

(GA) and seizure focus by retrospective chart review of 90 children (aged 6-18 years) with epilepsy, video-EEG recording, MRI, and neuropsychological testing (WISC-IV). Cognitive Proficiency Index (CPI) scores based on the WISC-IV Working Memory (WM) and Processing Speed (PS) indices were significantly lower than the General Ability Index (GAI) scores, comprising the WISC-IV Verbal Comprehension (VC) and Perceptual Reasoning (PR) indices. GAI>CPI differences were significantly greater in the right than left lateralized seizure group and also greater for the frontal than temporal group. CP was selectively compromised in those with seizures lateralized to the right hemisphere or localized to the frontal lobe. Right lateralization and frontal localization independently impact CP. GAI>CPI differences were significantly greater in the right-lateralized group than the generalized group and in the frontal-localized group than the generalized group. Deficits in CP are a defining neurocognitive characteristic of pediatric epilepsy in individuals with both focal and generalized onset, but especially when seizures originate from a primary epileptogenic focus within the right hemisphere or the frontal lobe. (Gottlieb L, Zelko FA, Kim DS, Nordli DR Jnr. Cognitive proficiency in pediatric epilepsy. *Epilepsy Behav* 2012;23:146-151). (Respond: Dr Frank A Zelko, Children's Memorial Hospital, #10, 2300 Children's Plaza, Chicago, IL 60614. E-mail: fzelko@childrensmemorial.org).

COMMENT. Children with epilepsy lateralized to the right hemisphere or localized to the right frontal lobe are at increased risk of cognitive deficits involving working memory and processing speed. Working memory maintains short-term information, and processing speed determines the amount of information that can be used and accommodated in working memory. Cognitive proficiency contributes to cognitive aptitude in learning and problem solving. A general availability of psychological services should add to the proficiency of epilepsy management in the clinic.

SHARED GENETIC BASIS OF EPILEPSY AND BEHAVIOR DISORDERS

Researchers from Columbia University, New York; UCLA; Northern Illinois University, DeKalb, IL; and Northwestern Children's Memorial Hospital, Chicago have examined whether the first-degree family history of unprovoked seizures in 308 probands with childhood onset epilepsy is associated with behavioral disorders. The association was assessed separately in uncomplicated and complicated epilepsy and for febrile seizures. Median age at onset was 4.2 years, and age at time of 9-year interview was 13.5 years. Epilepsy was uncomplicated in 213 (69.2%) and complicated in 95 (30.8%). Family history of unprovoked seizure was present in 24 probands with uncomplicated epilepsy (11.3%) and 9 probands with complicated epilepsy (9.5%). Family history of febrile seizures was present in 21 probands with uncomplicated (9.9%) and in 8 with complicated epilepsy (8.4%).

In probands with uncomplicated epilepsy, first-degree family history of unprovoked seizure was significantly associated with internalizing disorders, withdrawn/depressed, affective and anxiety disorders, aggressive and delinquent behavior, conduct disorder and oppositional defiant disorder. In probands with complicated epilepsy, family history of unprovoked seizure and behavioral problems

were not associated. Also, first-degree family history of febrile seizure was not associated with behavioral problems in probands with uncomplicated or in those with complicated epilepsy. The familial clustering of these disorders suggests that behavioral disorders may be another manifestation of the underlying pathophysiology involved in or related to epilepsy. (Hesdorffer DC, Caplan R, Berg AT. Familial clustering of epilepsy and behavioral disorders: evidence for a shared genetic basis. *Epilepsia* Feb 2012;53(2):301-307). (Respond: Dr Dale C Hesdorffer, GH Sergievsky Center, Columbia University, 610 West 168th St, P & S Unit 16, New York, NY 10032. E-mail: dch5@columbia.edu).

COMMENT. Epilepsy and behavioral disorders appear to have a common underlying genetic predisposition, whereas in the above study febrile seizure had no significant familial association with behavioral disorders. Previous reports of behavior disorder in children with febrile seizure have varied findings. Friderichsen C and Melchior J (*Acta Paediatr* 1954;43:307-317) found behavior disorders in 12 (4%) of 282 febrile seizure patients, and Millichap JG et al (*Neurology* 1960;10:643-653) in a prospective study of 110 febrile seizure patients reported recurrent episodes of aggressive behavior, temper tantrums, and hyperactivity in 35% patients. Patients with a history of birth trauma and those with cryptogenic epilepsies were excluded from the Friderichsen and Melchior series of febrile seizures but not from the study by Millichap and colleagues.

Risk of behavioral, developmental, and physical comorbidities with epilepsy/seizure disorder in a nationally representative sample of US children. (Russ SA, Larson K, Halfon N. *Pediatrics* February 2012;129(2):256-264). Estimated lifetime prevalence of epilepsy/seizure disorder was 1%, and of current epilepsy/seizure disorder was 6.3/1000. Children with current epilepsy/seizure disorder were significantly more likely than those never affected to have ADHD (23% vs 6%), developmental delay (51% vs 3%), autism (16% vs 1%), and headache (14% vs 5%). Those with prior but not current seizures had lesser risks.

NEUROMUSCULAR DISORDERS

SPINAL MUSCULAR ATROPHY II/III AND FEEDING PROBLEMS

Researchers at Kaohsiung Medical University Hospital, Taiwan studied the prevalence and risk factors of feeding and swallowing problems in 108 genetically confirmed patients with types II and III spinal muscular atrophy (SMA), age range 3-45 years, 60 with type II and 48 with type III. A questionnaire survey showed the 3 most common feeding and swallowing difficulties were choking (30.6%), difficulty conveying food to the mouth (20.4%), and difficulty chewing (20.4%). Motor function status (sitters vs walkers) was an independent risk factor for feeding and swallowing difficulties; 28 were walkers, 76 sitters, and 4 nonsitters. All 4 SMA II nonsitters had feeding and swallowing difficulties. Poor head control when feeding was a factor in 13 (12%) patients. Age was not an independent risk factor in this study; 10 patients, all with type II SMA and age <20 years (range 3-19 years), had feeding and swallowing difficulties and required respiratory management. Respiratory assistance or suction was required in 17