Diagnosis was confirmed by measles antibody in CSF and by electroencephalography. The mean age of patients was 13.4 years (range, 4-21 years) (mean of 15.7 years in 9 who had received measles vaccination and 12.4 years in the unvaccinated group). Loss of vision (cortical) in 7 cases, seizures in 6, and behavioral change in 7 were the presenting symptoms in 40% of patients. More classic symptoms of myoclonus and cognitive decline, present at first examination in 27 and 26 patients, respectively, were delayed for a mean of 8 months after onset of symptoms. MRI or CT in 21 was normal in one-third and showed abnormalities in 13, with predominantly white matter lesions in the parietal-occipital areas. Compared to a similar study of 39 patients at the same institution in 1974, the present series shows a later mean age of onset (13.4 cf 11.2 years), a similar male preponderance (24:8 cf 36:3), myoclonus (84% cf 95%). cognitive decline (81% cf 97%), less seizures (19% cf 33%), hemiparesis (16% cf 5%), chorioretinitis (13% cf 2%), and more prevalent visual loss (22% cf 0). (Khadilkar SV, Patil SG, Kulkarni KS. A study of SSPE: early clinical features. **J Pediatr Neurol** 2004;2(2):73-77). (Respond: Dr Satish V Khadilkar, Room 110, 1st Floor, MRC Building, Bombay Hospital, Mumbai, India).

COMMENT. The changing character and resurgence of SSPE in the US was referred to by Dyken PR in 1989 and is readdressed in an editorial (Dyken PR. Clinical expressivity in resurging SSPE: changing age of onset and new early symptoms. **J Pediatr Neurology** 2004;2(2):53-56). The above reports emphasize that measles and SSPE are not disappearing entities, even in "developed" countries, and that complacency among pediatric neurologists should be avoided. In addition to the typical subacute progressive form (SPF) that represents about 75% of reported cases, atypical cases are not stereotyped and consist of an acute progressive form (APF), a chronic progressive form (CPF), and two remitting forms, one chronic and one subacute. In the current report from India, 3 of 9 patients (9% of the total) who were vaccinated may have had an APF, and 6 with early onset loss of vision could be classified as chronic atypical (19% of total). Visual loss associated with posterior cerebral demyelination demonstrated in this series was not reported in earlier publications. The role of immunization in the changing character and age at onset of SSPE requires further study.

## BRAIN NEOPLASMS

### LONG-TERM SEQUELAE AFTER CEREBELLAR ASTROCYTOMA SURGERY

The long-term effects on neurologic, neuropsychological, and behavioral functioning in a consecutive series of 23 children treated surgically for cerebellar pilocytic astrocytoma without additional radio- and chemotherapy are determined in a study at Sophia Children's Hospital, Rotterdam, The Netherlands, and other medical centers. Follow-up ranged from 1 year to 8 years after tumor resection. Age at testing ranged from 6 to 22 years (mean, 12 years). Shunting of a preoperative hydrocephalus was required in 11 children (48%). Neurologic status at time of psychological assessment was normal to mildly impaired; 7 had mild ataxia and 2 a mild intention tremor. Orofacial apraxia occurred in 1 and upper limb neglect in 2. Dysarthria was present in 5 (22%), and language problems in 7 (30%). Two had suffered mutism and MSD syndrome immediately postoperative, with persistence of dysarthria. Neuropsychological tests showing significantly weaker performances in

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.