

# PEDIATRIC NEUROLOGY BRIEFS

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## MOVEMENT DISORDERS

### PATIENT PERCEPTION OF TICS

The subjective perception of abnormal movements was determined in 170 patients with various hyperkinesias at the Movement Disorders Clinic, Toronto Western Hospital, Toronto, Ontario. 102 of 110 patients with nontic disorders thought that the abnormal movements were entirely involuntary whereas 41 of 60 tic disorder patients stated that all of their motor and phonic tics were intentional or voluntary. A "voluntary response" could be used to predict the correct diagnostic category (tic versus nontic) in 8 of 9 patients for whom the initial diagnosis was incorrect. A large proportion of the motor and phonic symptoms experienced by tic patients are irresistably, but purposefully executed and resemble compulsions rather than involuntary hyperkinesias. (Lang A. Patient perception of tics and other movement disorders. Neurology Feb 1991; 41:223-228)

**COMMENT:** This recategorization of tics as intentional and purposeful actions is similar to that of compulsions which may be linked genetically.

### DYSTONIA AND PARTIAL CYTOCHROME B DEFICIENCY:

An 18 year old female with idiopathic torsion dystonia associated with mitochondrial encephalomyopathy is reported from the Departments of Neurology, Pediatrics and Biochemistry, Wayne State University School of Medicine, Detroit, Michigan. At the age of 2½ she developed weakness and disuse of the right leg with a febrile illness. Within one week the weakness extended to the right upper limb. During the next six months she developed progressive dysarthria and dystonia of

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the right leg and trunk. The dystonia progressively involved all extremities over the next four years. At the age of 7 years CT revealed bilateral putaminal hypodensities. Mitochondrial encephalomyopathy was suspected because of persistent lactic acidemia and myopathy. Studies of oxidative metabolism on skeletal muscle mitochondria revealed partial cytochrome b deficiency indicating a defect in cytochrome b-C1 complex (Nigro MA et al. Partial cytochrome b deficiency in generalized dystonia. Pediatr Neurol Nov/Dec 1990; 6:407-410).

COMMENT: Children with Leigh encephalopathy may develop dystonia and putaminal degeneration. H magnetic resonance spectroscopy was used to demonstrate regional variation in brain lactate in an 11 month old infant with Leigh syndrome at the University of Pennsylvania, Philadelphia. (Detre JA et al. Regional variation in brain lactate in Leigh syndrome by localized H magnetic resonance spectroscopy. Ann of Neurol Feb 1991; 29:218-221). Elevations in brain lactate were observed throughout the brain, but were most pronounced in the basal ganglia and brain stem regions with an abnormal MRI appearance. The increase in lactate was consistent with a deficiency in oxidative substrate utilization. The measurement of brain lactate is significant in patients where blood lactate levels may not always be elevated.

Symptomatic dystonias secondary to birth asphyxia, birth trauma, Wilson's disease, other degenerative disorders, encephalitis, neuroleptic drugs and mitochondrial disorders are distinguished from idiopathic or primary torsion dystonias. A genetic study of idiopathic focal dystonias has shown that 25% of index patients had relatives with dystonia. The results of segregation analysis suggested an autosomal dominant gene or genes with reduced penetrance as a common cause for focal dystonia and a single autosomal dominant gene mutation responsible for inherited dystonia in the majority of patients, irrespective of distribution or severity. Severity is largely determined by age of onset, with generalized idiopathic dystonia developing before the age of 20 and focal dystonias developing in adult life. (Waddy HM et al. Ann Neurol March 1991; 29:320-324).

## LEARNING DISORDERS

### STIMULANT MEDICATION FOR ADHD: THE CHILD'S PERSPECTIVE

The knowledge and attitudes of children receiving stimulant medication for attention deficit hyperactivity disorder were investigated at the Division of Ambulatory Pediatrics, Children's Hospital, Boston, MA. Of 45 respondents and parents who completed separate questionnaires concerning how they felt about receiving stimulant medication, 89% of the children felt that the medication was helpful