

COMMENT. Despite the potential adverse effects of corticosteroids in a patient already immunocompromised, a trial of betamethasone was considered appropriate in view of the poor neurologic prognosis in ataxia-telangiectasia. Limited trials of gabapentin, pregabalin and tiagabine were moderately effective in the control of ataxia in an adult with ataxia-telangiectasia (Gazulla J et al. New therapies for ataxia-telangiectasia. *Arch Neurol* April 2007;64:607-609).

HEADACHE DISORDERS

FAMILIAL HEMIPLEGIC MIGRAINE WITH *ATPIA2* MUTATIONS

Three children with prolonged hemiplegia following severe unilateral headache and having mutations in *ATPIA2* are reported from UCLA School of Medicine, Los Angeles, CA; University Children's Hospital, Zurich, Switzerland; and Wake Forest University School of Medicine, Winston-Salem, NC. Patient 1, a 9-year-old girl developed left-sided numbness and paralysis after 2 days severe right-sided headache. The hemiparesis lasted 5 days and then gradually resolved. Brain MRI and angiography and lumbar puncture were normal. EEG showed slowing in right hemisphere. No previous attacks were reported in the child, but her mother had a history of episodic hemiparesis lasting 30 min to an hour, and accompanied by headache and vomiting, since age 32 years. Patient 2, a 7-year-old boy had headache, somnolence, confusion and left hemiplegia, associated with viral gastroenteritis. Lumbar puncture and initial MRI were normal. Repeat MRI 2 days later showed increased signal intensity on diffusion weighted imaging in right parietal region. Hemiplegia persisted 7 days and gradually resolved over weeks. A generalized tonic-clonic seizure occurred 6 days after onset of hemiplegia. The child had a prior history of typical hemiplegic migraine, lasting 30 min and followed by unilateral headache, often induced by minor head trauma. Family history was negative. Patient 3, a 10-year-old girl developed a headache a few hours after striking her head in a fall while skating. She did not lose consciousness. Two-3 hours later, she developed right-sided numbness, hemiplegia and global aphasia. Symptoms persisted 10 days, the weakness resolved in 3 weeks, and language returned to normal after 7 weeks. Brain MRI on days 2 and 10 were normal. EEG showed slowing on the left that persisted several weeks. Her father had a history of typical hemiplegic migraine beginning at age 7 years; also, episodic visual aura without headache, and a single seizure.

Three distinct *ATPIA2* mutations were identified in the *FHM2* gene, but none in the *FHM1* gene of the patients. Affected parents of patients 1 and 3 also had the mutation. In Patient 2, neither parent had the mutation, indicating that the mutation in the child arose de novo. (Jen JC, Klein A, Boltshauser E et al. Prolonged hemiplegic episodes in children due to mutations in *ATPIA2*. *J Neurol Neurosurg Psychiatry* May 2007;78:523-526). (Respond: Dr Joanna C Jen, UCLA Neurology, 710 Westwood Plaza, Los Angeles, CA 90095).

COMMENT. Episodes of hemiplegic migraine usually last from 30 to 60 min, but patients with more prolonged hemiparesis are reported. *FHM1* is typically associated with cerebellar findings, and *FHM2* with seizures. More than 20 mutations in *ATPIA2* have been found to cause *FHM2*. The MRI is generally normal, but the EEG may show slowing over the affected hemisphere.

Altered arterial function in migraine of recent onset was demonstrated using ultrasound and applanation tonometry in 50 patients and 50 controls, at Ghent University, Belgium. Brachial artery diameter was decreased, while carotid arterial wall properties were unchanged during a headache-free interval. Flow-mediated vasodilation of the brachial artery was decreased in patients with migraine for >1 yr and <6 yrs duration, indicative of endothelial vasomotor dysfunction. (Vanmolkot FH, Van Bortel LM, de Hoon JN. *Neurology* April 2007;68:1563-1570). (Tietjen GE. Editorial. Migraine as a systemic disorder. *Neurology* April 2007;68:1555-1556).

MANAGEMENT OF ACUTE MIGRAINE IN CHILDREN

The management of acute pediatric migraine headaches in the pediatric emergency department (ED) and in a mixed pediatric and adult ED was compared at the University of Alberta, Edmonton, Canada. Of 382 patients identified with headache, aged 5 to 17 years, in 4 regional EDs during the 2003/2004 fiscal year, 186 (48.7%) met criteria for migraine. Simple oral analgesics in 20.7% and dopamine antagonists (metoclopramide and prochlorperazine) in 20.7% were prescribed first-line most commonly. Opiate medications (in 5.5%), ketorolac (4.7%), and dihydroergotamine (1%) were prescribed first-line infrequently. No treatment was given to 44.2%. The pediatric ED was more likely to prescribe a dopamine antagonist (12.9% vs 6.8%; $P=0.044$) while the mixed adult/pediatric EDs were significantly more likely to prescribe an opiate (28.1% vs 18.4%; $P=0.031$). Children with migraine in all EDs were significantly more likely to receive drug therapy (68.3% vs 42.9%; $P<0.001$) or a dopamine antagonist (32.3% vs 9.7%; $P<0.001$). Polypharmacy was used in 31.2%, and neuroimaging was obtained in 29.1%. In 109 studies, arachnoid cysts were diagnosed in 2 patients, optic glioma (1), sinus thrombosis (1), and sinusitis (1), a total of 5 (4.6%) abnormal scans. Lumbar puncture was performed in 9 patients (2.4%), with no difference between pediatric and adult cohorts. A headache associated infectious illness was diagnosed in 12%. Documentation of headache outcome was poor, but better in pediatric EDs compared with adult EDs (22.1% vs 13.6%). Complete headache resolution occurred in 13.9% and was significantly more common in the pediatric ED cohort compared with the adult ED cohort (20% vs 4%). (Richer L, Graham L, Klassen T, Rowe B. Emergency department management of acute migraine in children in Canada: a practice variation study. *Headache* May 2007;47:703-710). (Respond: Dr Lawrence Richer, University of Alberta-Pediatrics/Division of Neurology, 2C3, WMC 8440-112 St, Edmonton, Alberta, T6G 2R7, Canada).

COMMENT. Most children attending an ED in Canada with headache do not receive drug therapy. The type of medication most commonly prescribed is different in pediatric EDs compared to mixed adult/pediatric EDs. Dopamine antagonists are preferred in pediatric EDs and opiates in mixed EDs.