

LANGUAGE DISORDER AND POLYMICROGYRIA

The co-occurrence of developmental language disorder and reading impairment in members of three families with perisylvian polymicrogyria is reported from the State University of Campinas, and University of Sao Paulo, Brazil. The severity of language impairment correlated with the extent of the polymicrogyria, patients with the worst language deficit having diffuse bilateral perisylvian polymicrogyria while patients with mild impairment showing subtle MRI anomalies. (Oliveira EPM, Hage SRV, Guimaraes CA et al. *Characterization of language and reading skills in familial polymicrogyria. Brain Dev* April 2008;30:254-260). (Respond: Dr MM Guerreiro; E-mail: nmg@fcm.unicamp.br).

COMMENT. Polymicrogyria is a cerebral developmental anomaly, characteristically perisylvian in location, giving the cortical surface a pebbled, 'chestnut kernel,' or 'Moroccan leather' appearance. Genetic or acquired causes are described. A causative diagnosis was established in 20 of 48 cases recently reported (de Wit MCY et al. *Arch Neurol* March 2008;65:358-366; *Ped Neur Briefs* March 2008;22:24). A genetic cause was suspected in 6 patients with multiple congenital abnormalities and in 4 with consanguineous parents or multiple affected family members. A gestational insult was the probable cause in 7 patients. Polymicrogyria can be localized or diffuse, unilateral or bilateral. The cortex is thickened, without recognizable layers or with 4 layers in place of the usual 6. The brain stem may be hypoplastic, especially involving the pyramidal tracts. In severe cases, the child has spastic diplegia or hemiplegia, mental retardation, and seizures. The MRI has permitted the diagnosis and recognition of milder forms of polymicrogyria, some associated with language and reading disorders, as described in the above study.

NONAUTISTIC MOTOR STEREOTYPIES

Clinical features and long-term outcomes of 100 children (62 boys and 35 girls) with motor stereotypies were evaluated by review of records and telephone interviews at Johns Hopkins Hospital, Baltimore, MD. Mean age was 8.3 +/- 4.5 years. Age at onset was < 24 months in 81%. All children were in a regular classroom and were at least grade C in achievement. Six had a history of early language delay. Repetitive, rhythmic, involuntary movements consisted of finger wiggling and/or flapping of hands or arms; 20% also exhibited facial grimacing, and 8% had head nodding movements. Movements occurred once a day or more in 90% and lasted less than a minute in 62%. Triggers included excitement/happiness in 80%, anxiety and stress in 26%, and fatigue in 21%. Stereotypies ceased during sleep and when cued by calling his or her name. Family history was positive for most motor stereotypies in 17% first-degree relatives, but negative in patients with head nodding. Associated conditions included ADHD in 30%, tics in 18%, and OCD in 10%. Various medications prescribed in 20 patients, including clonidine, risperidone, and oxcarbazepine, were ineffective, and behavior modification in 14 resulted in modest improvements in 5 patients. Follow-up ranged from 2 months to 26 years, with a median of 6 years. Movements were persistent in 94 children, continuing for >10 years in 22%, and 6-10 years in 44%. Prognosis was better in children with head nodding than in those with hand/arm movements; head nodding resolved in one third, compared to only 3% with hand movements (P=0.001). (Harris KM, Mahone EM, Singer HS. *Nonautistic motor stereotypies:*