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VASCULAR DISORDERS

TREATMENT OF ARTERIAL ISCHEMIC STROKE

Current guidelines and evidence in the treatment of childhood arterial ischemic stroke (AIS) are reviewed by researchers at The Children's Hospital, Denver, CO, and other centers. Except for sickle-cell related guidelines that focus on antithrombotic therapy, evidence for management of AIS is based on two adult guidelines, the American College of Chest Physicians (ACCP) in the US and the Royal College of Physicians (RCP) in the UK. Hypoxia, hyperglycemia, fever, hypertension, and increased intracranial pressure are factors associated with adult AIS that have not been studied extensively in childhood stroke. Acute medical management RCP guidelines recommend (with strong consensus) that early neurosurgical referral be considered in children with depressed level of consciousness. Thrombolytic therapy and the use of intraarterial tissue plasminogen activator are not recommended. The efficacy of aspirin or other antiplatelet therapies is extrapolated from adult guidelines only. The RCP recommends initial treatment of acute AIS with aspirin, 5mg/kg/day, except for sickle cell patients and those with evidence of hemorrhage on imaging. Reye's syndrome is a concern with aspirin used in secondary prevention of recurrent stroke, especially during infections such as varicella and influenza. Both the RCP and the ACCP guidelines suggest aspirin as first-line treatment for acute AIS. ACCP also suggests treatment with heparin for 5-7 days and until cardioembolic stroke is excluded. For secondary prevention of recurrent idiopathic stroke, aspirin (1-3mg/kg/day (RCP) or 2-5mg/kg/day (ACCP) is recommended after anticoagulation therapy is discontinued. Retrospective studies suggest that low molecular weight heparin appears to be safe in childhood AIS, without a high risk for intracranial hemorrhage. AIS in moyamoya, sickle cell disease, vasculitis and genetic/metabolic disorders require distinct therapeutic

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approaches. Moyamoya is treated with surgical revascularization. AIS with sickle cell disease requires urgent erythrocyte exchange transfusion to reduce hemoglobin S levels to less than 30% and increase hemoglobin to 10-12gm/dl. For primary prevention of AIS in sickle cell disease, patients older than 2 years are screened yearly with transcranial Doppler and placed on chronic transfusion regimen if found to have increased velocity of blood flow due to arterial narrowing. Multicenter cohort studies of acute, preventive and rehabilitation therapies are essential to establish evidence-based guidelines for treatment of AIS in childhood. (Bernard TJ, Goldenberg NA, Armstrong-Wells J, Amlie-Lefond C, Fullerton HJ. Treatment of childhood arterial ischemic stroke. **Ann Neurol** June 2008;63:679-696). (Respond: Dr Bernard, The Children's Hospital, Denver, CO 80045. E-mail: timothy.bernard@uchsc.edu).

COMMENT. Risk factors for arterial ischemic stroke in childhood are different from those in adults and include cerebral arteriopathies, vascular anomalies, congenital heart disease, infection (herpes zoster virus and varicella), head and neck trauma, sickle cell anemia, genetic/metabolic disease, and prothrombotic abnormalities. A mortality rate of 2-11% and persistent neurologic deficit in 68-73% are cited. Among the risk factors for AIS in childhood, infections and inflammatory disorders are relatively common, occurring in 27% to 34% in 2 large patient cohorts (Lee YY et al. **Brain Dev** 2008;30:14-19; Riikonen R et al. **Neuropediatrics** 1994;25:227-233). In a review of recent developments in childhood AIS, Amlie-Lefond C et al (**Lancet Neurol** 2008;7:425-435) stress the role of infection, the association of arteriopathy and varicella, and research in the use of antiviral, antibacterial, and anti-inflammatory therapy for AIS.

HYPOTHYROIDISM AND STURGE-WEBER SYNDROME

Two children out of 83 (2.4%) with Sturge-Weber syndrome, who attended a multidisciplinary center at Johns Hopkins Hospital, Baltimore, MD, were found to have central hypothyroidism. The prevalence of hypothyroidism was 500-10,000 times that in the general population. Both patients (ages 7 and 12 years) received carbamazepine or oxcarbazepine for seizures, anticonvulsants known to cause abnormal thyroid function. Six (7.2%) of the 83 reported maternal hypothyroidism. An admitted limitation of the study was the lack of routine thyroid function studies during the study period of 2000-2007, leading to a posible underestimate of thyroid dysfunction. The 2 patients tested had clinical symptoms or signs of hypothyroidism and a family history of hypothyroidism. Thyroid function tests revealed a low free thyroxine and normal TSH. Months after initiation of levothyroxine, free thyroxine level returned to normal, skin, hair loss, and energy levels improved, headaches and recurrence of stroke-like episodes resolved, but the frequency and severity of seizures were unchanged. Evaluation of possible growth-hormone deficiency is planned following linear growth deceleration. (Comi AM, Bellamkonda S, Ferene LM, Cohen BA, Germain-Lee EL. Central hypothyroidism and Sturge-Weber syndrome. **Pediatr Neurol** July 2008;39:58-62). (Respond: Dr Comi, Department of Neurology, Kennedy Krieger Institute, 707 North Broadway, Tower 100A, Baltimore, MD 21205. E-mail: comi@kennedykrieger.org).