

PEDIATRIC NEUROLOGY BRIEFS

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MALFORMATIONS

ENCEPHALOCELES: PROGNOSIS

The outcome of 34 infants with a diagnosis of cephalocele was reviewed at the University of Colorado Health Sciences Center, Denver, and was compared with that of previously published series. Of 22 posterior defects, 5 were meningoceles and of 12 anterior defects, 8 were meningoceles. Additional major abnormalities were present in 15 (44%) of 34 infants, 7 (58%) with anterior defects and 8 (36%) with posterior defects. Those with anterior defects had abnormalities of the cranium, while those with posterior defects had extracranial abnormalities with greater frequency. Mortality was 29% (10/34) and confined to infants with posterior cephalocele. Microcephaly was noted in 7 children all with posterior defects. Nine had hydrocephalus. Seizures occurred in 14 children, 5 with anterior defects and 9 with posterior defects. Outlook depended on site, operability, and additional major abnormalities. Mortality was confined to posterior defects, inoperability, congenital heart disease, infection, and shunt malfunction. Microcephaly, seizures, and hydrocephalus were associated with a worse outcome. Anterior defects have a better prognosis than posterior defects (Brown MS, Sheridan-Pereira M. Outlook for the child with a cephalocele. Pediatrics Dec 1992; 90:914-919). (Reprints: Mark S. Brown, M.D., P/SL Medical Center, 1719 E. 19th Ave., Denver, CO 80218.)

COMMENT. Cephaloceles (cranium bifidum) occur most frequently in the midline. The primary morbidity in children with anterior defects has included facial disfigurement, anosmia and visual problems. Although there were no deaths in infants with anterior defects in this series, significant long-term disability was present in 50% and was associated with severe facial clefting, intracranial abnormalities and visual impairments. In infants with posterior defects, severe long-term

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disabilities occurred in 25% and mild handicaps in 38% of surviving infants.

TERATOGENICITY OF ANTICONVULSANTS (AEDs)

The risks of teratogenic effects of AEDs are reported from Erasmus University, Rotterdam, The Netherlands. These consist of major malformations, minor anomalies, intrauterine or postnatal growth failure, and psychomotor retardation. The absolute risk of 7-10% is about 3-5% higher than that in the general population. None of the currently available AEDs is free of possible adverse effects on the fetus. Valproate and carbamazepine are associated predominately with spina bifida and hypospadias. Barbiturates and phenytoin are associated with congenital heart malformations and facial clefts. Risk factors include a high daily AED dosage, high maternal serum AED levels, low folate levels, or polytherapy. Genetic predisposition plays a role but no tests are available for identifying parents or fetuses at high risk. Prenatal diagnosis consists of fetal ultrasound during week 18-20, *a*-fetoprotein analysis of amniotic fluid in week 16 in mothers receiving VPA or CBZ (Lindhout D, Omtzigt JGC. Pregnancy and the risk of teratogenicity. Epilepsia 1992; 33 (Suppl 4):S41-S48). (Reprints: Dr. Dick Lindhout, Department of Clinical Genetics, Erasmus University, P.O. Box 1738, 3000 DR Rotterdam, The Netherlands.)

COMMENT. Prevention of teratogenic AED side effects should include the evaluation of the patient before conception, the need for AED therapy, monotherapy with the lowest possible dosage, avoidance of high peak levels by dividing daily dosage into 2 or 3 doses, folic acid supplement in cases of obvious deficiency and exclusion of vitamin B₁₂ deficiency. Minor anomalies tend to lessen or disappear with age (e.g. nail hypoplasia). Pregnancy and teratogenesis in epilepsy was the subject of a Neurology supplement 5, April 1992. The effects of prenatal exposure to anticonvulsants on intellectual functioning of 4-8 year olds are reported by Vanoverloop D et al. from the Department of Pediatrics, Harvard Medical School, Boston (Neurotox Teratol Sept/Oct 1992; 14:329-335). None of the children was mentally retarded, but scores for performance IQ and full scale IQ were lowered.

INFECTIOUS DISORDERS

BRAIN ABSCESS: MANAGEMENT

A review of 130 children with brain abscesses treated over 21 years is reported from Hacettepe University School of Medicine, Sıhhiye, Ankara, Turkey. Four (3%) were infants, 45% were aged 3-8 years, and 30% were 6-8 years. The incidence decreased between ages 8 and 18 years. Chronic ear infection with mastoiditis occurred in 28 (21%) and congenital heart disease in 26 (20%). Infants had meningitis or ventriculitis. The abscess was supratentorial in 113. In children with congenital heart disease, the parietal lobes were commonly involved with a left-sided predilection (77%). Multiple