Deficit Hyperactivity and Learning Disorders. Chicago, PNB Publ, 1998. Multiple biological causes for autism and autistic-like disorders have been described. The EEG may be abnormal in 50% of cases and neuroimaging may show structural abnormalities in the brain in 25%.


Failure of Secretin treatment for autism is reported in 56 patients (49 boys, 7 girls; mean age 6 years) treated at Lake Forest Hospital, Illinois (Chez MG, Buchanan CP, Bagan BT et al. Autism and Dev Disorders April 2000;30:87-94).

DEVELOPMENTAL DISORDERS

FAMILIAL PERISYLVIAN POLYMICROGYRIA

The clinical presentation and possible mode of inheritance of familial perisylvian polymicrogyria (FPP) are described in twelve affected kindreds presenting at 10 medical centers. Among 42 patients, clinical and radiological findings were variable in families and within members of the same family, except for abnormal tongue movements and/or dysarthria correlating with bilateral MRI findings. The main clinical features, pseudobulbar palsy, cognitive deficits, epilepsy, and perisylvian abnormalities, were not present uniformly, and varied in severity. The syndrome showed an X-linked transmission, except for 2 families with autosomal dominant inheritance and decreased penetrance. (Guerreiro MM, Andermann E, Guerrini R et al. Familial perisylvian polymicrogyria: a new familial syndrome of cortical maldevelopment. Ann Neurol July 2000;48:39-48). (Respond: Dr E Andermann, Montreal Neurological Institute, 3801 University Street, Montreal, Quebec, Canada H3A 2B4).

COMMENT. A new X-linked syndrome, named familial perisylvian polymicrogyria, is characterized by pseudobulbar palsy, cognitive deficits, epilepsy, and cortical maldevelopment. The syndrome is genetically heterogeneous, and the clinical presentation is variable.

ETIOLOGY OF AGENESIS OF CORPUS CALLOSUM

Agenesis of the corpus callosum was found in 7 of 135 children (aged 3 months to 15 years) with structural cerebral defects on MRI. All 7 showed dysmorphic features, psychomotor retardation, and neurologic abnormalities, and 6 had epilepsy (infantile spasms in 3). Causal factors in 4 children were partial trisomy of chromosome 13, partial duplication on the long arm of chromosome 10, Aicardi's syndrome, and intracranial bleeding in the fetus due to injury. Dandy-Walker malformation was also present in one other patient. (Marszal E, Jamroz E, Pilch J et al. Agenesis of corpus callosum: clinical description and etiology. J Child Neurol June 2000;15:401-405). (Respond: Prof Elzbieta Marszal, Pediatric Neurology Clinic, Silesian School of Medicine, ul Medykow 16, 40-752 Katowice, Poland).

COMMENT. Agenesis of the corpus callosum occurs as part of chromosomal syndromes, trisomy 8, 13, 18, or 21, and with X-linked syndromes, especially Aicardi's syndrome. It may be associated with other brain malformations, and it sometimes accompanies various inborn errors of metabolism. Exogenous factors such as intracranial bleeding and infection may also be involved in etiology.