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1 Review of pictures of the Dionne monozygote quintuplets confirmed facial changes related to epilepsy in one – G.M. Rémillard, A. Sherwin, E. Andermann, F. Andermann, B.G. Zifkin (Montréal, QC, Canada)

Pictures of the famous Dionne monozygotic quintuplets can be viewed on the Internet at http://www.city.north-bay.on.ca/quints/digitize/archivef.htm. They were born in Canada in 1934. They were the product of a sixth pregnancy, delivered 2 months prematurely. Marie, the last born, was the smallest and Émilie was the only one who was left-handed. She developed epilepsy at the age of 8 years, and was reported to have one attack every week until she choked and died during one of her seizures at the age of 20. The Dionne quintuplets were the subject of intense media attention in their childhood and adolescence. Review of the many pictures posted on the Internet, taken from birth to the end of their second decade, showed changes in Émilie's facial features appearing during adolescence and apparently caused by first-generation antiepileptic drugs, particularly phenytoin. The photographs also show that during her adolescence she developed left lower facial asymmetry seen when smiling for the camera. We presume that she had right temporal lobe epilepsy responsible for inversion of the constitutional right facial asymmetry seen in her mother and four sisters. We interpret Émilie's facial asymmetry as a form of hypodynamia related to right temporal lobe epilepsy during posed emotional facial expression.

2 Factor V Leiden and the hemi-convulsions, hemiplegia, epilepsy syndrome: a report of two cases – Morris Scantlebury, Lionel Carmant (Saint Justine Hospital, Montreal, QC, Canada)

The Factor V Leiden (FVL) mutation is the most common cause of venous thrombosis in European populations, being more prevalent than all other causes of hereditary thrombosis. In the pediatric population, it has been associated with cerebrovascular thrombosis, cerebral palsy and prosencephaly in neonates. Its association with other neuropathological conditions is undetermined and there has been no association with the hemi-convulsions, hemiplegia, epilepsy syndrome (HHE) reported in the literature. We present two cases of HHE with differing precipitating and complicating factors and whose cause is likely due to cerebrovascular thrombosis secondary to the FVL mutation. In one case, the initial presentation was at 18 months, preceded by a febrile illness and associated with a past history of prolonged febrile convulsions, whereas in case 2 the initial presentation occurred at 11 years old, was not preceded by a febrile illness, and there was no past medical history of febrile convulsions. Furthermore, a dual pathology of FVL and MTHFR C677T was diagnosed in case 2. We suggest that patients presenting with HHE should be routinely investigated for FVL and if positive, careful consideration should be given to therapeutic and prophylactic anti-coagulation as this may speed recovery and improve outcome.

3 EEG and epilepsy in some chromosomal disorders – Paul A. Hwang, Chen Li, Jane Shaw, Philip Wyatt (Department of Paediatrics, Bloorview Epilepsy Research Program, University of Toronto, North York General Hospital, Toronto, ON, Canada)

In assessing children with epilepsy, the EEG and neuroimaging studies often reveal underlying structural and/or functional abnormalities sufficient for clinical diagnosis and management. In a subset of children with dysmorphic features and developmental delay, additional cytogenetic testing may be required. In a retrospective review of over 1500 patients with epilepsy, mostly children and young adults, only about 2% have chromosomal anomalies detected by karyotype analysis, with fluorescent in situ hybridization techniques (FISH): Down syndrome with trisomy 21, and balanced Robertsonian translocation (2 cases), Angelman syndrome with and without deletions at 15q11–13 (15 cases), the Smith-Magenis syndrome (1 case) with deletion at chromosome 17p-, and the Wolff-Hirschorn Syndrome with deletion at 7p- (1 case). The EEG studies were quite variably abnormal: disturbance of background activity, multifocal epileptiform discharges, pseudoperiodic complexes and frequent epileptic seizures. The clinical seizures are variable and of many different types: partial, generalized atypical absences, tonic and myoclonic, often associated with stereotypic and autistic behavioral disorder, global developmental delay, while computed tomography (CT) or magnetic resonance imaging (MRI) may show some non-specific changes. The long-term prognosis is often quite dismal, and the seizures remain refractory to antiepileptic drugs. In the long-term follow-up of children with refractory seizures, it may be necessary to reassess earlier non-diagnostic studies, including chromosomal analysis (with FISH), diagnostic neuroimaging (preferably MRI rather than CT), and follow-up EEG studies (preferably with sleep or video-EEG monitoring), as the developing brain matures into adulthood, and technological advances provide a better 'window into brain mechanisms.'

4 High frequency transients in kindling identified using a calibrated amplitude wavelet transform (CAWT) – E.L. Ohayon[a,b], A.J. Mendonça[a], P.W. Tsang[c], H.C. Kwan[c], W.M. Burnham[a,b] ([a]Bloorview Epilepsy Research Program, [b]Institute of Medical Science and [c]Department of Physiology, University of Toronto, Toronto, ON, Canada)

Although spectral algorithms are a staple of electroencephalogram (EEG) analysis, the investigation of spectral changes in the most common animal model of complex partial seizures, the kindling model, has been surprisingly limited. Existing studies of kindling have been restricted to examining low frequencies or long periods thereby excluding the identification of high frequency transients. The oversight may be attributable to historical limitation in both the maximal sampling speed of older data acquisition equipment as well as the inherent shortcomings of commonly applied algorithms. In order to address these issues, recordings were collected using a custom-programmed data acquisition system using high sampling rates (2 kHz). The data were subjected to a novel form of transient analysis. Specifically, we altered a wavelet analysis algorithm so that it is consistently sensitive to signal amplitude across frequencies. We then examined hippocampal activity just prior to and following stimulus in stage 5 kindled rats. The resultant charts clearly demonstrate a dramatic rise in events in the 20–40 Hz band, with major frequency components going as high as 200 Hz. Whether these high-frequency transients reflect a fundamental mechanism or are best considered an epiphenomena may be indeterminate. However, the very identification and measurement of these afterdischarge-related high-frequency phenomena attests to the importance of such events as candidate markers for

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seizure detection and categorization. The successful use of this Calibrated Amplitude Wavelet Transform (CAWT) illustrates that the technique may be applied more broadly to other high-frequency phenomena that have been traditionally overlooked, including potential transients in human EEG studies.

5 Intractable temporal lobe epilepsy (TLE) with rare or absent interictal spikes ('oligospikers') – A. Rosati, A. Bernasconi, F. Andermann, N. Bernasconi, A Olivier, J. Gotman, F Dubeau (Montreal Neurological Hospital and Institute, 3801 Rue University, Montreal, QC, Canada H3A 2B4)

Background: TLE patients usually have a high incidence of positive interictal EEG tracings with frequent interictal spikes (IS). Only 2% of these patients have no ISs despite prolonged scalp EEG recordings.

Rationale: To analyze clinical, electrophysiological, and neuroradiological characteristics of a group of patients with non-lesional intractable TLE and rare or absent IS (oligospikers).

Design/Methods: We selected 31 oligospikers (mean age, 34 years; 11 men) investigated at the Montreal Neurological Institute and Hospital (MNI) between 1990 and 2000. We compared the clinical and laboratory characteristics of these individuals with a group of 27 age-matched (mean age, 38 years; 10 men) randomly selected non-lesional TLE patients who had frequent IS.

Results: Compared to TLE patients with frequent IS, oligospikers show a later seizure onset age (19 vs. 10 years, $P = 0.004$), shorter disease duration (14 vs. 28 years, $P < 0.001$), lower incidence of secondarily generalized tonic-clonic (SGTC) seizures (10 vs. 81%, $P < 0.001$), and no status epilepticus (0 vs. 22%). Volumetric magnetic resonance imaging also demonstrated that hippocampal atrophy was less common in oligospikers (55 vs. 96%, $P = 0.001$). However, there were no differences in family history of epilepsy, risk factors, antecedent of febrile convulsions, and type of medication between the two groups. Equally favorable surgical outcome (Engel's class I) was present in 18/23 oligospikers and 21/25 TLE patients with frequent IS (79 vs. 84%).

Conclusion: This study showed that a subgroup of patients with non-lesional TLE has no or few interictal spikes. These patients have a less severe epileptic disorder as suggested by a later age of seizure onset, shorter duration of epilepsy, less sever seizure types, and less frequent hippocampal atrophy. These features suggest that in some respect the epilepsy is less severe but still intractable and usually incapacitating. The absence or paucity of IS and the absence of hippocampal atrophy may explain the smaller number of oligospikers that were referred for surgery. Nevertheless, oligospikers have the same favorable surgical outcome compared to patients with frequent IS.

6 Vagus nerve stimulation for the treatment of medically refractory pediatric epilepsy – T.E. Thomas, A. Bouthillier, A. Lortie, J.-C. Marchal, C. Mercier, D. Chartrand, L. Carmant (Sainte-Justine Hospital, Montreal, QC, Canada)

Introduction: Left vagus nerve stimulation (VNS) is an increasingly popular surgical alternative for medically refractory epilepsy. However, specific indications, as well as efficacy in the pediatric population have yet to be clarified. The goal of this study was to evaluate the efficacy of VNS in a cohort of children implanted at our center, and to identify factors predictive of outcome.

Methods: The first 10 children who were implanted at our institution were followed prospectively 2 months before, and each month after VNS surgery. Selection criteria included age less than 18 years, seizure control refractory to multiple drug trials, and absence of a resectable seizure focus. Seizure frequency, stimulation parameters, efficacy of self-stimulation, and side effects were monitored at each visit. In addition, neuropsychological and quality-of-life evaluations were performed on each patient both before and 12 months after surgery. Average follow-up is 12.7 months (range, 2–17 months). Medical treatment was not modified during the first 6 postoperative monhs.

Results: Five boys and 5 girls had VNS surgery, with a mean age of 8.3 ± 3.3 years. Six patients had partial epilepsies, and 4 had generalized epilepsies, including one with Lennox-Gastaut syndrome. Preoperative seizure frequency was 275 per month. Postoperative seizure frequency showed a 63% decrease at 1 month ($P < 0.001$), with a trend towards further reductions in subsequent months up to 6 months. Mean seizure reduction at 6 months was 95% ($P < 0.001$), and this reduction was maintained at 12 months. Eight of 10 children had a $> 85\%$ decrease in seizure frequency. Two patients are seizure-free. Overall behavioral improvement was observed in all patients, but only one child demonstrated an improvement in global IQ. No factor was identified as a marker for favorable outcome; however, the extremely high baseline seizure rate, the relatively young age at surgery of this cohort, and the maintenance of full antiepileptic drug coverage during the first 6 months may all have been contributory.

7 Distribution and clinical impact of sleep disorders among the epilepsies – Alcibiades J. Rodriguez, Rahul Gupta, Michele R. Sammaritano, Grant W. Su, Bruce L. Ehrenberg (Department of Neurology, New England Medical Center, Boston, MA, USA)

In the kindled feline epilepsy model, Shouse has found an alteration in sleep patterns, but these studies do not show that an exacerbation in seizure frequency may result. Epilepsy is clearly exacerbated by sleep apnea, but heretofore the resultant sleep disruption has been correlated neither to seizure severity nor to type of epilepsy. We studied seizure type and severity vis-à-vis two common sleep disorders: obstructive sleep apnea (OSA) and periodic limb movements disorder of sleep (PLMS). Polysomnogram (PSG) results on 233 epilepsy patients (1990–1999) defined three groups: OSA ($n = 19$, 15 male), PLMS ($n = 22$, 8 male) and combined sleep disorders (CSD [$n = 6$, 5 male]) which were compared to age-/sex-matched epilepsy controls by number of antiepileptic drugs, PSG findings, and by seizure type and number during the preceding year. Total sleep time, sleep efficiency, REM sleep and stages 3/4 were significantly decreased in patients with OSA and PLMS compared to controls. Stage 1 and total arousals per hour were significantly increased in both disorders. The percentage of patients with focal seizures was significantly higher in OSA (85%), PLMS (70%) and CSD (100%) compared to controls (56, 48 and 50%, respectively) ($P < 0.05$ for all). Patients with exclusively nocturnal seizures were significantly over-represented in OSA ($P < 0.025$) but not PLMS. Seizure rate and number of drugs in use were significantly higher in OSA (21.6, 2.1) than controls (5.5, 1.3)($P < 0.05$, 0.02). In addition, the PLMS patients showed a trend ($P = 0.10$) toward a higher seizure rate than controls (7.0 vs. 2.6).

8 Antimyoclonic efficacy of piracetam in idiopathic generalized epilepsy – Y.A. Agha Khani, F. Andermann (Department of Neurology and Pediatrics, McGill University, and Epilepsy Service, Montreal Neurological Hospital, 3801 Rue University, Montreal, QC, Canada H3A 2B4)

We used piracetam, a derivative of γ-aminobutyric acid (GABA), in treatment of myoclonus in a 47-year-old woman with idiopathic generalized epilepsy (IGE). She had had only two generalized tonic-clonic seizures; the last was 10 years earlier. Morning myoclonic jerks were inconvenient. Her obesity precluded use of the sodium valproate. She had a dramatic response to piracetam with sustained cessation of myoclonus, and no side effects during 1.5 years of follow up. Further trials for control of myoclonus in patients with IGE are justified.