MOVEMENT DISORDERS

OUTCOME OF OPSOCLONUS-MYOCLONUS SYNDROME

Long-term neurologic sequelae and predictors for disease outcome were identified in 101 patients diagnosed with opsoclonus-myoclonus syndrome (OMS) over a 53-year period at Royal Hospital for Sick Children, Glasgow; Great Ormond Street Hospital, London; and Guy's and St Thomas Evelina Children's Hospital, London, UK. Median age at disease onset was 18 months (range 3 months to 8.9 years). Neuroblastoma was detected in 21% of patients (40% in those born after 1990). A preceding illness was reported in 56 patients (upper respiratory tract infection, gastroenteritis, and nonspecific), and 8% had been vaccinated within one month of symptom onset. Treatment of OMS consisted of steroids in 87%, none in 12%, and IVIg in 1 case. Median follow-up was 7.3 years (range 3-32 years). Response was good in 35% and moderate in 60%. The course was chronic-relapsing in 61% patients and monophasic in 7%, and acute exacerbations were frequent in 32%. At last review, 60% had residual motor problems, 66% speech abnormalities, 51% learning disability, and 46% behavior problems. One third had normal intellectual outcome and were asymptomatic. A severe initial presentation in 82% patients predicted a chronic course and later learning disability. Cognitive impairment occurred in patients younger at disease onset. A chronic-relapsing course was associated with motor, speech, cognitive, and behavior problems. (Brunklaus A, Pohl K, Zuberi SM, de Sousa C. Outcome and prognostic features in opsoclonus-myoclonus syndrome from infancy to adult life. **Pediatrics** August 2011;128:e388-e394). (Respond: Andreas Brunklaus MD, Neurosciences Unit, Royal Hospital for Sick Children, Dalnair Street, Glasgow G3 8SJ, UK. E-mail: brunklaus@nhs.net).

COMMENT. OMS is a chronic and debilitating illness with frequent long-term motor, speech, and cognitive disabilities, especially in patients of younger age at onset and severe initial presentation. Early diagnosis and treatment with immunomodulating therapy are recommended.

Outcome of OMS in 11 children, 8 having occult neuroblastoma, is reported from Children's Memorial Hospital, Chicago (Hammer MS, Larsen MB, Stack CV. **Pediatr Neurol** 1995;13:21-24). Opsoclonus and ataxia responded to ACTH but recurred when treatment was discontinued. Symptoms were not improved by removal of neuroblastoma. At follow-up, ranged from 12 to 115 months, 8 patients had developmental delay, motor incoordination, and speech and cognitive delay. Development was normal in 2 of 3 patients without neuroblastoma and in only 1 of 8 with neuroblastoma.

A persisting disability at long-term follow-up is reported in 88% of 54 patients with OMS (dancing eye syndrome) reported from the Hospital for Sick Children, Great Ormond Street, London, UK (Pohl KRE, Pritchard J, Wilson J. **Eur J Ped** 1996;155:234-244).

NMDA receptor autoimmune encephalitis presenting with opsoclonus-myoclonus is reported in a 27-year-old woman with a history of episodic migraine. (Smith JH, et al. **Arch Neurol** Aug 2011;68(8):1069-1072). No neoplasm was detected. The encephalopathy responded to plasmapheresis.