

EARLY DEVELOPMENTAL OUTCOME FOLLOWING CONVULSIVE STATUS EPILEPTICUS

Investigators at the Developmental Cognitive Neurosciences Unit, Institute of Child Health, London, other centers in the UK, and Dartmouth School of Medicine, NH, USA, prospectively recruited children aged between 1 and 42 months from North London who had at least one episode of convulsive status epilepticus (CSE) and classified them as prolonged febrile seizures (PFS) or nonfebrile CSE. Of 54 patients receiving neuropsychological and MR imaging tests within 6 weeks (mean 38 days) of CSE (baseline) and at 1-year follow-up, 27 had PFS (mean age 18.4 months) and 27 had nonfebrile CSE (mean age 15.5 months). Children with nonfebrile CSE had similar seizure characteristics but a worse developmental outcome than children with PFS ($p < 0.002$). The PFS group had a worse developmental outcome than healthy controls (mean age 20.49 months) ($p = 0.002$). Tests at 1-year follow-up showed no significant improvement from baseline. Seizure characteristics were not predictive of performance. (Martinos MM, Yoong M, Patil S, et al. Early developmental outcomes in children following convulsive status epilepticus: A longitudinal study. *Epilepsia* 2013 Jun;54(6):1012-9). (Resp.: Dr Marina M Martinos. E-mail: m.martinos@ich.ucl.ac.uk).

COMMENT. Following convulsive status epilepticus (CSE), children including those with prolonged febrile seizures (PFS) are developmentally delayed. The impairments in development are still present 1 year post CSE, suggesting that premorbid abilities may be overshadowing any transient direct effects of CSE itself on outcome.

FS duration and Developmental Delay. The FEBSTAT Study Team investigated the association between FS duration and baseline characteristics of development in 158 children with a first FS, median duration 4 minutes (Hesdorffer DC, et al. *Ann Neurol* 2011 Jul;70(1):93-100). One population was identified that accounted for 82.3% of FSs and had a mean duration of 3.8 min (short FS) and a second population accounting for 17.7% of FSs with a mean duration of 39.8 min (long FS). Long FSs were significantly associated with developmental delay ($p = 0.01$) and younger age at first FS ($p = 0.048$). The data provide support for redefining a simple febrile seizure, limiting the duration to no longer than 10 min.

SLEEP DISORDERS AND EPILEPSY

An investigator from University of Oxford, UK, reviews the effect of sleep disorders on epilepsy and the effects of epilepsy on sleep. The occurrence of seizures is modified by the circadian sleep-wake cycle, the stage of sleep, sleep deprivation, and obstructive sleep apnea. Certain seizures, such as Rolandic epilepsy, are accentuated during sleep. Nocturnal epilepsy may cause arousals in sleep, and circadian sleep-wake rhythms may be disrupted and cause impaired cognitive function and behavior during the day. Antiepileptic drugs may have indirect effects on sleep. For example, barbiturates cause daytime sleepiness, and phenytoin causes insomnia.

Sleep disorders confused with epilepsy include parasomnias, arousal disorders, sleepwalking, sleep terrors, rhythmic movement disorders (headbanging), nightmares,

panic attacks, and REM sleep behavior disorder. Epilepsies confused with sleep disorders include Rolandic epilepsy, Panayiotopoulos syndrome, occipital epilepsy of Gastaut, and nocturnal frontal lobe epilepsy (NFLE) in which seizures occur mainly during sleep. Guidelines are suggested for differentiating non-epileptic parasomnias from nocturnal seizures, and NFLE from arousal disorders. Non-convulsive status is sometimes misinterpreted as a sleep disorder. (Stores G. Sleep disturbance in childhood epilepsy: clinical implications, assessment and treatment. **Arch Dis Child** 2013 Jul;98(7):548-51). (Response: Professor Gregory Stores, Department of Psychiatry, University of Oxford, UK. E-mail: gregory.stores@psych.ox.ac.uk).

COMMENT. The author concludes that screening for sleep disturbance should be routine in children with epilepsy, and the accurate diagnosis determined, according to the **International Classification of Sleep Disorders: Diagnostic and Coding Manual** (2nd ed. Westchester, IL: American Academy of Sleep Medicine; 2005).

HEREDO-DEGENERATIVE / METABOLIC DISORDERS

CHARACTERISTICS OF INDIVIDUALS AT RISK FOR SPINOCEREBELLAR ATAXIA

Investigators at University Hospital of Bonn and 13 other centers in Germany, France, and Italy conducted a prospective, longitudinal observational study (2008-2011) of offspring or siblings of patients with spinocerebellar ataxias (SCA)-1, 2, 3, and 6. Study individuals had no ataxia and were aged 18-50 years (35-70 years, if directly related to individuals with SCA6). Relations between outcome variables and time from onset (present age to estimated age at ataxia onset) were analyzed using clinical scales, questionnaires, and coordination tests. In 264 participants, estimated time to ataxia from onset was -9 years in 50 carriers of the SCA1 mutation, -12 years in 312 SCA2 mutation carriers, -8 years in 26 SCA3 mutation carriers, and -18 years in 16 SCA6 mutation carriers. Compared with non-carriers of each mutation, SCA1 and SCA2 mutation carriers had higher median scores for ataxia. SCA2 carriers had lower functional index scores than did non-carriers and worse composite cerebellar functional scores than did their non-carrier counterparts. In 83 individuals (30%) who underwent MRI, brainstem and cerebellum showed grey-matter loss in SCA1 and SCA2 mutation carriers. (Jacobi H, Reetz K, Montcel ST, et al. Biological and clinical characteristics of individuals at risk for spinocerebellar ataxia types 1, 2, 3, and 6 in the longitudinal RISCA study: analysis of baseline data. **Lancet Neurol** 2013 Jul;12(7):650-8). (Response: Prof T Klockgether. E-mail: klockgether@uni-bonn.de).

COMMENT. The authors summarize the baseline findings of the RISCA study designed to define the preclinical stages of SCAs in individuals at risk and to identify functional and brain structural abnormalities before the onset of ataxia. Carriers of SCA1 and SCA2 mutations have mild coordination functional deficits and brain structural abnormalities before the onset of clinically manifest ataxia. Recognition of the preclinical manifestations of neurodegenerative diseases may permit early therapeutic intervention.