CONGENITAL CEREBRAL MALFORMATIONS

CORPUS CALLOSUM SIZE AND VERBAL INTELLIGENCE

Quantitative MRI measurements of the sagittal surface area of the corpus callosum (CC) in 72 preterm individuals tested at adolescence (14-15 years) were compared with verbal skills on neuropsychological tests in a study at the Institute of Psychiatry, De Crespigny Park, London, UK. Total CC area in very preterm adolescents was 7.5% smaller than in controls (p=0.015). In preterm individuals who had a periventricular hemorrhage in the neonatal period, the decrease in CC area was greater. Verbal IQ and verbal fluency scores were lower in male adolescents with reduced total mid-sagittal CC size and mid-posterior surface area. (Nosarti C, Rushe TM, Woodruff PWR, et al. Corpus callosum size and very preterm birth: relationship to psychological outcome. **Brain** September 2004;127:2080-2089). (Respond: Chiara Nosarti PhD, Division of Psychological Medicine, Section of General Psychiatry, PO Box 63, Institute of Psychiatry, De Crespigny Park, Denmark Hill, London SE5 8AF, UK).

COMMENT. The development of the corpus callosum (CC), especially the posterior quarter adjacent to the periventricular region, is adversely affected by very preterm birth. The decreased size of the CC is correlated with lower verbal IQ and verbal fluency scores in preterm boys at adolescence.
A decrease in size of the splenium of the corpus callosum is reported in ADHD children compared to controls (Semrud-Clikeman et al. 1994; see **Ped Neur Briefs** July 1994).

CAUSES, TYPES, AND OUTCOME OF HOLOPROSENCEPHALY

Recent advances in genetics and neuroimaging of children with holoprosencephaly (HPE) are reviewed from Stanford University School of Medicine, CA. Four major types of HPE are delineated by MRI findings: alobar, semilobar, lobar, and middle interhemispheric (MIH) variant, the lobar and MIH types being most prevalent. In etiology, both genetic and environmental factors are identified. *Chromosomal anomalies* account for 40% and include trisomies (> half of these cases), duplications, deletions, and ring arrangements. Outcome in HPE cases with cytogenetic abnormalities is very poor, only 2% surviving beyond 1 year, compared to 30-54% for those without genetic causes. Smith-Lemli-Opitz syndrome carries an increased risk of HPE. Mutations have been identified in 15% to 20% of HPE cases with normal karyotypes. In the autosomal dominant form of HPE, *SHH* is the most frequently identified gene defect. *Environmental causes* include antiepileptic drugs, alcohol, smoking, statins, gestational diabetes, and cytomegalovirus infection. Some teratogens interfere with sonic hedgehog signaling pathways by disturbing cholesterol metabolism. *Neuroimaging abnormalities* include non-separation of deep gray nuclei, especially thalamus, monoventricle, dorsal cyst, hydrocephalus, Sylvian fissure abnormalities, and cortical dysplasia. *Clinical manifestations* include mental and developmental retardation, epilepsy, spasticity, dystonia, choreoathetosis, and endocrine disorders (diabetes insipidus, growth hormone deficiency). Craniofacial malformations are severe (cyclopia, midline cleft lip and