# PEDIATRIC NEUROLOGY BRIEFS
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J. GORDON MILLICHAP, M.D., F.R.C.P., EDITOR

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## DEMYELINATING DISORDERS

### CEREBELLAR MUTISM IN ACUTE DISSEMINATED ENCEPHALOMYELITIS

Researchers at a pediatric multiple sclerosis clinic, the Neurological Institute, Buffalo General Hospital, New York, report six (32%) of 19 patients diagnosed with acute disseminated encephalomyelitis (2005-9) who had protracted speech and language deficits and 3 having cerebellar mutism after follow-up from 6 months to 4 years. One third of the 19 patients had cerebellar symptoms, all under the age of 3 years at presentation. The dentate nucleus was involved in all 6 with cerebellar symptoms, but only 3 had mutism. None showed all of the features of posterior fossa syndrome (cerebellar cognitive affective syndrome), but 2 with most extensive cerebellar involvement had marked emotional lability and behavior disturbance. All 3 patients with mutism continue to exhibit expressive speech, neurocognitive, and behavioral problems. Of 3 children with cerebellar involvement without mutism, 1 has reading and coordination difficulties, and 1 has verbal memory and attention deficits. One patient with neurocognitive deficits at follow-up had MRI evidence of cerebellar atrophy. Quantitative MRI should be considered in patients with acute disseminated encephalomyelitis who have persistent cognitive deficits at follow-up. ((Parrish JB, Weinstock-Guttman B, Yeh EA. Cerebellar mutism in pediatric acute disseminated encephalomyelitis. **Pediatr Neurol** April 2010;42:259-266). (Respond: Dr Parrish, Department of Neurology, Buffalo General Hospital, 100 High Street, Suite D-6, Buffalo, NY 14203, E-mail: jparrish@thejni.org).

COMMENT. Cerebellar involvement in acute disseminated encephalomyelitis is common but speech and language problems are infrequent and cerebellar mutism is rare.

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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 dentatothalamic 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.