

20mg/daily, but improvement was not maintained. The biochemical changes resembled the abnormalities observed in X-linked ALD and differed from those in the neonatal form. Comparison with other known peroxisomal disorders suggested a unique example of this category of disease. (Naidu S et al. Neonatal seizures and retardation in a girl with biochemical features of X-linked adrenoleukodystrophy: a possible new peroxisomal disease entity. Neurology July 1988;38:1100-7).

**COMMENT.** For an excellent review of the various entities now classified as generalized peroxisomal disorders, please refer to Naidu, Moser, Moser. Pediatr Neurol 1988;4:5 (reviewed in Ped Neur Briefs 1988;2:30). Immunopathological factors have been postulated in the pathogenesis of CNS lesions in X-linked adrenoleukodystrophy. Cyclophosphamide administered to 4 boys between 6 and 11 years of age with proven ALD failed to slow the rate of neurological progression. (Naidu S et al. Arch Neurol August 1988;45:846. Also, Stumpf et al. Arch Neurol 1981;38:48.

#### OUTCOME OF NEONATAL CONVULSIONS

The risk factors, causes, and prognosis of convulsions in 156 neonates are reviewed at the Mater Misericordiae Mothers Hospital, South Brisbane, Queensland, Australia. The incidence of early neonatal convulsions was 3/1000 live births. Compared to infants who did not convulse, the leading risk factors for convulsions were prematurity, intra-uterine growth retardation, low 5 min Apgar score, pre-eclampsia, antepartum hemorrhage, twin pregnancy, and breech presentation. The cause was hypoxic-ischemic encephalopathy (HIE) in 40%, intracranial hemorrhage (30%), metabolic (12%), infection (8%), malformation (3%), misc (7%). Mortality (31%) was related to etiology: 57% for intracranial hemorrhage, 33% infection, 27% HIE. Of the 107 infants who survived, long-term disability occurred in 43%; severe in 25 infants, moderate in 8 and mild in 10. The highest mortality and morbidity are associated with prolonged convulsions, tonic and multifocal clonic convulsions, convulsions due to asphyxia and intracranial hemorrhage, and an abnormal neurologic examination at discharge. (Tudehope DI et al. Clinical spectrum and outcome of neonatal convulsions. Aust Paediatr J August 1988;24:249-253).

**COMMENT.** The outcome of neonatal convulsions in this study is similar to that reported in a Dublin Collaborative Study of neonatal asphyxial seizures in which 43% had a poor outcome (Curtis PD et al. Arch Dis Childh September 1988;63:1065-8). In the Dublin study, asphyxial seizures occurring within 48 hours of birth in 0.87/1000 live births were correlated with antenatal complications, primiparity, and prolonged pregnancy. The incidence of seizures ranged from 0.55-1.2/1000 in the 3 participating maternity hospitals, reflecting differences in management policies in regard to frequency of cesarean section, induced labor, and forceps delivery. The mortality rate was 18% and of those who survived, 28% were handicapped at 1 year. Outcome was correlated with the infants' feeding habits at 1-2

weeks, those requiring tube feeding for more than 14 days being handicapped at follow up.

#### SURGERY FOR NEONATAL-ONSET SEIZURES

Four children with intractable neonatal-onset seizures treated successfully by hemispherectomy at 1 1/2-5 years of age are reported from UCLA School of Medicine, Los Angeles, California. Positron emission tomography (PET) with fluoro-D-glucose provided accurate localization of seizure foci whereas CT and MRI were either normal or showed mild generalized cerebral atrophy. The report illustrates the important role of PET in the evaluation of children with intractable epilepsy of neonatal onset. (Chugani HT et al. Surgical treatment of intractable neonatal-onset seizures: The role of positron emission tomography Neurology August 1988;38:1178-88).

COMMENT. The criteria for hemispherectomy were as follows: 1. Intractable unilateral seizures with diffuse epileptic activity in the affected hemisphere. 2. Persistent neurologic deficit on the contralateral side. 3. Malfunction of the affected hemisphere and intact function of the opposite hemisphere as tested by interictal EEGs, evoked potentials, thiopental test, and PET. At UCLA the results of surgery are impressive: the patients were seizure-free for periods up to 1 1/2 years and 3 patients were off all anticonvulsants. The surgical approach to treatment of refractory seizures appears superior to the conservative method with potentially toxic anticonvulsant drugs. The authors are to be complemented for their aggressive approach and search for alternate forms of early treatment.

#### NEONATAL CEREBRAL HEMORRHAGE AND ISCHEMIC LESIONS

##### SHUNTS FOR POST-HEMORRHAGIC HYDROCEPHALUS

The outcome of 19 infants who underwent cerebrospinal shunting for post-hemorrhagic ventricular dilatation is reported from the Department of Pediatrics and Neonatal Medicine, Hammersmith Hospital, London W12. Periventricular hemorrhages were diagnosed by ultrasound scanning, and surgery was considered necessary if the hydrocephalus could not be controlled by intermittent lumbar or ventricular tapping and the CSF pressure was above 6mmHg. Complications of ventriculo-peritoneal shunts included seizures at the time of the surgery in 8 infants, postoperative infection in 12 of 58 (20%) procedures and blockage of 29 shunts. Shunt infection with Staphylococcal epidermidis occurred in almost half the patients in spite of prophylactic antibiotics. Shunt blockage occurring in 70% of infants was less frequent in those over 2.5kg and with CSF protein below 1g/l. Long-term outcome was poor: 3 died, 4 were quadriplegic and mentally retarded, and only 4 (20%) were developmentally normal. Outcome was correlated with pre-operative parenchymal brain lesions diagnosed by ultrasound scans. (Hislop JE et al. Outcome of infants