## SEIZURE DISORDERS

## INTRACTABLE SEIZURES AFTER ANTIEPILEPTIC WITHDRAWAL

The frequency of relapse and occurrence of intractable epilepsy following withdrawal of antiepileptic drug (AED) treatment in a Nova Scotia population-based cohort of children with epilepsy was studied at Dalhousie University, Halifax, NS, Canada. Patients (n=367), ages 1 month to 16 years, had partial or convulsive seizures, and at least 5 years follow-up. Those with benign rolandic epilepsy were excluded. Intractability was defined as one or more seizures every 3 months during the last year of follow-up despite maximum tolerated doses of 3 or more AEDs. Seventy one percent (260/367) had become seizure free for 1 to 4 years before discontinuing AEDs. Thirty percent had recurrence of seizures, but only 3 (1%) with recurrences developed intractable seizures. Two were eventually controlled, 1 after temporal lobectomy. Factors predictive of relapse were not determined. (Camfield P, Camfield C. The frequency of intractable seizures after stopping AEDs in seizure-free children with epilepsy. **Neurology** March (2 of 2) 2005;64:973-975). (Reprints: Dr Peter Camfield, IWK Health Centre, 5850/5980 University Ave, PO Box 9700, Halifax, NS, B3K 6RS, Canada).

COMMENT. While one third have seizure recurrences, only 1% of children discontinuing treatment after 1 to 4 years of AED control will develop intractable epilepsy. The authors comment that their study group was evaluated before MRIs were available, and the advent of MR neuroimaging may have provided clues to predict cases at risk of epilepsy intractability.

The EEG and Risks of AED withdrawal. The EEG may be of value in predicting relapse during withdrawal of AEDs, according to a study of 128 patients (mean age 28 years) with partial epilepsies. (Tinuper P et al. Neurology 1996;47:76-78; Ped Neur Briefs Aug 1996). Worsening of EEG abnormalities after starting drug withdrawal was more predictive of relapse than pre-withdrawal dysrhythmias or slowing. Several studies of the EEG and drug withdrawal have shown conflicting results regarding its value in predicting outcome (see Progress in Pediatric Neurology I and III, PNB Publishers, 1991 and 1997). Interictal EEG slowing was predictive of seizure recurrence in a prospective cohort study of 264 children (Shinnar S et al. Ann Neurol 1994;35:534-545; Ped Neur Briefs June 1994). Approximately one third relapsed during AED withdrawal, after 2 or more years of seizure control, a similar relapse rate to that reported in the current Camfield study.

## CORTICAL TRIGGERS IN GENERALIZED REFLEX EPILEPSIES

The electroclinical patterns of generalized reflex seizures (GRS) triggered by sensory or cognitive stimuli are reviewed in a report from Montreal Neurological Hospital, Canada; and Universita degli Studi di Messina, Italy. Four major patterns of GRS having a focal cortical trigger are identified: 1) photosensitive seizures are subserved by the occipital cortex; 2) tacile or noise-induced seizures, by the sensorimotor cortex; 3) cognitive-induced (mental arithmetic, block design) by the non-dominant parietal lobe; and 4) reading, talking, and writing-induced epilepsies, by dominant frontotemporal lobes. Clinically, the seizures induced by these stimuli are idiopathic generalized epilepsies, especially juvenile myoclonic epilepsy, and the cortical mechanisms are complex, ultimately involving cortico-reticular or cortico-cortical pathways. (Ferlazzo E, Zifkin BG, Andermann E, Andermann F. Cortical triggers in generalized reflex seizures and epilepsies. **Brain** April 2005;128:700-710). (Respond: Benjamin G Zifkin MDCM FRCPC, Epilepsy Clinic, Montreal Neurological Institute and Hospital, 3801 University Street, Room 128, Montreal, QC H3A 2BA, Canada).

COMMENT. The authors hypothesize that patients with cortical triggers of GRS present regions of cortical hyperexcitability overlapping with the areas activated during sensory stimulation and cognitive or planned motor (praxis) activities. When these hyperexcitable areas are activated sufficiently, epileptic activity is produced that involves cortico-reticular or cortico-cortical pathways, resulting in a generalized reflex seizure. Genetic or acquired lesions may be responsible for the neuronal hyperexcitability.

Pattern sensitive epilepsy, discussed in Ped Neur Briefs February 2005, has a higher incidence of focal symptomatic seizures than the generalized idiopathic reflex epilepsies. The EEG in pattern-sensitive epilepsies shows focal epileptiform discharges, whereas photosensitive epilepsies are usually accompanied by generalized polyspike-wave and spikewave complexes.

## PAROXYSMAL TONIC UPGAZE

This oculomotor syndrome of childhood, originally termed "benign paroxysmal tonic upgaze of childhood" (Ouvrier and Billson, 1988) is reviewed from the Children's Hospital of Westmead, Sydney, NSW, Australia. Since the original description of 4 cases, a total of 49 cases have been reported. The clinical features listed in the original cases were as follows: onset before 1 year of age; conjugate upward deviation of the eyes, with neck flexion; downbeating compensatory saccades; normal horizontal eye movements; fluctuation of symptoms during the daytime and relief in sleep; exacerbation during febrile illness; intermittent or persistent ataxia; otherwise normal neurologic exam; no deterioration and eventual improvement with long-term follow-up; normal EEG, CT, and CSF neurotransmitters. Some subsequent reports have described etiological factors, including: genetic, autosomal dominant or recessive inheritance in 4 families; fetal valproate exposure in 3 cases; and cerebral abnormalities in 5 (hypomyelination (2 cases), periventricular leukomalacia, Vein of Galen malformation, pinealoma). About 40% have learning or mild cognitive deficits, and 10% are moderately to severely retarded. Several have a history of febrile convulsions, 2 had epilepsy, but the EEG shows no epileptic activity during the tonic upgaze. Apart from the few cases with structural pathology involving the upper brainstem, a localization-related lesion is not evident. About 50% of cases have a favorable outcome, 25% have residual ataxia, and 25% may have strabismus or nystagmus as a sequel. Treatment with L-dopa is sometimes successful, but AEDs, including acetazolamide and ACTH, are of no benefit. (Ouvrier R, Billson F. Paroxysmal tonic upgaze of childhood - a review. Brain Dev April 2005;27:185-188). (Respond: Dr Robert Ouvrier, E-mail: roberto@chw.edu.au).

COMMENT. A tonic upgaze may be caused by a space-occupying lesion involving the brainstem, and an MRI is indicated in these infants, especially when ataxic.