the right hemisphere in children between 1 and 3 years of age, whereas the left hemisphere was functionally dominant at 4 years and later. The change from right to left asymmetry was due to a shift in activity of the posterior associative area. This shift of asymmetry appeared to be functionally related to the visuospatial abilities subserved by the right hemisphere in the first year followed by emergence of language abilities at 3 years of age, a left hemisphere function. (Chiron C, Jambaque I, Nabbout R et al. The right brain hemisphere is dominant in human infants. Brain June 1997;120:1057-1064). (Respond: Dr Catherine Chiron, Service de Neuropediatrie, Hopital Saint Vincent de Paul, 82 avenue Denfert Rochereau, 75674 Paris cedex 14, France).

COMMENT. Right hemisphere cerebral blood flow (CBF) is higher than left at 1 to 3 years of age, whereas left CBF is higher in older children. Right hemisphere of infants is functionally dominant whereas the left becomes dominant after 3 years, when language develops. These significant CBF asymmetries are detected in sensorimotor cortex, Broca's area and the posterior associative regions which serve handedness and language.

RETT SYNDROME

EARLY BRAIN GROWTH IN RETT SYNDROME

Neuropathological and neurochemical studies supporting the concept of Rett syndrome (RS) as a neurodevelopmental disorder are reviewed from the Kennedy Krieger Institute and Johns Hopkins Hospital, Baltimore, MD. Prevalence in females is 1:10,000 to 1:22,000. Incomplete, forme fruste, cases are common. Twin data support a genetic basis but its nature is undetermined and no biological marker has been defined. Reduction in velocity of head growth begins at 2 to 4 months of age. Brain weight and size are reduced by 12 to 34% compared to age-matched controls in autopsy studies, but without evidence for progressive decrease in brain weight with age. Immunological studies suggest that certain pyramidal neurons are preferentially affected. Low levels of dopamine receptors and transporters are associated with decreased dopamine levels in the neocortex and basal ganglia. Choline acetyltransferase is reduced in the neocortex, hippocampus, putamen, and thalamus. Abnormal neurotransmitter systems can account for the clinical manifestations of RS. The early rapid evolution of symptoms followed by a static course suggest a neurodevelopmental disorder. (Naidu S. Rett syndrome: a disorder affecting early brain growth. Ann Neurol July 1997;42:3-10). (Respond: Dr SakkuBai Naidu, Kennedy Krieger Institute, #500 707 N Broadway, Baltimore, MD 21205).

COMMENT. Rett syndrome as a specific disease still eludes definition. Its static course after age five years is at variance with most genetic neurodegenerative disorders, and an abnormality in neurodevelopment appears more likely. My colleague Dr John Wilson at Great Ormond Street Hospital, London, suggests that the age-specific decelerating head growth in RS may be an apoptotic phenomenon.(In: Progress in Pediatric Neurology III, PNB Publ, 1997;p567).

A prevalence rate for RS of 2.17 per 10,000 girls is reported in a Norwegian study. (Skjeldal OH et al. Brain Dev June 1997;19:258-261). This higher than usual rate was associated with clustering in certain restricted geographical areas, a finding important in identifying a possible genetic basis

for RS.

Four cases of RS with a single family tree and prevalence rate of 2.1 per 10,000 are reported from Northern Tuscany, Italy. (Pini G, Milan M, Zappella M. Clin Genet Dec 1996;50:486-490). This study supports a genetic basis for RS.

METABOLIC DISORDERS

SUBEPENDYMAL CYSTS AND BIOTIN DEFICIENCY

Subependymal cysts were identified by cranial ultrasound and confirmed by MRI in an infant with holocarboxylase synthetase (HCS) deficiency, presenting with lactic acidosis, shock, and hypertonia, and responsive to biotin therapy (10mg daily), in a report from Devos Children's Hospital, Grand Rapids, MI. At delivery the infant had apnea and bradycardia, and Apgars were 7 and 8. Within hours she developed tachypnea, fever, and shock. Laboratory studies showed metabolic acidosis, hypoglycemia, lactic acidosis, and elevated pyruvate and ammonia. Urinary organic acid assays revealed increased 3-hydroxyisovaleric acid, 3-methyl-crotonoylglycine, and methylcitric acid, diagnostic of multiple carboxylase deficiency. Treatment with bicarbonate and biotin was followed by remission of symptoms and a normal development at 14 month follow-up. MRI at age 6 months showed resolution of the bilateral subependymal cysts and normal myelination. (Squires L, Betz B, Umfleet J, Kelley R. Resolution of subependymal cysts in neonatal holocarboxylase synthetase deficiency. Dev Med Child Neurol April 1997;39:267-269). (Respond: Liza Squires MD, Pediatric Neurology, Devos Children's Hospital, 330 Barclay NE, Grand Rapids, MI).

COMMENT. Subependymal cysts uncovered by cranial ultrasound in a sick neonate require investigation of possible metabolic disorders. Prompt diagnosis and specific therapy can prevent fatalities and permit normal development.

Glutaric aciduria type 1 is cited in association with cerebral arachnoid cysts (Millichap JG. Neurology 1997;48:1435); and L-2-hydroxyglutaric aciduria is reported in 6 Portuguese children presenting with mental deficiency, cerebellar ataxia, progressive macrocephaly and seizures. (Barbot C, Fineza I, Diogo L et al. L-2-hydroxyglutaric aciduria: clinical, biochemical and magnetic resonance imaging in six Portuguese pediatric patients. Brain Dev June 1997;19:268-273). A thalamic tumor, a diffuse fibrillary astrocytoma, was found in one of these cases, the second in the literature described with hydroxyglutaric aciduria.

MITOCHONDRIAL ENCEPHALOPATHY AND CYTOCHROME C

Benefits from treatment of mitochondrial encephalomyopathy (MEM) with cytochrome C (6.25 mg) and vitamins B1 (25 mg) and B2 (12.5 mg), in daily injections, are reported from Osaka University Medical School and other centers in Japan. Symptomatic improvements in 8 of 9 patients included decrease in muscle fatigability, and lessening of motor disability and severity of stroke-like episodes. Intermittent courses of injections were needed to maintain clinical improvement. (Tanaka J, Nagai T, Arai H et al. Treatment of mitochondrial encephalomyopathy with a combination of cytochrome C and vitamins B1 and B2. Brain Dev June 1997;19:262-267). (Respond: Dr Junko Tanaka, Sakai Municipal Hospital, Minamiyasui-cho 1-1-1, Sakai, Osaka 590, Japan).