early indicator. EDS is mistaken for a ‘problem behavior’ on entry to school, when napping is not permitted. Narcolepsy presents in response to school based concerns about EDS and inattentiveness in 78% of childhood cases over 5 years (Guilleminault, 1998). Hypersomnolence may also present as hyperactivity and irritability (Wise, 1998). Like EDS, cataplexy is rarely classic in presentation. Abnormal clumsy behavior, falls or bumps, dropping and breaking of objects, ‘drop attacks’ occur, but rarely in association with laughter or excitement. In one report of 77 childhood cases, cataplexy developed on average within 1 year after EDS symptoms, and the time lag is considerably shorter than in adults. The reporting of sleep paralysis and hallucinations is largely dependent on the age of the child. Estimates suggest that in children with narcolepsy at a mean age of 9 years, 97% have experienced EDS, 80% cataplexy, 39% hallucinations and 29% sleep paralysis, 13% having the full tetrad of symptoms (Challamel, 1994), a finding similar to that in adults. Differential diagnoses include: vaso-vagal or cardiogenic syncope, nocturnal insomnia with obstructive sleep apnea, Pickwickian syndrome, Chiari type 1, hypothalamic dysfunction or tumor, Prader-Willi syndrome, disordered circadian rhythm, atonic seizures, Coffin-Lowry syndrome, pontine tumor, and Niemann Pick disease (diagnosed in 12 of 97 cases of narcolepsy (Challamel, 1994). Genetic markers HLA-DR2 and DQB1 have been associated with narcolepsy but genetic penetrance is low. Recent work has pointed to a degeneration of the neuropeptide orexin in the hypothalamus as the cause. A definitive diagnosis in adults is made with attacks of EDS, cataplexy, and/or sleep onset REM (SOREM) periods measured using polysomnography and the Multiple Sleep Latency Test (MSLT). A mean sleep latency of <5 minutes and/or 2 or more sleep onset REM periods is diagnostic. The MSLT is unreliable in children below 8 years, although polysomnography may help in exclusion of nocturnal sleep disorders. In treatment, stimulants are only partially effective, tricyclics (clomipramine) have been advocated for cataplexy, and a daytime nap schedule is effective in preschoolers. Psychosocial issues are common including emotional lability, aggressive behaviors, social isolation, bullying, academic underachievement and depression. In a cohort of 40 children aged 7 and older, 90% described a sense of shame and 20% had reactive depression (Guilleminault, 1998). School transition is a critical period of adjustment, requiring psychosocial intervention strategies. (Hood BM, Harbord MG. Narcolepsy: diagnosis and management in early childhood. J Pediatr Neurol 2004;2(2):65-71). (Respond: Dr Bernadette Hood, Department of Psychology, Victoria University, PO Box 14428 MCMC 8001, Melbourne, Australia).

COMMENT. Narcolepsy in young children is usually atypical and frequently misdiagnosed. Diagnosis is based primarily on clinical symptoms, and polysomnography is usually inconclusive. Medications are often less effective than in adults, and psychosocial issues on entering school need wider recognition and attention.

INFECTIONOUS DISEASES

CHANGING CLINICAL FEATURES OF SUBACUTE SCLEROSING PANENCEPHALITIS (SSPE)

The early clinical features, age at onset, and relation to immunization of 32 patients (24 males, 8 females) with SSPE are reported from Grant Medical College, Mumbia, India.
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 resurring 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