antibodies (3), and antiphospholipid antibodies and plasminogen deficiency (1). Five children (50%) with SVT tested positive: 1 had inherited protein S deficiency, 3 had acquired antithrombin deficiency, and 1 had antiphospholipid antibodies. No cases of activated protein C resistance or factor V Leiden mutation were found. (Bonduel M, Sciuccati G, Hepner M et al. Prethrombotic disorders in children with arterial ischemic stroke and sinovenous thrombosis. Arch Neurol 1999;56:967-971). (Reprints: Mariana Bonduel MD, Hopital de Pediatria "Prof Dr Juan P Garrahan," Combate de los Pozos 1881, Buenos Aires, Argentina 1245).

COMMENT. The majority of children with cerebral thrombosis and stroke have underlying disease or risk factors and hereditary or acquired prethrombotic disorders. A complete hemostatic evaluation is recommended for all children with arterial ischemic stroke and sinovenous thrombosis, including those with obvious clinical risk factors.

TRAUMATIC DISORDERS

MRI IN ACUTE NON-ACCIDENTAL HEAD INJURY

The value of magnetic resonance imaging (MRI) in the diagnosis of acute non-accidental head injury (NAHI) was studied retrospectively in 12 infants and children, ages 1 to 34 months (average, 5.7 months), admitted to the Royal Hospital for Sick Children, Edinburgh, Scotland. Injuries were whiplash-shaking with impact in 4 cases and without impact in 7, and a compression injury in 1. Subdural hematomas, commonly subtemporal, were present in all cases, and MRI findings were similar in cases with and without evidence of impact. MRI is superior to CT scan in defining subtemporal hematomas, tearing of bridging veins, and demonstrating hemorrhages at the grey-white matter junction, lesions common to cases of acute NAHI involving rotational acceleration/deceleration forces. (Barlow KM, Gibson RJ, McPhillips M, Minns RA. Magnetic resonance imaging in acute non-accidental head injury. Acta Paediatr July 1999;88:734-740). (Respond: Dr RA Minns, Department of Paediatric Neurosciences, Royal Hospital for Sick Children, 9 Sciences Road, Edinburgh, UK, EH9 1LF).

COMMENT. In infants with suspected non-accidental head injury, a CT scan on admission should be followed by MRI at 3 to 7 days after the acute injury, and as a guide to resolution of the hemorrhage. Whiplash-shaking injury syndrome without impact can result in MRI evidence of contusion and subdural hematomas, similar to that resulting from injury with impact.

Serial CT and MRI in outcome prediction of mild to moderate head injury. A series of 67 adults had CT on admission and MRIs within 1-3 and 6-12 months after injury. Outcome was worse in patients with edema and lesions on CT, and also in those with MRI lesions. Early MRI showing frontal lesions and late MRI with focal atrophy in frontotemporal regions, in combination with duration of amnesia, were predictive of outcome. (van der Naalt J, et al. Ann Neurol July 1999;46:70-78).

HEREDO-DEGENERATIVE DISEASES

IRON STORAGE IN FRIEDREICH'S ATAXIA

To test the hypothesis that iron is increased in the cerebellum of patients with Friedreich's ataxia (FA), a multigradient echo magnetic resonance sequence for the three-dimensional imaging of brain iron-induced contrast was used in 12 patients and 23 normal subjects examined at the National Institutes of Health, Bethesda, MD. Relaxation rate (R2) values, the inverse of T2, in the unaffected globus pallidus were equal in FA patients and controls, but R2 values in the dentate nucleus of FA patients were significantly higher. These R2 values reflect an increased iron concentration in the dentate, which supports the hypothesis of oxidative damage as the mechanism for FA. (Waldvogel D, van Gelderen P, Hallett M. Increased iron in the dentate nucleus of patients with Friedreich's ataxia. Ann Neurol July 1999;46:123-125). (Respond: Dr Mark Hallett, NINDS, NIH, Bldg 10, Rm 5N226, 10 Center Drive, MSC-1428, Bethesda, MD 20892).

COMMENT. Increased iron in the dentate nucleus of patients with Friedreich's ataxia, demonstrated by magnetic resonance, points to oxidative damage as the pathogenesis.

LAMOTRIGINE THERAPY IN NEURONAL LIPOFUSCINOSIS

Lamotrigine (LTG) long-term anticonvulsant therapy was evaluated in 29 patients, aged 6-28 years (mean, 14 years), with juvenile neuronal ceroid lipofuscinosis (JNCL), followed for 1-6 years (mean, 3 years) at the Hospital for Children and Adolescents, University of Helsinki, Finland. An initial dose of 0.1-0.5 mg/kg/day was increased every 2 weeks up to a maintenance dose of 1.25-15 mg/kg/day. After 1 year, seizures were decreased by more than 50% in 10, and seizures became less severe in 9 of 22. General patient well-being was improved in 18 of 28. LTG monotherapy was continued in 13 of 19 patients. (Aberg L, Kirveskari E, Santavuori P. Lamotrigine therapy in juvenile neuronal ceroid lipofuscinosis. Epilepsia June 1999;40:796-799). Reprints: Dr L Aberg, Hospital for Children and Adolescents, Pediatric Neurology, PL 280, 00029 HYKS, Finland).

COMMENT. The classical form of JNCL has an onset between 5 and 8 years with visual loss followed by progressive psychomotor retardation, epilepsy beginning at 8 to 13 years, extrapyramidal signs, and behavior disorders. Seizures are usually generalized tonic-clonic and partial, occasionally absence or atonic, and in the final stages of the disease, sometimes myoclonic. Lamotrigine may be an effective anticonvulsant in patients with JNCL.