CORTICOSPINAL DYSGENESIS AND CONGENITAL HEMIPLEGIA

A diffusion tensor imaging technique was compared with conventional MRI to measure and quantify corticospinal dysgenesis in 12 patients with congenital hemiplegia and 12 matched control subjects, in a study at Universite Catholique de Louvain, Brussels, Belgium. A symmetry index computed between the area of the contralateral and ipsilateral corticospinal tracts was similar for the two methods, but the diffusion tensor imaging indexes were significantly smaller. This suggests that the use of the conventional MRI measurement of the cross-sectional area of cerebral peduncles on T1 MRI might lead to an underestimate of cortical dysgenesis. Hand-movement deficits, particularly precision grasping, and stereognosis were examined and correlated with the neuroimaging findings. The symmetry index computed from MRI peduncle measurements correlated solely with deficits in stereognosis, while the diffusion tensor imaging index correlated with stereognosis, digital and manual dexterities, and a measure of manual ability in daily life activities. (Bleyenheuft Y, Grandin CB, Cosnard G, Olivier E, Thonnard J-L. Corticospinal dysgenesis and upper-limb deficits in congenital hemiplegia: a diffusion tensor imaging study. **Pediatrics** Dec 2007;120(6):e1502-e1511). (Respond: Jean-Louis Thonnard PhD, Universite Catholique de Louvain, Unite de Readaptation, Ave Mounier 53, 1200 Bruxelles, Belgium).

COMMENT. Diffusion tensor imaging symmetry index may prove useful in predicting motor and sensory deficits in children with congenital cerebral dysgenesis.

SEIZURE DISORDERS

SURGERY FOR INTRACTABLE TEMPORAL LOBE EPILEPSY IN YOUNG CHILDREN

The results of temporal resection for medically intractable epilepsy in 20 children less than age 5 years with at least 2 years follow-up are reported from Miami Children's Hospital, Florida. The mean age at surgery was 26 months, and the mean age at seizure onset was 12 months. Seizures were typical psychomotor in 4 patients, with staring and oral or gestural automatisms; and psychomotor plus in 7, with aura and frightened appearance, staring, decreased responsiveness followed by movements that were contraversive, lateralized tonic or clonic, and asymmetric tonic posturing. Motor symptoms were prominent in 3, with tonic asymmetric posturing followed by circling and vegetative signs. Clusters of epileptic spasms occurred in 6, mainly clonic. Interictal EEGs showed lateralizing abnormalities in 15 that were concordant, and ictal EEGs were lateralizing and concordant in 18 and nonlateralizing in 2. Brain MRI revealed localizing pathology in 16, and ictal SPECT was concordant in 4/8 cases. Invasive EEG recording was performed in 6 children to delineate the epileptogenic zone and map language cortex. Electrocorticography was performed in the remaining 14 cases. At mean follow-up of 5.5 years following surgery, 65% were seizure-free and 15% had >90% seizure reduction. The etiologic pathology was a tumor in 8 cases, benign developmental in 4 and malignant astrocytoma in 4. Focal cortical dysplasia was found in 6 cases, one with neurofibromatosis. Hippocampal sclerosis was identified in 4. Other pathologies included encephalitis, prior hypoxic-ischemic event, tuberous sclerosis, and

white matter gliosis, 1 of each. Patients with the most favorable outcome had psychomotor type seizures, tumor, and complete resection without complications. Stroke occurred in 2 and infection and hydrocephalus in 1. (Maton B, Jayakar P, Resnick T, Morrison G, Ragheb J, Duchowny M. Surgery for medically intractable temporal lobe epilepsy during early life. **Epilepsia** Dec 2007;49(1):80-87). (Reprints: Michael Duchowny MD, Miami Children's Hospital, Department of Neurology, 3200 SW 60th Court, Suite 302, Miami, FL 33155).

COMMENT. Cortical resection limited to one temporal lobe for refractory temporal lobe epilepsy is rare in children less than 5 years. This report indicates that surgery in this age group can be associated with favorable outcome, similar to that in older children.

Surgery for epilepsy in children ages 1 – 15 years. A report from Milan, Italy, found that 60% of 113 patients younger than 16 years (mean age at surgery of 8.8 years) were seizure free following excision of the epileptogenic zone for refractory focal seizures (Cossu M et al. **Epilepsia** Dec 2007;49(1):65-72). Variables associated with a significantly lower risk of seizure recurrence were unifocal lesion on MRI, older age at seizure onset, temporal unilobar resection and complete lesionectomy, and glial-neuronal tumor pathology. Results of surgery were strongly dependent on presurgical identification and resection of the epileptogenic zone.

Propeller MRI sequencing for detailed imaging of hippocampal sclerosis. This method is superior to routine MRI sequencies for identifying subtle hippocampal sclerosis (HS), and negates the effects of movement during scans (Eriksson SH et al. **Epilepsia** Dec 2007;49(1):33-39). Signs of HS on MRI are increased hippocampal signal on T2-weighted images and loss of hippocampal volume on T1-weighted images. Periodically Rotated Overlapping Parallel Lines with Enhanced Reconstruction (PROPELLER) sequence compensates for head motion during the MRI scan.

GENETIC VARIATION IN CALCIUM CHANNEL GENE IN IDIOPATHIC GENERALIZED EPILEPSIES

Researchers at Women's And Children's Hospital, North Adelaide, and other centers in Australia and Canada screened 240 individuals from 167 families with idiopathic generalized epilepsy and generalized epilepsy with febrile seizures plus (GEFS+) and 95 controls for variants in the *CACNA1H* gene. They identified 19 novel variants causing amino acid changes associated with the following epilepsy syndromes: childhood absence epilepsy, juvenile absence, juvenile myoclonic, idiopathic generalized with tonic-clonic seizures, and temporal lobe epilepsy. The variants also occurred in unaffected individuals. In some families, the variant segregated with epilepsy, but not in others. It is concluded that variants in *CACNA1H* gene that alter channel properties occur in patients with various generalized epilepsy syndromes, contributing to epilepsy susceptibility but not sufficient to cause epilepsy themselves. (Heron SE, Khosravani H, Varela D, et al. Extended spectrum of idiopathic generalized epilepsies associated with CACNA1H functional variants. **Ann Neurol** Dec 2007;62:560-568). (Respond: Sara E Heron, Department of Genetic Medicine, Women's and Children's Hospital, 72 King William Road, North Adelaide SA 5006, Australia).