

cardiac rhabdomyoma: Implications for evaluation and follow-up. Ann Neurol Oct 1993;34:617-619). (Respond: Dr Weig, Department of Pediatrics, University of Massachusetts Medical Center, Worcester, MA 01655).

COMMENT. The authors cite one previous report of heart block secondary to erythromycin-induced CBZ toxicity in a 10-year-old boy. This child had no evidence of underlying cardiac disease. Additional cases in adults have been reported. Although CBZ cardiotoxicity is rare, an echocardiogram is recommended in patients with tuberous sclerosis treated with carbamazepine, and cardiac rhythm should be carefully monitored.

ICTAL SPECT IN LOCALIZING EPILEPTOGENESIS

Ictal single-photon emission computed tomography (SPECT) was used successfully in the localization of an epileptic focus in two children, ages 9 and 7 years, with intractable epilepsy reported from the University of Alabama at Birmingham Epilepsy Center. CT and initial MRIs were normal, and repeated EEG-video monitoring studies failed to localize the seizures. Resection of the abnormal areas shown by SPECT as hyperperfusion in the frontal lobes revealed a cortical dysplasia. The children had no neurological deficits and remained seizure free 1 year after surgery. (Kuzniecky R et al. Ictal single-photon emission computed tomography demonstrates localized epileptogenesis in cortical dysplasia. Ann Neurol Oct 1993;34:627-631). (Respond: Dr Kuzniecky, Department of Neurology, UAB Station, Birmingham, AL 35294).

COMMENT. Ictal SPECT performed exactly at the onset of a clinical seizure precisely localized the primary epileptogenic area in the cortical dysplasia and permitted a discrete resection of the focus without neurologic deficit.

TEMPORAL LOBE TUMOR AND LANDAU-KLEFFNER SYNDROME

A 7-year-old boy with a left temporal lobe tumor and Landau-Kleffner syndrome (acquired aphasia and epilepsy) who improved dramatically after surgery is reported from the New York Hospital-Cornell University Medical Center. A verbal auditory agnosia with mutism had evolved over a 3 month period after onset of refractory partial complex seizures at 5 years. EEG showed left temporal and right occipital spike and S/W discharges. Language comprehension was absent on the Peabody Picture Vocabulary test and Token test. Following removal of a grade 1 astrocystoma demonstrated on MRI, seizures stopped, language returned, and after 1 year speech and comprehension were almost normal for age. (Nass R et al. Landau-Kleffner syndrome: Temporal lobe tumor resection results in good outcome. Pediatr Neurol July/Aug 1993;9:303-5). (Respond: Dr Nass, New York Univ Med Ctr, 400 E 34th St RR 212, New York, NY 10021).

COMMENT. The favorable result of surgery in this patient contrasts with the poor outcome of most patients with Landau-Kleffner syndrome treated medically. MRI and ictal SPECT should be considered in all patients with this syndrome to rule out localized structural pathology amenable to surgery.

MOVEMENT AND BEHAVIOR DISORDERS

EARLY SIGNS OF TOURETTE'S SYNDROME

The early course of tics and behavioral disturbances in 101 children with Tourette's syndrome, aged 12 years (range, 8 to 21 years), are reported from the University of Rochester School of Medicine, Rochester, NY. The age at onset was 6.4 ± 3.1 years. At initial evaluation, 45% already had ADHD and 50% had obsessive-compulsive disorder (OCD). During the observation period of 1.6 years (range, 0.5 to 7 years), tics became controlled with medication in 81%, 13% developed ADHD, 8% OCD, 28% disruptive behavior, and 25% school problems. Of those with behavioral disorders initially, about 50% were controlled over time with treatment. Tic suppressants were withdrawn from 12% of patients. (Park S, Kurlan R et al. The early course of the Tourette's syndrome clinical spectrum. Neurology Sept 1993;43:1712-1715). (Reprints: Dr Kurlan, Department of Neurology, University of Rochester School of Medicine, 601 Elmwood Avenue, Rochester, NY 14642).

COMMENT. If ADHD and OCD are absent at the onset of tics, it is unusual for these behaviors to develop later. Disruptive behavior and school problems are more likely to develop over time.

DESMOPRESSIN ACETATE AND NOCTURNAL ENURESIS

A literature review of 18 randomized controlled trials of desmopressin for resistant enuresis involving 689 patients is reported from the University of Manitoba, Canada. Frequency of wetting decreased in all studies, ranging from 10% to 91%, and 25% of subjects became completely dry. In the long-term, only 5.7% of responders remained dry after medication was withdrawn. Children older than 9 years do better than those treated at an earlier age. A dose-response effect was apparent, and side-effects consisting of nasal stuffiness, headache, epistaxis, and abdominal pain were infrequent. No cases of water intoxication were reported. In one comparative study, patients treated by conditioning alarms had 10% fewer wet nights and a better long-term result. (Moffatt MEK et al. Desmopressin acetate and nocturnal enuresis: How much do we know? Pediatrics Sept 1993;92:420-425). (Reprints: Dr Moffatt, Univ Manitoba, Room S100-750 Bannatyne Avenue, Winnipeg, Manitoba R3E 0W3, Canada).

COMMENT. If we need to treat, alarms seem superior to drugs.