56 cases (37 adult, 19 pediatric) reported to the British Neurological Surveillance Unit (BNSU) and surveyed at the Department of Psychological Medicine, Institute of Psychiatry, London, UK. Male to female ratio was 2:1 for adult cases and 3:2 in children. Approximately two-thirds (57%) had epilepsy, half the adults were intellectually impaired, and 35% had a psychiatric disorder. Macrocephaly may have accounted for an unusually high (35%) incidence of obstetric complications. The obstetric history was not correlated with the complication of epilepsy, low IQ, or psychiatric disorder. (Taylor M, David AS. Agenesis of the corpus callosum: a United Kingdom series of 56 cases. J Neurol Neurosurg Psychiatry Jan 1998;64:131-134). (Respond: Professor AS David, Department of Psychological Medicine, Institute of Psychiatry, De Crespigny Park, Denmark Hill, London SE5 8AF, UK).

COMMENT. In addition to epilepsy, cognitive impairments and mental retardation, agenesis of the corpus callosum may be complicated by neuropsychiatric disorders, including attention deficit hyperactivity disorder and psychoses. Corpus callosal agenesis is a manifestation of Aicardi and other developmental syndromes. It may also be associated with schizencephaly.

SCHIZENCEPHALY AND CYTOMEGALOVIRUS INFECTION

Two cases of schizencephaly associated with cytomegalovirus (CMV) infection are reported from the University La Sapienza, Rome, and Dibit Scientific Institute, Milan, Italy. In patient 1, MRI showed a left schizencephaly with a polymicrogyric cortex lining the clefts; CMV DNA was demonstrated in the serum on the second day of life. Patient 2 was referred at 4 years of age, and MRI showed a large, open left schizencephaly, surrounded by polymicrogyric cortex, and cerebellar hypoplasia. CMV DNA was detected in the serum, the early history revealed intrauterine growth retardation, microcephaly, petechiae, cataract, hepatospenomegaly,anemia, and periventricular calcifications were present on CT at 3 months of age. Tests for a genetic origin of the schizencephaly failed to demonstrate the characteristic mutation in the homeobox gene EMX2. (Iannetti P, Nigro G, Spalice A, Faiella A, Boncinelli E. Cytomegalovirus infection and schizencephaly: case reports. Ann Neurol Jan 1998;43:123-127). (Respond: Dr Paola Iannetti, Child Neurology Division, Pediatric Department, University La Sapienza, Viale Regina Elena 324, 00161 Rome, Italy).

COMMENT. Prenatal cytomegalovirus infection may be a factor in the pathogenesis of some cases of schizencephaly and other neuronal migration disorders. Yakovlev and Wadsworth who coined the term schizencephaly (1946) timed the formation of the clefts in the 2nd month of fetal development.

PAROXYSMAL DISORDERS

NARCOLEPSY AND CATAPLEXY

The clinical manifestations and treatment of narcolepsy diagnosed in 51 prepubertal children (29 boys) are reported from the Stanford Sleep Disorder Clinic, Stanford University Medical Center, CA. The mean age was 8 years (range, 2 to 11). Diagnosis was based on daytime sleepiness as the initial symptom in 39, 10 presented with cataplexy, and 2 had hallucinations and sleep paralysis. In 5 patients who presented at or before 5 years of age, epilepsy was the first diagnosis considered; 3 reported abrupt unexplained falls, clumsiness, day dreaming, night terrors, and intermittent irritability and aggressiveness. The history of daily naps was obtained only after visits to 3 different specialists. Of 46 children older than 5