comparison with norms included sustained attention, executive functioning, visual-spatial function and memory. Severe hydrocephalus before surgery was associated with lower scores on visual-spatial skills and complex figure drawing. Behavioral disturbances were observed in 15 children (65%), including overanxious disorder, Asperger syndrome, ADHD, and posttraumatic stress disorder. Flattened affect, disinhibited or clingy behavior, loss of speech spontaneity, and verbal hyperspontaneity were also seen. The cerebellar cognitive affective syndrome (CCAS) following cerebellar resection in children is a loose complex with variable composition. The high percentage requiring special education reflects the severity and persistence of the cognitive deficits. (Aarsen FK, Van Dongen HR, Paquier PF et al. Long-term sequelae in children after cerebellar astrocytoma surgery. **Neurology** April (2 of 2) 2004;62:1311-1316). (Reprints: FK Aarsen MA, Pediatric Neuropsychologist, Department of Pediatric Neurology, Erasmus MC/Sophia Children's Hospital, PO Box 2060, 3000 CB Rotterdam, The Netherlands).

COMMENT. The authors stress the need to inform parents and teachers about behavioral and cognitive sequelae following cerebellar surgery, so that timely social and educational intervention is made available. The cerebellum is now known to be involved in cognitive and emotional responses. A posterior fossa syndrome (PFS) is a neurobehavioral syndrome consisting of transient short-term deficits arising 1 to 6 days postoperatively and gradually resolving over months. Mutism and subsequent dysarthria (MSD) is the most common example of the PFS. The cerebellar cognitive affective syndrome (CCAS) as seen in adults consists of 4 major symptoms: disturbances of executive function, impaired spatial cognition, linguistic difficulties, and personality changes. In children following cerebellar tumor resection, CCAS is atypical but apparently more prevalent than previously reported.

BEHAVIOR DISORDERS

METABOLIC CAUSE OF HYPERKINETIC BEHAVIOR

An 8-year-old female child with congenital ornithine transcarbamylase deficiency (OTCD) who presented with a hyperkinetic behavior disorder at 3 years is reported from the Medical School Hanover, Germany. Methylphenidate therapy was used for 1 year without benefit. At age 6, she developed increasing somnolence, hypotonia and dyskinesia during high fever. The diagnosis of OTCD was based on hyperammonemia, increased glutamine, low normal citrulline and decreased arginine in the blood, and elevated orotic acid and uracil in urine. Mutation analysis of the OTC gene revealed heterozygosity for 274C; R92X. Despite therapy with a low protein diet, glucose, arginine, benzoate and phenylbutyrate, she became comatose for a week before slowly waking with flapping tremor and seizures. MRI showed cerebellar atrophy and white matter lesions. One year after the metabolic stroke, metabolic control and clinical improvement were observed. (Lucke T, Illsinger S, Hartmann H et al. Ornithine transcarbamylase deficiency in a girl with hyperkinetic behaviour. **J Pediatr Neurol** 2004;2(2):97-100). (Respond: Thomas Lucke MD, Department of Pediatrics, Medical School Hanover, Carl-Neuberg-Str 1, D-30623 Hanover, Germany).

COMMENT. In a young child with hyperkinetic behavior unresponsive to methylphenidate, a metabolic cause should be considered in the differential diagnosis.