

MOVEMENT DISORDERS

BRAIN GROWTH IN CHILDREN AT RISK FOR HUNTINGTON DISEASE

Researchers at the University of Iowa and Washington University, St Louis, MO, studied the effect of the mutant Huntington gene (mHTT) on measures of growth in children at risk for Huntington disease (HD). Measurements of growth (height, weight, body mass index [BMI], and head circumference) in 20 at risk gene-expanded children, aged 7-18 years, with no symptoms were compared with measurements in 14 gene-nonexpanded children and 138 age-matched healthy controls. Children with a CAG repeat length \geq 39 were designated as gene-expanded. At risk gene-expanded children had significantly lower measures of head circumference, weight, and BMI. Head circumference was abnormally low even after correcting for height, suggesting a specific defect in brain growth, rather than a global growth abnormality. mHTT may play a role in atypical somatic, and particularly, brain development. (Lee JK, Mathews K, Schlaggar B, et al. Measures of growth in children at risk for Huntington disease. **Neurology** 2012 Aug 14;79(7):668-74). (Response: Jessica Lee. E-mail: jessica-k-lee@uiowa.edu).

COMMENT. Children tested as HD gene expanded were an estimated >3 decades from onset of the disease. Constant caloric burn due to chorea is a possible cause of weight loss in HD patients but not in preHD children. A primary abnormality in mitochondrial function and metabolic rate is more likely. (Damiano M, et al. Mitochondria in Huntington's disease. **Biochim Biophys Acta** 2010 Jan;1802(1):52-61 | Also cited by Lee JK).

LONG-TERM CLINICAL COURSE OF TOURETTE SYNDROME

Researchers at Catania University, and University Tor Vergata, Rome, Italy studied the course of Tourette syndrome (TS) after 10 years follow-up in 100 children. Of the "pure TS" group (n=38), 58% were unchanged, whereas 42% changed to TS+OCD phenotype. Of the "TS+ADHD" group (n=48), 62% changed to pure TS, 35% to TS+OCD, and 2% to TS+ADHD+OCD. Medication was required in 65%. Patients with comorbid condition at onset had a more severe prognosis and lower quality of life scores. (Rizzo R, Gulisano M, Cali PV, Curatolo P. **Brain Dev** 2012 Sep;34(8):667-73). (Respond: Dr Rizzo. E-mail: rerizzo@unict.it).

COMMENT. Patients with pure TS at onset have a favorable prognosis, whereas patients with TS and comorbid disorders, ADHD and OCD, are at risk of impaired quality of life. TS presents with a variety of clinical phenotypes that may change over time. Appropriate treatment of ADHD in children with TS may prevent behavioral problems in adulthood. Comorbidity with ADHD, occurring in 50% of TS patients, causes more disability than motor tics alone. (Spencer T et al. **Am Acad Child Adolesc Psychiatry** 1995 Sep;34(9):1133-9). Children with TS and ADHD have a 32% risk of learning disabilities. (Schuerholz LJ et al. **Neurology** 1996 Apr;46(4):958-65).