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ATAXIAS

"HAPPY PUPPET" SYNDROME

The diagnostic features of 36 cases of Angelman's "happy puppet" syndrome are reported from the Hospital for Sick Children, Great Ormond Street, London. These include ataxia, developmental delay, paroxysmal laughter, seizures, and microcephaly. The facial appearance is characterized by a prominent jaw, wide mouth, and pointed chin. The ataxia of gait is associated with jerky limb movements and hand flapping. Muscle tone is variable, deep tendon reflexes normal, and plantar responses flexor. This series included 3 sibships of 3 affected sisters, 2 affected brothers, and 2 affected sisters, respectively. Males and females were equally affected. The electroencephalogram was abnormal in all cases and showed rhythmic slow waves at 4-6HZ and runs of 2-3 HZ anteriorly. On closure of eyes, spikes and 2-4 HZ slow waves occurred posteriorly. CT showed mild cerebral atrophy in 8 of 23 patients tested. Chromosome deletions of 15q11-13 were detected in 5 patients. (Robb SA et al. The "happy puppet" syndrome of Angelman: review of the clinical features. Arch Dis Child Jan. 1989;64:83-86).

> COMMENT. Prior to this report, approximately 50 cases of Angelman's syndrome had been described in a period of 20 years. First considered to be sporadic it now appears to be genetic in nature. The authors stress the importance of the recognition of the syndrome so that genetic counseling can be offered.

ASTASIA-ABASIA

Twenty-seven children and adolescents diagnosed as having

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conversion reaction manifested as an inability to stand or walk are reported from the Department of Pediatrics, Mayo Clinic, Rochester, MN. There were 9 males and 18 females, ages 8 to 16 years, and duration of the symptoms varied from 1 to 60 weeks. Precipitating events in 17 patients included minor illnesses, school and home related problems, and minor injuries. Onset of symptoms showed seasonal variation with maximal frequency in the winter months and none in the summer and at time of school vacation. The duration of symptoms, estimated at the time of follow-up some 5 to 22 years later, was from a few days to a maximum of 2.5 years. Twenty-two (80%) said they were healthy and the remainder had minor complaints. None had organic illnesses that might have explained the ataxia. Three had continued mental problems and one was in jail for having killed his parents. (Stickler GB, Cheung-Patton A. Astasia-abasia. A conversion reaction. Prognosis. Clin Pediat Jan 1989;28:12-16).

COMMENT. A prompt and firm diagnosis on the basis of positive in addition to exclusion criteria is advocated, and a neurologic examination and appropriate laboratory tests are essential as a reassurance for both patient and treating physician. Dystonia musculorum deformans can be a pitfall, a disorder frequently misdiagnosed as a conversion reaction initially. In a study of 7 children with neurological symptoms diagnosed as hysterical conversion reactions and reported from Duke University Medical Center (Bangash IH et al AJDC Nov 1988;142:1203) all but one had been misdiagnosed as having organic diseases. When the correct diagnosis was made, all recovered and none relapsed after 3 to 11 months follow-up.

CHIARI TYPE I MALFORMATION

A 13-year-old girl with post-traumatic cerebellar ataxia, transient upper extremity weakness, and lower cranial nerve dysfunction was found to have a Type I Chiari malformation on MRI at the Depts of Neurosurgery and Neurology, San Francisco General Hospital, California. She lost consciousness and had a cardiopulmonary arrest after hitting her neck and head on the windshield in an automobile accident. On the 2nd day, her right upper limb was paralyzed; on day 4 she had upbeating nystagmus and was unable to swallow, phonate, or protrude her tongue; and on day 12 right vocal cord paralysis was noted which recovered by day 15. She continued to have improvement of the lower cranial nerve weakness but the ataxia persisted at the time of discharge. It was thought that the initial cardiopulmonary arrest after trauma and the brain stem dysfunction and ataxia were causally related to the Type I Chiari malformation. The continued improvement argued against surgical decompression. (Mampalam TJ et al. Presentation of Type I Chiari malformation after head trauma. Neurosurgery 1988;23:760-762).

COMMENT. This sudden manifestation of symptoms and signs of Chiari I malformation after head and neck trauma is unusual. Previously reported cases include a 3-year-old child who died 48 hours after a mild head injury, a