There are **more questions than answers**, according to an editorial comment by Shapiro ED of Yale University, New Haven, CT, and Gerber MA, University of Connecticut, Farmington. They consider that the clinical significance of CSF abnormalities in Lyme disease-associated facial palsy has not been established, and routine spinal tap and *parenteral* antibiotic therapy, recommended by some authorities in cases with CSF pleocytosis, is unnecessary and not justified.

Facial palsy in Kawasaki syndrome is reported from Children's Hospital, Boston, MA. (Bushara K, Wison A, Rust RS. Pediatr Neurol Nov 1997;17:362-364). A 12-week-old-boy with coronary artery aneurysms and Kawasaki syndrome (KS) developed a facial palsy which resolved after treatment with IV immunoglobulin. KS is a vascular inflammatory disease of young children of unknown cause, presenting with unexplained fever, conjunctivitis, red lips, tongue and pharynx, skin rash, cervical adenopathy, erythematous hands and feet, and 25% have coronary aneurysms. Encephalopathy, seizures, stroke, ataxia, myositis, and facial palsy are rare neurological complications.

VASCULAR DISORDERS

SUBTYPES OF ISCHEMIC STROKE: CHILDREN CF YOUNG ADULTS

Children aged 1 to 18 years with acute ischemic stroke, seen at Indiana University, and young adults aged >18 to 45 years, identified from the Indiana University and Northwestern University Young Adults Stroke Registries, were classified in subtypes as atherothrombotic (AT), cardioembolic (CE), small-vessel (SV), or other determined or unknown causes. The percentages of these stroke subtypes in children cf young adults were as follows: AT 0/16, CE 15/14, SV 0/3, other 49/44, and unknown 36/23. Prothrombotic causes (sickle cell disease) were more common in children (25/14%), and dissections more common in young adults (3/15%). Causes of stroke in the 15 to 18 year group of children were more similar to the young adults. (Williams LS, Garg BP, Cohen M, Fleck JD, Biller J. Subtypes of ischemic stroke in children and young adults. Neurology Dec 1997;49:1541-1545). (Reprints: Dr Linda S Williams, Department of Neurology, IUMC, 541 Clinical Drive, CL 365, Indianapolis, IN 46202).

COMMENT. Children with cardioembolic stroke have cyanotic heart disease, and sickle cell disease and moyamoya are the most common causes of the "other determined" subtype in this age group. Young adults with CE stroke have right-to-left atrial shunts due to patent foramen ovale, and arterial dissection and antiphospholipid antibodies are the most common "other determined" causes. Age 15 years is the most appropriate dividing line for subtyping ischemic stroke in children and young adults.

Protein C and S deficiency are risk factors for stroke, according to a study at the Childrens Hospital, Los Angeles, CA. (Koh S, Chen LS. Pediatr Neurol Nov 1997;17:319-321). Among 37 children with ischemic stroke, protein C and S deficiencies were the only identified risk factors in 2 (5.4%) and 5 (14%) patients, respectively.

Transcranial doppler screening for long-term stroke risk in chidren with sickle cell disease was studied at the Medical College of Georgia, Augusta, GA. Elevated TCD velocities, 200 cm/sec or greater, predict increased stroke risk. (Adams RJ, McKie VC, Carl EM et al. Ann Neurol Nov 1997;42:699-704).

Language delay in children with sickle cell disease and stroke is

reported in 10 children studied at the University of Arkansas, Little Rock. (Davis P, Landers A, Gentry B et al. Perceptual and Motor Skills Dec 1997;85:809-810).

MOYAMOYA SYNDROME AND CONGENITAL HEART DISEASE

The association of moyamoya syndrome and congenital heart disease is described in 5 patients at Children's Hospital, Boston, MA. Stroke was the presenting sign in 3 and seizures in 2. Coarctation of the aorta with septal or valvular defects was diagnosed in 3, and tetralogy of Fallot in 2. Moyamoya, diagnosed after surgery for congenital heart disease, was treated by cerebral revascularization. (Lutterman J, Scott M, Nass R, Geva T. Moyamoya syndrome associated with congenital heart disease. Pediatrics Jan 1998;101:57-60). (Respond: Tal Geva MD, Department of Cardiology, Children's Hospital, 300 Longwood Ave, Boston, MA 02115).

COMMENT. Moyamoya syndrome should be considered in the differential diagnosis of stroke or seizures associated with congenital heart disease, both before or after surgery.

METABOLIC DISORDERS

BIOTINIDASE DEFICIENCY: EARLY PRESENTATION

Two infants with manifestations of biotinidase deficiency presenting at age 3 weeks and 2 weeks are reported from the University of Aarhus, Roskilde County Hospital, and Herning Central Hospital, Denmark. Patient 1, born to related Kurdic parents, developed a generalized skin rash at 3 weeks, generalized tonic-clonic seizures up to 20 times daily at 6 weeks, and visual inattention, hypertonia and hyperreflexia on admission at 8 weeks. EEG showed epileptiform activity. Valproic acid was ineffective. Metabolic screening showed urinary B-hydroxyisovalerate and B-methylcrotonylglycine, and very low serum biotinidase activity. After oral biotin (5mg x 3 daily) the seizures stopped within a few days, and at 2 year follow up psychomotor development was normal except for hearing loss. MRI showing cerebral atrophy initially was normal at 12 months. Patient 2, the second child of related Kurdic parents, presented at 1 hour after birth with respiratory distress and septicemia. She had dry and squamous skin at 2 weeks, loss of hair at 4 weeks, and on readmission at 6 weeks she was lethargic, hypotonic, and hypothermic. Visual inattention, trembling, tense fontanelle, tonic clonic seizures, conjunctivitis, and alopecia were noted. EEG showed diffuse slowing and a right occipital spike focus. CT was suggestive of periventricular leukodystrophy. Serum lactate and pyruvate were elevated. Organic aciduria and absent serum biotinidase confirmed the diagnosis of biotinidase deficiency. Oral biotin (10mg daily) was begun at 7 weeks, and seizures were controlled and other manifestations improved within 2 weeks. At 18 month follow-up, development and CT were normal. The authors advocate routine neonatal screening for biotinidase deficiency in Denmark. (Haagerup A, Andersen JB, Blichfeldt S, Christensen MF. Biotinidase deficiency: two cases of very early presentation. Dev Med Child Neurol Dec 1997;39:832-835). (Respond: Dr Annette Haagerup, Institute of Human Genetics, University of Aaarhus, DK-8000 Aarhus C, Denmark).

COMMENT. Biotinidase deficiency is an autosomal recessive disorder causing multiple carboxylase deficiency and usually manifested at 3 to 6 months of age with intractable seizures, hypotonia, skin rash, alopecia, and developmental delay. Lactic acidosis leads to coma and death in untreated cases.