

COMMENT. Familial BAVM is rare with only 25 families reported. Familial BAVMs present at a younger age than sporadic cases, and clinical anticipation of the disorder may occur in children with BAVM born to affected parents.

Presenting symptoms, surgical management and outcome of 39 pediatric cases of AVM are reviewed by Klimo P Jr et al. *Child's Nerv Syst* 2007;23:31-37; *Ped Neur Briefs* Jan 2007;21:1-2.

## SEIZURE DISORDERS

### LONG-TERM OUTCOME OF NEONATAL SEIZURES

Neonates with seizures prospectively diagnosed in a population-based setting in Newfoundland, Canada, between 1990 and 1995, were followed to a median age of 10 years. The incidence of epilepsy, and physical and cognitive impairments were analyzed for 62 term and 26 premature infants. Outcome was normal in 31 children (35%); 36 (41%) had neurodevelopmental impairments; and 21 (24%) had died. The median age at death was 13 months; range 2 days to 14 years. Gestational age was a better predictor of outcome than birth weight. Normal outcome occurred more often in term than preterm infants ( $p=0.003$ ). Postneonatal epilepsy developed in 27 (34%), including 29% of term and 48% preterm infants. Median age at onset of epilepsy was 9 months in term and 4 months in preterm infants. Epilepsy complicating mental retardation (MR) and cerebral palsy (CP) was correlated with early death. CP occurred in 35% of those surviving infancy, MR in 32%, and learning disorders in 23%. Neonatal encephalopathy occurred in 42%, usually following hypoxic-ischemic events. Infants with Sarnat stage III encephalopathy either died or had MR, CP, and symptomatic epilepsy. Infections occurred in 19%; outcome was normal in term infants with infection, but poor in premature infants or with CMV infection.

Abnormal neonatal EEGs, usually an abnormal background activity, correlated closely with poor outcome ( $p<0.0001$ ). Other variables associated with a poor prognosis included cerebral dysgenesis, intraventricular hemorrhage, and need for multiple AEDs. Purely clonic seizures without facial involvement in term infants had a favorable prognosis, whereas generalized myoclonic seizures in preterm infants were associated with high mortality. Subtle seizures, generalized tonic seizures, and number of seizures were not predictive of outcome. The severity and timing of the brain insult are the major determinants of outcome. (Ronen GM, Buckley D, Penney S, Streiner DL. Long-term prognosis in children with neonatal seizures. *Neurology* Nov 6, 2007;69:1816-1822). (Reprints: Dr Gabriel M Ronen, HSC 3N11, 1200 Main Street West, Hamilton, Ontario, Canada L8N3Z5).

COMMENT. In an editorial, Dlugos D and Sirven JI (Prognosis of neonatal seizures. "It's the etiology, Stupid" – or is it? *Neurology* 2007;69:1812-1813) refer to the study by Mizrahi E and Kellaway P. (*Neurology* 1987;37:1837-1844) who found, using video-EEG, that focal clonic seizures and focal tonic seizures were consistently associated with ictal activity on EEG. In contrast, generalized myoclonic events showed ictal EEG correlation in 60% of cases. Motor automatisms (subtle seizures), generalized tonic events, and focal myoclonic jerks were not associated with EEG ictal activity, suggesting that these clinical events may not be epileptic. A distinction was made between clinical events that were epileptic and those associated with an encephalopathy but were not neonatal seizures. The

report by Ronen and associates stresses the unfavorable prognosis of neonatal seizures, especially in preterm infants, and with the exception of clonic seizures without facial involvement. Outcome is determined primarily by gestational age and the severity and timing of the encephalopathy resulting in seizures. The authors emphasize the need for preventive measures and antiepileptic medications more specific for neonatal seizures.

A study of the etiology and neurodevelopmental outcome of seizures in *term* newborn infants (Tekgul H et al. **Pediatrics** 2006;117:1270-1280; **Ped Neur Briefs** April 2006;20:29-30) found that neonatal mortality was 7%, and neurologic outcome of survivors at 1 year was favorable in 72%. Predictors of a favorable outcome were a normal neonatal neurologic exam and normal EEG. Global cerebral hypoxic-ischemia is the most frequent cause (40%) of neonatal seizures and a strong predictor of poor long-term outcome. The evidence favors the etiology as the most important factor in prognosis and not the neonatal seizure per se. (Camfield PR. **Epilepsia** 1997;38:735-737). In developing laboratory animals, a major tonic-clonic seizure with post-ictal depression cannot be induced until the animal is older and cortical organization more mature. (Millichap JG. **Proc Soc Exper Biol & Med** 1957;96:125-129).

## PAROXYSMAL NONEPILEPTIC EVENTS

Paroxysmal events that mimic epilepsy, and their precipitants, prodromes, and distinguishing features are reviewed by researchers at Texas Tech University, Lubbock, TX, and American University of Beirut, New York. They include syncope, long QT disorders, breath-holding episodes, compulsive self-induced valsalva syncopal convulsions, apneic episodes, hyperkplexia, familial hemiplegic migraine, basilar migraine syncope, benign paroxysmal vertigo, alternating hemiplegia, Alice in Wonderland syndrome, psychogenic seizures, motor tics, paroxysmal torticollis, paroxysmal dyskinesias, dystonic choreoathetosis, kinesigenic choreoathetosis, paroxysmal dystonia induced by exercise, episodic ataxia, benign myoclonus of early infancy, shuddering attacks or tremors, neonatal jitteriness, Sandifer syndrome (gastroesophageal reflux), stereotypies (repetitive movements), paroxysmal tonic upward gaze, oculomotor apraxia, spasmus nutans, opsoclonus, daydreaming, sleep disorders, nonrapid eye movement arousal disorders, rapid eye movement nightmares or sleep paralysis, sleep transition head banging and body rocking, benign neonatal sleep myoclonus, and narcolepsy-cataplexy syndrome. These potential mimickers of epilepsy may be classified according to age of occurrence and clinical presentation. (Obeid M, Mikati MA. Expanding spectrum of paroxysmal events in children: potential mimickers of epilepsy. **Pediatr Neurol** Nove 2007;37:309-316). (Dr Mikati, Epilepsy Program, Department of Pediatrics, American University of Beirut, 850 3<sup>rd</sup> Ave, 18<sup>th</sup> Floor, New York, NY 10020).

COMMENT. As presented in this extensive review, paroxysmal nonepileptic events in infants and children comprise an expanding spectrum. The misdiagnosis of epilepsy in children with apparent refractory seizures, reported to be as high as 39% (Uldall P et al. **Arch Dis Child** 2006;91:219-221; cited by reviewers), leads to unnecessary investigation and potentially toxic antiepileptic therapy. Diagnosis of a nonepileptic event requires a careful history and a normal ictal EEG.