April 1992; 42:784-788). (Reprints: Dr. C.G. Goetz, Department of Neurological Sciences, Rush-Presbyterian-St. Luke's Medical Center, 1725 West Harrison St., Chicago, IL 60612.)

COMMENT. In a study of TS in monozygotic twins, Hyde et al. at the National Institute of Mental Health, Washington, D.C. observed a significant effect of birth weight on the phenotypic expression of TS. Tic scores were greater in the lower birth weight twin as compared with the heavier group. Postnatal factors were not identified and did not play a major role in the subsequent expression of TS (Neurology March 1992; 42:652-658). Global neuropsychological performance was more impaired in the more severely affected twin particularly in tests of attention and visuospatial perception (Hyde TM et al. Neurology April 1992; 42(Suppl 3):396). Hypnosis was of benefit in reducing involuntary tics in 6 of 7 unmedicated children age 6 to 15 years with TS (Hollander H et al. Neurology April 1992; 42(Suppl 3):239). Deprenyl, an MAO-B inhibitor, was a safe and effective treatment of attention deficit disorder with hyperactivity in 80% of 20 children with TS. None of the patients noted exacerbation of their tics or other adverse side effects at doses up to 15 mg/day (Jankovic J. Neurology April 1992; 42(Suppl 3):238).

MUSCLE DISORDERS

MITOCHONDRIAL MYOPATHY AND HYPOTONIA

Three children with hypotonia, cardiac impairment, and defects of the mitochondrial respiratory chain complexes, but no ragged red fibers, are reported from the Hopital de la Timone, Chemin de l'Armee, d'Afrique, Marseille, France. Case 1 shared clinical and metabolic features with fatal infantile myopathy associated with cytochrome *c* oxidase deficiency as described by DiMauro: neonatal hypotonia and weakness, respiratory failure, and severe lactic acidosis. Post-mortem studies at age 7 weeks showed complex IV reduced in the liver but not in the heart and quantitative analysis of mtDNA showed depletion in muscle. Case 2 showed intractable cardiomyopathy, cyclic neutropenia, and 3-methylglutaconic aciduria. The boy died suddenly at age 27 months. Case 3 presented at age 16 months as an acute hypokinetic hypertrophic cardiomyopathy with transient hypotonia and mild lactic acidosis. After the acute episode the boy gradually improved and the neurological and cardiac examinations were normal at the age of 3 years. All cases showed lipid storage myopathy and decreased cytochrome *c* oxidase. Biochemical studies confirmed the cytochrome *c* oxidase deficiency in muscle in all cases (Figarella-Branger D et al. Defects of the mitochondrial respiratory chain complexes in three pediatric cases with hypotonia and cardiac involvement. (J Neurol Sci March 1992; 108:105-113.) (Correspondence: Dr. D. Figarella-Branger, Laboratoire d'Anatomie Pathologique, Hopital de la Timone, Chemin de l'Armee d'Afrique, F-13005 Marseille, France.)

COMMENT. Histochemical staining of cytochrome *c* oxidase may be used for the diagnosis of mitochondrial myopathy when ragged red fibers are lacking. Schon EA et al. and Munnich A et al. review the mitochondrial myopathies and the clinical aspects of mitrochondrial disorders in the current issue of International Pediatrics 1992; 7:23-33. The diagnosis of mitochondrial disorder should be considered with (1) an unexplained association of symptoms; (2) an early onset and a rapidly progressive course; and (3) involvement of unrelated organs which share no common embryologic origin and no common biological functions. Determination of lactate/pyruvate and ketone body molar ratios in plasma may help to select patients at risk for further investigation. In a family with infantile mitrochondrial myopathy and cardiomyopathy reported from Rome, Italy, no correlation was found in the muscle between biochemistry and severity of the clinical intrafamilial phenotype (Bertini E et al. Neurology April 1992; 42(Suppl 3):267).

DYSTROPHIN IN FETAL MUSCLE

Dystrophin, the product of the Duchenne muscular dystrophy gene, was studied in human fetal skeletal muscle from 9 to 26 weeks of gestation at the Jerry Lewis Muscle Research Centre, Hammersmith Hospital, London. Dystrophin was localized to the sarcolemma of myotubes in fetal muscle from at least 9 weeks of gestation. Dystrophin immunostaining increased and became more uniform with age and sarcolemmal staining of myotubes was intense by 26 weeks of gestation. Western blot analysis showed a lower relative molecular mass protein in fetal tissue compared to adult tissue. This persisted until 26 weeks of gestation but switched to the adult form by 9 months of age. The data demonstrated several specific isoforms of dystrophin present in developing skeletal muscle (Clerk A et al. Characterisation of dystrophin during development of human skeletal muscle. Development Feb 1992; 114:395-402).

COMMENT. Tome FMS et al. in Paris, France and Boston, MA report on the distribution and localization of dystrophin and dystrophin-related protein during skeletal muscle development and changes in Duchenne muscular dystrophy fetuses. Dystrophin of the same size as in adults was present in normal fetuses as soon as 8 weeks gestation in this study. It was absent in Duchenne muscular dystrophy fetuses. The dystrophin-related protein was quantitatively and qualitatively comparable in normal and Duchenne fetuses. The authors concluded that the prenatal diagnosis of Duchenne muscular dystrophy can be confirmed by immunocytochemical or biochemical analysis of muscular dystrophin in aborted fetuses. Dystrophin-related protein has a particular role in synaptic function which is not disturbed in Duchenne muscular dystrophy (Neurology April 1992; 42(Suppl 3):227-228).