

Cerebellar mutism occurs primarily as a complication of posterior fossa tumor resection. Other nonsurgical causes of cerebellar mutism in children include cerebellitis, stroke, cavernous malformation, and hemolytic uremic syndrome. Cerebellar mutism usually presents as a feature of the posterior fossa (cognitive affective) syndrome, with neurobehavioral and personality changes. MRI reveals damage to the dentate nucleus and dentothalamic tract. The outcome is variable.

Postoperative cerebellar mutism syndrome (CMS): MRI features and origin are reported in 28 children with medulloblastoma treated at Children's National Medical Center, Washington, DC. (Wells EM et al. *JNS Peds* April 2010;5(4):online; Respond: Roger J Packer MD. E-mail: rpacker@cnmc.org). Preoperative MR images show a significant association with brainstem invasion and a trend toward cerebellomedullary angle involvement. Immediately postoperative images show cerebellar edema in 92%, especially middle and superior cerebellar peduncle edema. At 1 year after surgery, patients with CMS show atrophy/gliosis of total cerebellum, vermis, and brainstem, and cognitive deficits. Mean IQ was 16 points lower in patients with CMS compared with those without. Long-term damage to the cerebellum and poor cognitive outcome are not predicted by immediate postoperative MR imaging.

PAROXYSMAL DISORDERS

BENIGN NEONATAL SLEEP MYOCLONUS

The literature on benign neonatal sleep myoclonus (BNSM) was reviewed and synthesized by researchers at University of Bern, Switzerland. The diagnostic criteria were neonatal onset, myoclonic jerks (sudden, brief, jerky involuntary movements) occurring only during drowsiness or sleep, cessation with arousal, and absence of concomitant epileptiform EEG activity. All articles published in English after the original description by Coulter and Allen in 1982 were analyzed. Based on 24 reports that included 164 term-(96%) or near term-born (4%) infants, BNSM occurred in all sleep stages, disappeared after spontaneous or provoked arousal, and was induced by rocking (7 cases) the infant or repetitive sound stimuli (8 infants). Jerks worsened by holding the limbs (5 reports) or by administration of antiepileptic drugs (33 cases). BNSM resolved by age <3 months in 64%, and by age <6 months in 95%; it persisted after 3 months of age in one-third of infants. Incidence is unknown but is estimated between 0.8 and 3.0 cases per 1000 births. A positive family history of BNSM was reported in 3 cases and parasomnias in 9 cases. Mothers had no history of illicit drug use. (Maurer VO, Rizi M, Bianchetti MG, Ramelli GP. Benign neonatal sleep myoclonus: a review of the literature. *Pediatrics* April 2010;125:e919-e924). (Respond: Mario G Bianchetti MD, San Giovanni Hospital, 6500 Bellinzona, Switzerland. E-mail: mario.bianchetti@pediatrician.ch).

COMMENT. An awareness of the characteristics of benign neonatal sleep myoclonus and the clinical diagnosis should differentiate it from epilepsy. Parents may be advised to avoid unnecessary stimulation by rocking and noise and be reassured of an early spontaneous recovery.

Paroxysmal nonepileptic events in children and adolescents vary with age (Kotagal P et al. *Pediatrics* 2002;110(4):e46). 1) *Infant, Toddler, and Preschool Group* (2 mos – 5 years; 26 patients), the most common diagnoses were stereotyped movements, hypnic jerks, parasomnias, and Sandifer syndrome (spasmodic torticollis, hiatus hernia, and gastroesophageal reflux). Concomitant epilepsy was present in 12 patients (46%); 2) *The School-Age Group* (5 – 12 years; 61 patients), Most common diagnoses were conversion disorder (psychogenic seizures), inattention or daydreaming, stereotyped movements, hypnic jerks, and paroxysmal movement disorders. Concomitant epilepsy in 15 patients (25%); 3) *The Adolescent Group* (12 – 18 years) 48 patients of whom 40 (83%) were diagnosed with conversion disorder. Nine (19%) had concomitant epilepsy. Conversion disorder was most common in the adolescent group and in females, whereas males predominated in the school-age group. Concomitant epilepsy with nonepileptic events occurred in all age groups.

BENIGN INFANTILE SEIZURES (FUKUYAMA SYNDROME)

Benign infantile seizures (BIS), first described by Fukuyama (1963) as generalized convulsions were later reported as focal or partial seizures, mainly non-familial, and more recently as familial with autosomal dominance. The two types, familial and non-familial, generalized or focal, are now presented as one syndrome of Fukuyama, Watanabe, and Vigevano. The occurrence of the syndrome in Saudi Arabia is reported by researchers at King Fahad Military Hospital, and King Saud University. Inclusion criteria for BIS were as follows: 1) age of onset 2-24 months; 2) normal development; 3) normal interictal EEG; 4) normal brain imaging; and 5) good response to treatment. Of 116 infants, between 2 and 24 months, with epilepsy, 14 (12%) showed electroclinical features consistent with BIS; 11 patients fulfilled criteria for benign non-familial infantile seizures (BNFIS), and 3 had pedigrees consistent with benign familial infantile seizures (BFIS). All responded to anticonvulsants, and 50% responded within 3 months. (Saardeldin IY, Housawi Y, Al Nemri A, Al Hifzi I. Benign familial and non-familial infantile seizures (Fukuyama-Watanabe-Vigevano syndrome): A study of 14 cases from Saudi Arabia. *Brain Dev* May 2010;32:378-384). (Respond: Dr IY Saardeldin.E-mail: eyssaad@yahoo.co.uk).

COMMENT. First described in Japan, the syndrome of BIS is now reported in other countries, including Saudi Arabia. In the new classification described below, BIS or benign infantile epilepsy is included under the electroclinical syndromes defined by age, but grouped with West syndrome and Dravet syndrome, both having a poor prognosis.

ILAE REVISED CLASSIFICATION AND TERMINOLOGY OF SEIZURES AND EPILEPSIES, 2005-2009

The International League Against Epilepsy (ILAE) Commission on Classification and Terminology has revised concepts, terminology, and approaches for classifying seizures and forms of epilepsy, last updated in 1981 for seizures and in 1989 for epilepsies. Changes in the 1981 *classification for seizures* include the following: