born with severe muscular hypotonia (92%), apneic spells (92%), cardiomyopathy (76%), lactic acidosis (92%), hyperammonemia (86%), and 3-methylglutaconic aciduria (100%). Ten died within the first 6 weeks after birth. Surviving infants had persistent muscular hypotonia and psychomotor delay, with microcephaly in 13/22. Boys had hypospadia in 54% and cryptorchidism in 67%. (Honzik T, Tesarova M, Mayr JA et al. Mitochondrial encephalocardio-myopathy with early neonatal onset due to TMEM70 mutation. **Arch Dis Child** 2010;95:296-301). (Respond: Professor J Zeman, Charles University of Prague, Czech Republic. E-mail: jzem@if1.cuni.cz).

COMMENT. ATP synthase deficiency due to TMEM70 mutation is a novel mitochondrial disease of neonatal onset with muscle hypotonia, hypertrophic cardiomyopathy, lactic acidosis, hypospadius, hyperammonemia, and 3-methylglutaconic aciduria. Progressive CNS impairment affects most patients who survive the neonatal period, but the severity of the phenotype may vary. Molecular genetic diagnosis is available without need for muscle biopsy. TMEM70 deficiency should be considered in critically ill hypotonic neonates.

ATTENTION DEFICIT DISORDERS

SACCADE EYE MOVEMENTS AS MEASURE OF FRONTOSTRIATAL DYSFUNCTION IN ADHD

Saccade latency and accuracy were tested in 50 normal subjects (6-35 years), 19 ADHD patients (6-11 years), and 4 patients with frontal lesions (13-15 years) in an investigation of reflexive/voluntary control of saccades in ADHD, at University of Yamanashi, Japan. Subjects were seated in a dental chair with a chin rest in the dark, facing a display 100 cm away. A central fixation point (FP) appeared in the straight-ahead position, and visual targets were presented at 20 degrees to right or left of FP. Saccade tasks involved visually-guided (VGST), memory-guided (MGST), and antisaccade tasks. In normal controls, saccade latency and accuracy error rates were significantly correlated with age and maturity. The ADHD group showed significantly higher percentage of anticipatory errors and direction errors. Saccade eye movements do not fully mature until adolescence, and ADHD patients show dysfunction in "response inhibition", which is modulated by the frontal lobe. (Goto Y, Hatakeyama K, Kitama T et al. Saccade eye movements as a quantitative measure of frontostriatal network in children with ADHD. **Brain Dev** May 2010;32:347-355). (Respond: Dr Masao Aihara: E-mail: maihara@yamanashi.ac.jp).

COMMENT. The core symptoms of ADHD result from failure to inhibit or delay appropriate behavioral responses to stimuli. Evidence of faulty inhibition in ADHD is demonstrated by neuropsychological tasks (Go/NoGo, delayed response task, Stroop Color Word Interference Test), and from neuroimaging studies (MRI, SPECT, PET, and functional MRI). Abnormal saccade eye movements provide further evidence of frontostriatal dysfunction in children with ADHD.