

the number of patients with complicated epilepsy who have underlying neurologic abnormalities and are susceptible to infections, especially pneumonia. The authors conclude that the increase in mortality in this group of patients might be prevented by supportive care and improved infection control, not solely by improvement in seizure management. The mortality rate in children with uncomplicated epilepsies is not significantly greater than that for the general population.

Of 13 deaths related to seizures (almost 20% of all deaths in this combined cohort of new-onset childhood epilepsies) 10 (77%) were attributed to SUDEP. Risk of SUDEP varies with age and is higher in adults. The authors of the current study draw attention to the increased risk during the transition from adolescence to adulthood, a period associated with reduced sleep, irregular adherence to anticonvulsant dose schedules, excess alcohol, and other stresses that increase susceptibility to seizures.

In a recent prospective study of 245 children with childhood-onset epilepsy followed for almost 40 years, 60 subjects had died, and 33 (55%) deaths were epilepsy-related including SUDEP in 23/60 (38%), status epilepticus in 4 (7%), and accidental drowning in 6 (10%). The higher mortality rates reported in this cohort are related to duration of follow-up, most of the mortality beginning in adolescence and years after the onset of epilepsy (Sillanpaa M, Shinnar S. **Epilepsy Behav** 2013 Aug;28(2):249-55).

POLYMICROGYRIA-ASSOCIATED EPILEPSY

Investigators from the Boston Children's Hospital, New York University, Brown University, and Birmingham School of Medicine, AL, studied the clinical epilepsy and imaging features of 87 patients with polymicrogyria (PMG) and epilepsy, recruited through the Epilepsy Phenome/Genome Project. Median age of seizure onset was 3 years (range <1 month to 37 years). Seizures were focal in 87.4%, some in combination with generalized seizures (23%). Of generalized seizures, infantile spasms were the most prevalent, occurring in 45.2%. MRI showed a bilateral PMG pattern in 56.7% and perisylvian in 77%. Generalized PMG presented with an earlier age of seizure onset (median age of 8 months) and an increased prevalence of developmental delay prior to seizure onset (57.1%). Perisylvian PMG was unilateral in 43.3%. Seizures lateralized to the same hemisphere as the PMG or the hemisphere with greater involvement in those with unilateral or asymmetric PMG, with trend toward more right-sided involvement. (Shain C, Ramgopal S, Fallil Z, et al. Polymicrogyria-associated epilepsy: A multicenter phenotypic study from the Epilepsy Phenome/Genome project. **Epilepsia** 2013 Aug;54(8):1368-75). (Response: Annapurna Poduri, Epilepsy Genetics Program, Division of Epilepsy and Clinical Neurophysiology, Fegan 9, Boston Children's Hospital, Boston, MA 02115. E-mail: Annapurna.poduri@childrens.harvard.edu).

COMMENT. Polymicrogyria (PMG), a developmental brain malformation, is associated with variable clinical findings dependent on the localization of the lesion. In addition to generalized epilepsy, bilateral PMG may also feature pseudobulbar signs, cognitive impairment, and developmental delay. Seizures are often medically refractory and not surgically responsive or appropriate. Prior to the introduction of the MRI, PMG may have been misdiagnosed as ulegyria or cerebral cortical sclerosis due to perinatal or

postnatal hypoxic-ischemia. Ulegyria that is bilateral and perisylvian may also be manifested by epilepsy and pseudobulbar palsy.

Perisylvian Ulegyria Pseudobulbar syndrome. In a report of 12 patients with perisylvian ulegyria, medically refractive seizures responded to resective surgery in 4 patients, despite the bilateral distribution of cerebral sclerosis. The recognition of ulegyria and distinction from PMG as the cause of a perisylvian pseudobulbar palsy syndrome is important in treatment and prognosis of the complicating medically refractive seizures. (Schilling LP, Kieling RR, Pascoal TA, et al. Bilateral perisylvian ulegyria: An under-recognized, surgically remedial epileptic syndrome. **Epilepsia** 2013 Aug;54(8):1360-7). (Response: Andre Palmini. E-mail: apalmini@uol.com.br).

HEADACHE DISORDERS

CRANIAL AUTONOMIC SYMPTOMS IN PEDIATRIC MIGRAINE

Investigators at the University of California, San Francisco, examined the frequency of cranial autonomic symptoms in all pediatric and adolescent patients with migraine seen in 4 different clinical settings during July 2010 to June 2012. Of 125 patients, mean age 13.1 yrs (range 4-17), 60% were female, 46% had chronic migraine and 54% episodic migraine. Headache was unilateral in 16%, bilateral in 52% and variable in 26%. The majority (49%) was seen in a pediatric headache subspecialty clinic, and 22% in a general child neurology clinic. At least one cranial autonomic symptom was experienced by each of 91 (73%) patients. In order of decreasing frequency, aural fullness was experienced by 30%, facial flushing/sweating 25%, lacrimation 25%, conjunctival injection 23%, ptosis 20%, grittiness in eye in 20%, nasal congestion in 15%, rhinorrhea in 10%, and periorbital edema in 8%. The majority had more than one cranial autonomic symptom, usually bilateral. The likelihood of autonomic symptoms was not related to age, sex, headache laterality, aura, or episodic vs chronic headache. Autonomic symptoms are the rule rather than the exception in pediatric/adolescent migraineurs, and headache with autonomic symptoms involving eye and nose should not be misdiagnosed as sinus headache. (Gelfand AA, Reider AC, Goadsby PJ. Cranial autonomic symptoms in pediatric migraine are the rule, not the exception. **Neurology** 2013 Jul 30;81(5):431-6). (Response: Dr Gelfand. GelfandA@neuropeds.ucsf.edu).

COMMENT. Cluster headache or trigeminal autonomic cephalalgia is typically suspected when headache is complicated by cranial autonomic symptoms. This study shows that pediatric/adolescent migraine is also frequently associated with autonomic symptoms. A sense of aural fullness, the most common autonomic symptom in the San Francisco cohort, will be added to the list of cranial autonomic symptoms in the International Classification of Headache Disorders, 3rd edition-beta **Cephalalgia** 2013 Jul;33(9):629-808 (cited by Gelfand AA et al. **Neurology** 2013 Jul 30;81(5):431-6).