

SEIZURE DISORDERS

SURGICAL MANAGEMENT OF SEIZURES WITH FOCAL CORTICAL DYSPLASIA

Anatomical-clinical correlations and EEG findings in 10 children, aged 26 months to 11 years (median 6 years), with drug resistant partial epilepsy and focal cortical dysplasia, operated on 1996-2000, were analysed at the Epilepsy Surgery Centre "C Munari" in Milan, Italy. MRI abnormalities were unilobar in 5, bilobar in 2, and multilobar in 1. Two with negative MRIs had frontal lobe seizures. Surgery consisted of corticectomy plus lesionectomy in all cases. Seizures were completely controlled at 25 month follow-up or longer in 70% patients. Outcome was poor in multilobar patients. Development improved after surgery in all patients. Stereo EEG exploration in the most complex cases helped to define the epileptogenic zone in frontal, extratemporal and multilobar epilepsies. Stereo EEG facilitated resection of extralesional cortex. (Francione S, Vigliano P, Tassi L, et al. Surgery for drug resistant partial epilepsy in children with focal cortical dysplasia: anatomical-clinical correlations and neurophysiological data in 10 patients. J Neurol Neurosurg Psychiatry Nov 2003;74:1493-1501). (Respond: Dr Stefano Francione, Centro per la Chirurgia dell'epilessia "Claudio Munari," Ospedale Niguarda Ca' Granda, Pz Osperdale Maggiore 3, 20162 Milan, Italy).

COMMENT. Seizures are controlled following surgery in 70% of children with drug-resistant partial epilepsies and histologically confirmed focal cortical dysplasia. Stereo invasive EEG recording delineates the epileptogenic zone in patients with extratemporal epilepsies, and especially those with negative MRI.

CONGENITAL MALFORMATIONS

CUTIS MARMORATA TELANGIECTATICA AND CHIARI TYPE I

A 10-year-old girl born with telangiectasias of the lip, trunk, extremities, palms, and soles was found to have an asymptomatic Chiari I malformation without syringomyelia on MRI, and is reported from Children's Hospital, Alabama. Additional diagnoses in this patient included left hemihypertrophy, thoracic scoliosis, obsessive compulsive disorder, Tourette's syndrome, and an absence seizure disorder. The Chiari I malformation showed 22 mm of tonsillar ectopia. (Tubbs RS, Smyth MD, Wellons JC, III, Blount JP, Oakes WJ. Cutaneous manifestations and the Chiari I malformation. Pediatr Neurol September 2003;29:250-252). (Respond: R Shane Tubbs PhD, Pediatric Neurosurgery, Children's Hospital, Birmingham, AL).

COMMENT. Cutaneous manifestations associated with Chiari I malformation (CIM) are rare. The authors cite other reports including macrocephaly Cutis Marmorata Telangiectatica Congenita (M-CMTC), Leopard syndrome with CIM, blue rubber bleb nevus syndrome with CIM, acanthosis nigricans, neurofibromatosis type 1, and Waardenburg syndrome with congenital leukoderma.