Unit, Tel Aviv Medical Center, Israel. Ataxia, present since 1 year of age, and irregular, "jerky" eye movements, noted on admission, became worse over a 2 month observation period. An abdominal mass found at 20 months and removed at operation was a ganglioneuroblastoma. Following surgery, steroids for 3 weeks, and chemotherapy 1 year, blood pressure returned to normal immediately and the ataxia and opsoclonus disappeared within 6-7 weeks. At a 2 year follow-up, the neurological and general examinations were normal. (Harel S et al. Cerebellar ataxia and opsoclonus as the initial manifestations of myoclonic encephalopathy associated with neuroblastoma. Child's Nerv Syst 1987;3:245-247).

COMMENT. Opsomyoclonus or "dancing eye syndrome", also known as myoclonic encephalopathy of infancy, is frequently of undetermined etiology. It may follow viral infection and it is sometimes associated with occult malignancies, notably neuroblastoma. Normal urinary catecholamines do not exclude the presence of tumor and repeat evaluations including radiographs of abdomen and chest are indicated. The acute stage of the dancing eye syndrome usually responds best to ACTH followed after a few weeks by prednisone. Steroids may need to be continued for several months.

DEGENERATIVE ATAXIC DISORDERS

Harding AE at the Institute of Neurology, London, author of the Hereditary Ataxias and Related Disorders (Edinburgh, Churchill Livingstone, 1984) reviews the classification, causes, clinical characteristics and treatment of degenerative ataxias. A combination of genetic and environmental factors is the most common origin for this complex group of over 50 distinct diseases, subdivided according to clinical and genetic features. Metabolic defects such as arylsulfatase-A in metachromatic leukodystrophy are recognizable but untreatable but some deficiency diseases (e.g. Vitamin E) are amenable to treatment with supplements. The cause of Friedreich's ataxia, an autosomal recessive disorder, is unknown and reported deficiencies of pyruvate dehydrogenase and mitochondrial malic enzymes have not been confirmed. Similarly, in olivoponto-cerebellar atrophy, a late onset ataxia, recent studies have not confirmed an earlier report of reduced leucocyte glutamate dehydrogenase activity. Most attempts at treatment of degenerative ataxias have been disappointing but promising results using thyrotropin releasing hormone have been reported from Japan. (Harding A. Degenerative ataxic disorders: still perplexing. BMJ 1987; 295:1223-4).

COMMENT. Degenerative ataxias resembling Friedreich's ataxia that may be amenable to treatment include Vitamin E, B12, folate and biotin deficiencies and Refsum's disease, responsive to a diet low in phytol and phytanic acid.