bronchopneumonia. The children lived for 10, 23 and 32 months. At autopsy in 2 patients, the brain weighed approx 500 gms and showed marked hypoplasia of the corpus callosum, aqueductal stenosis, enlarged 3rd and 4th ventricles, widened cavum septi pellucidi, and diffuse degenerative changes with astrocytosis. The literature on anomalies associated with callosal defects is reviewed. (da-Silva EO. Callosal defect, microcephaly, severe mental retardation, and other anomalies in three sibs. Am J Med Genet April 1988;29: 837-843).

COMMENT. Congenital callosal defects occur alone or with other brain anomalies, e.g. septum pellucidum defect, hydrocephalus, porencephaly, polymicrogyria, and cerebellar hypoplasia. Mental retardation, seizures, and failure to thrive are commonly associated, but the callosal defect itself may be asymptomatic. At least 4 distinct syndromes include callosal agenesis as a major component: Aicardi (with infantile spasms, hypsarrhythmia, chorioretinal lacunae and coloboma), Schinzel acrocallosal syndrome (with macrocephaly, and polydactyly), Anderman's syndrome (with anterior horn cell disease), and Shapiro syndrome (with recurrent hypothermia). (See Ped Neur Briefs, March 1988;2:17).

HYPOMELANOSIS OF ITO

The neurological complications in 34 Spanish children with hypomelanosis of Ito are described from the Paediatric Neurology Service, Hospital Infantil La Paz, and La Universidad Autonoma, Madrid, Spain. Most were referred because of mental retardation (65%) and seizures (53%), and the ages at time of the first visit were 2 months to 10 years. Skin lesions, observed within the first year of life in 70% of patients, consisted of hypomelanotic depigmented patches, cafe-au-lait spots, and angiomatous nevi; changes in hair color and alopecia also occurred. Noncutaneous abnormalities, observed in 94%, included macrocephaly, microcephaly, hemihypertrophy, kyphoscoliosis, coarse facial features, and hypertelorism. Autosomal dominant inheritance was demonstrated in some. (Pascual-Castroviejo I et al. Hypomelanosis of Ito. Neurological complications in 34 cases. Can J Neurol Sci May 1988; 15:124-129).

COMMENT. The incidence of this disease was estimated at 1 per 1000 new patients consulting a pediatric neurology service, or 1 per 10,000 unselected patients in a children's hospital. It affects all races, but fair-skinned individuals may require a Woods lamp examination to detect the cutaneous lesions.

BRAIN LESIONS IN THE NEWBORN

NEUROPATHOLOGY OF PRENATAL BRAIN DAMAGE

Autopsy results of 89 infants who died at 7 days of age or less were analysed in the Depts of Pathology and Pediatrics, University of California School of Medicine, Davis, CA. Twenty-two (25%) showed evidence of prenatal brain lesions; 10 (16%) were preterm and 12 (48%) were term infants. Term infants were affected more often than

those born prematurely. The prenatal injuries in both premature and term infants were characterized by cerebral white matter necrosis and gliosis without hemorrhage. Hydramnios was the only maternal condition that predicted prenatal damage. Apgar scores were low, seizures were rare, and acute intracranial hemorrhage occurred equally often in infants with and without prenatal injury. the causes of death were primarily cardiorespiratory. The findings support growing evidence for the prenatal onset of brain injury in many infants who survive and later develop cerebral palsy. (Ellis WG et al. Neuropathological documentation of prenatal brain damage. AJDC Aug 1988;142:858-866).

COMMENT. Cordocentesis has been employed to detect prenatal hypoxia (see Ped Neur Briefs, June 1987;1:1) but the test is attended by technical risks and cannot be used routinely. As the authors comment, neonatal care will increase survival for prenatally damaged infants and the incidence of cerebral palsy may rise unless fetal/maternal abnormalities in late gestation are identified and corrected. There is need for a non-invasive and repetitive test for prenatal diagnosis of fetal hypoxia.

PAROXYSMAL DISORDERS

TREATMENT OF STATUS EPILEPTICUS

Very-high-dose phenobarbital was used for refractory status epilepticus in 50 children treated in the Neurology Division, Children's Hospital, University of Southern California School of Medicine, Los Angeles, CA. Intravenous boluses of 5-20 mg/kg, in increments of 10 mg/kg at 30-60 min intervals, produced a linear increase in drug level of 9.7 mcg/ml over a 24- to 48-hour time span. All patients were intubated prior to treatment. Side-effects, principally depression of respiratory drive and cardiac suppression with hypotension, were influenced more by the severity of the underlying disease and the seizures than by the use of the drug. Phenobarbital controlled seizures in all cases where limits were not imposed on the maximum dose by uncontrollable hypotension. Seven patients died, none during a period of rising drug level. Maximum serum levels ranged from 70-344 mcg/ml. (Crawford TO et al. Very-high-dose phenobarbital for refractory status epilepticus in children. Neurology July 1988;38:1035-1040).

COMMENT. Phenobarbital in adequate amounts given intravenously is a relatively safe and effective treatment for status epilepticus, but in those cases not responding to initial doses of 10-20 mg/kg, further amounts should be used only after the patient has been intubated. Pressor agents may be required to treat hypotension and assisted ventilation for respiratory depression. When the intravenous administration of drugs is not possible or practical, rectal therapy with paraldehyde, diazepam, or valproic acid has been recommended (see Ped Neur Briefs, Jan 1988;2:7) for termination of prolonged or serial seizures. The treatment of status epilepticus in children with rectal sodium valproate was reported from the Children's