respiratory illness and those associated with minor head trauma are the most common forms seen in the ER. A serious neurological disorder requiring admission to hospital is encountered in 9% of headache patients. CT scans that are indicated when headache is linked to neurologic disorders may be positive in 15% of cases; none was positive among patients with migraine or nonneurological headaches. Prescription analgesic therapy for ED cases of headache is rarely indicated.

The prevalence of headache, including migraine, increased significantly in school age children, from 23% to 71% during an eighteen year time period (Sillanpaa M, Anttila P. Headache 1996;36:466-470). The highest increases occurred in headache related to tension, stress and social instability, while the prevalence of febrile illness and head trauma, common precipitating causes in ED headache, was unchanged.

The decision to obtain a CT scan in a child presenting in the ED with headache is sometimes difficult, and many cases, approximately 50%, require careful neurological evaluation. Absolute indications include the following: head trauma with skull fracture and/or impaired consciousness; symptoms and signs of increased intracranial pressure; ventriculo-peritoneal shunt malfunction; and abnormal increase in head circumference. Probable indications for CT include: atypical recurrent headaches; a recent change in the character of headaches; early morning headaches, especially if associated with vomiting; abnormal neurological findings; and occurrence of headache in younger age groups. Patients with a known diagnosis of migraine and those with viral and respiratory illness can be treated symptomatically and followed, but those with suspected neurological disorders deserve complete work up before discharge. (see Maytal J et al. Pediatrics 1995;96:413-416; and Progress in Pediatric Neurology III PNB Publ, 1997; pp185-190, for reviews of indications for imaging studies and prevalence of headache).

MUSCLE DISORDERS

FREQUENCY OF CHILDHOOD MITOCHONDRIAL MYOPATHIES

The frequency of mitochondrial diseases among patients with childhood encephalopathies and myopathies in a defined population of Northern Finland was investigated at the University of Oulu, Finland. Among a total of 116 consecutive patients with unexplained psychomotor retardation enrolled during a 7-year period, the frequency of ultrastructural mitochondrial abnormalities was 71% (ragged-red fibres in 4 cases), oxidative phosphorylation defect (OXPHOS) occurred in 28% (complex I and IV most commonly), and mutations in mitochondrial DNA (mtDNA) in only 1 patient (.9%). A diagnosis of mitochondrial disease was possible (with ultrastructural changes in muscle mitochondria) in 71%, probable (with defects in OXPHOS enzymes in addition to ultrastructural changes) in 15%, and definite (having pathogenic mutations in mtDNA) in 0.9%. Clinical manifestations were mental retardation alone or with hypotonia, ataxia, epilepsy, or spasticity in 74%, and muscular hypotonia or ataxia in 14%. EMG and NCS showed myopathy in 13%, nerve degeneration in 7%, and anterior horn cell disease in 2%. MRI or CT showed cortical atrophy in 18% and calcifications in 6%. Blood lactate was elevated in 58%. (Uusimaa J, Remes AM, Rantala H et al. Childhood encephalopathies and myopathies: a prospective study in a defined population to assess the frequency of mitochondrial disorders. Pediatrics March 2000;105:598-603). (Reprints: Kari Majamaa MD, University of Oulu, Department of Neurology, Kajaanintie 52 A, FIN-90220 Oulu, Finland).

COMMENT. Biochemical and ultrastructural abnormalities indicative of mitochondrial disease are sufficiently frequent to recommend muscle biopsy as an important diagnostic examination in children with unexplained mental retardation. Both ultrastructural mitochondrial abnormalities and decreased activity of 1 or more respiratory chain enzymes are required for a probable diagnosis of mitochondrial disease. The commonly known mtDNA mutations are a rare cause of childhood encephalomyopathies, in contrast to the adult form that frequently shows the MELAS mutation.

Mitochondrial DNA (mtDNA) defects in neuromuscular disorders are reviewed by Marin-Garcia J. and Goldenthal MJ (Pediatr Neurol February 2000;22:122-129) at the Molecular Cardiology Institute, Highland Park, NJ. Mitochondrial mtDNA deletions were found in 1 child with Kearns-Sayre disease, 1 with stroke/CADASIL, and 1 with progressive external ophthalmoplegia, hypotonia, developmental delay, and lactic acidosis. Reduced mtDNA levels occurred in 2 children with encephalomyopathy, hypotonia, lactic acidosis, and mtDNA depletion. Pathogenic mtDNA point mutations are maternally inherited, and most are located in tRNA and rRNA genes.

CLINICAL APPROACH TO METABOLIC MYOPATHIES

The clinical and laboratory evaluation of the patient with suspected metabolic myopathy is reviewed from the Department of Neurology, Children's Hospital, Boston, MA. Myopathies are classified as *static* characterized by proximal weakness, generalized weakness, and developmental delay; and *dynamic* with recurrent episodes of reversible muscle dysfunction, sometimes myoglobinuria, related to exercise intolerance, fasting, exposure to cold, anesthesia, intercurrent infection, or a low-carbohydrate, high-fat diet. Both forms are common in mitochondrial myopathies. The type or duration of exercise inducing weakness may be specific: Prolonged, low-intensity activity (eg walking) - induced weakness occurs with fatty acid oxidation (FAO) defects; high-intensity exercise (eg weight lifting or sprinting) - glycogen or glucose metabolism defects. Myoglobinuria may be induced by inborn errors of glycogen/glucose metabolism, FA metabolism, and some mitochondrial cytopathies. Laboratory tests include CK, elevated in glycogen defects and lactate dehydrogenase deficiency; blood lactate and pyruvate elevated in mitochondrial myopathies; liver transaminases elevated in FAO defects; and abnormal carnitine, acylcarnitine, free fatty acids, and hypoketotic hypoglycemia in lipid metabolic disorders. EMG, Forearm Ischemic Exercise Test, and muscle biopsy with molecular studies may be required in diagnosis. (Darras BT, Friedman NR. Metabolic myopathies: a clinical approach; Part I. Pediatr Neurol February 2000;22:87-97). (Respond: Dr Darras, Neuromuscular Program, Neurology Department, Fegan 11, Children's Hospital, 300 Longwood Avenue, Boston, MA 02115).

COMMENT. A helpful algorithm for the step-by-step diagnosis of metabolic myopathies is provided by the authors.

SEIZURE DISORDERS

GELASTIC EPILEPSY AND HYPOTHALAMIC HAMARTOMA

Three patients with small hypothalamic hamartomas and a recurrent "pressure to laugh," often without actual laughter, are reported from the University of Melbourne, Australia, and McGill University, Canada. Giggling