Essential Tremor Among Children

To characterize the clinical and therapeutic aspects of essential tremor (ET) among children ET an autosomal dominant disorder has been studied extensively among adults, but little is known regarding its occurrence, clinical characteristics, treatment and prognosis in pediatric populations. Often stigmatized as a disorder of the elderly, ET may be mis-diagnosed among children. Previous studies of childhood onset ET were limited by small sample sizes. Clinical data, including gender, age at nonset, family history, associated disorders and response to treatment were collected for consecutive patients diagnosed with childhood-onset ET at the Movement Disorders Clinic at Baylor College of Medicine. In the present study 39 patients with ET, 29 (74.4%) were male. The mean age at onset was 8.8 ± 5.0 years, and the mean age at evaluation was 20.3 ± 14.4 years. A family history of tremor was noted for 79.5% of the patients. Eighteen (46.2%) had some neurologic comorbidity, such as dystonia, which was noted for 11 patients (28.2%). Only 24 of the patients (61.5%) were treated with a specific antitremor medication; 5 of the 12 patients treated with propranolol experienced improvement. Concomitant movement disorders, such as dystonia, are common among patients with childhood onset ET, which supports the concenpt that ET is a heterogeneous disorder. Treatment strategies used for adult patients with ET seem to be effective also for children with ET, although controlled therapeutic trials in this population of patients with ET are lacking.

Abstracted from Joseph Jankovic, Jaswanth Madisetty and Kevin Dat Vuong, *Pediatrics* 2004; 5 : 1203.

The United Kingdom Infantile Spasms Therapy Trial

Background Infantile spasms, which comprise a severe infantile seizure disorder, have a high morbidity and are difficult to treat. Hormonal treatments (adrenocorticotropic hormone and prednisolone) have been the main therapy for decades, although little evidence supports their use. Vegabatrin has been recorded to have a beneficial effect in this disorder. We aimed to compare the effects of vigabatrin with those of prednisolone and tetracosactide in the treatment of infantile spasms, in a multicentre, randomised controlled trial in 150 hospitals in the UK. The primary outcome was cessation of spasms on days 13 and 14. Minimum doses were vigabatrin 100 mg/kg day, oral prednisolone 40 mg per day, or intramuscular tetracoactide depot 0.5 mg (40 IU) on alternate days. Analysis was by intention to treat. Findings of 208 infants screened and assessed, 107 were randomly assigned to vigabatrin (n=52) or hormonal treatments (prednisolone n=30, tetracosactide (n=25). None was lost to follow-up. Proportions with no spasms on days 13 and 14 were: 40 (73%) of 55 infants assigned hormonal treatments [(prednisolone 21/30 (70%)], tetracosactide 19/25 (76%) and 28 (54%) of 52 infants assigned vigabatrin (difference 19%, 95% CI 1%-36%, p=0.043). Two infants allocated tetracosactide and one allocated vigabatrin received prednisolone. Adverse events were reported in 30 (55%) of 55 infants on hormonal treatments and 28 (54%) of 52 infants on vigabatrin. No deaths were recorded. Cessation of spasms was more likely in infants given hormonal treatments than those given vigabatrin. Adverse events were common with both treatments.

Abstracted from Andrew L. Lux, Stuart W. Edward *et al*, *Lancet* 2004; 364 : 1773.
