

# PEDIATRIC NEUROLOGY BRIEFS

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### SEIZURE DISORDERS

#### RISKS OF MORTALITY IN NEW-ONSET EPILEPSY

Investigators at Lurie Children's Hospital of Chicago; Mayo Clinic, Rochester, MN; and centers in Connecticut, the Netherlands and Nova Scotia, pooled data from 4 large pediatric cohorts and obtained direct estimates of the mortality risk overall and from specific causes of mortality, particularly seizure related SUDEP. Of 2239 subjects followed for >30,000 person-years, 79 died at average age of 11.6 y, an overall-death rate of 228 per 100,000PY (10 with neurometabolic disorders were excluded). The death rate in the complicated group of epilepsies (743 per 100,000) was higher than in the uncomplicated group (36 per 100,000) ( $P<0.0001$ ). Ten deaths were attributed to SUDEP (5 definite, 3 probable, and 2 possible). Three additional deaths were seizure-related (1 status epilepticus, 1 complications of treatment, and 1 aspiration), seizure-related deaths accounting for 19% of all deaths. Seizure-related death rates were 43 overall, 122 for complicated epilepsy, and 14 for uncomplicated epilepsy. Of 48 deaths from other natural causes, 35 were due to pneumonia, 3 sepsis, 3 from shunt malfunction, 1 each from tracheotomy malfunction, pulmonary embolism, and cardiomyopathy, and 2 each from cerebral hemorrhage and cancer. Five deaths were due to non-natural causes: 2 accidents, 2 suicides, and 1 homicide. Compared with the general population, mortality in children with complicated epilepsy is significantly higher but not in those with uncomplicated epilepsy. Most excess death in young people with epilepsy is not seizure-related. (Berg A T, Nickels K, Wirrell EC, et al. Mortality risks in new-onset childhood epilepsy. *Pediatrics* 2013 Jul;132(1):124-31). (Respond: Dr Anne T Berg, Ann & Robert H. Lurie Children's Hospital of Chicago, 225 E Chicago Ave, Box 29, Chicago, IL 60611. E-mail: [atberg@luriechildrens.org](mailto:atberg@luriechildrens.org)).

COMMENT. The increased death rate in children and young adults (1- to 29-year-olds) followed from their initial diagnosis of childhood epilepsy is accounted for by

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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](mailto: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