

**1669** OUTCOME IN CHILDREN WITH PRENATALLY DIAGNOSED CONGENITAL HYDROCEPHALUS. Charles C. Duncan, Frank A. Chervenak, Laura R. Ment, David T. Scott, Richard A. Ehrenkranz (Spon. by H. A. Pearson) Yale Univ Sch Med, Dept of Neurosurg, Peds, Neurol, and Obs, New Haven, CT.

At our institution in a 5 year interval between 1978 and 1983, all 14 children with prenatally diagnosed congenital hydrocephalus, defined as excessively increasing ventricular size and biparietal diameter on at least two sequential examinations, were available for outcome evaluation. Others in whom this diagnosis was made either did not survive gestation or the initial neonatal period due to interruption of the pregnancy or intrapartum demise. GA at diagnosis varied from 26 to 38 wks with a mean of 31 wks. Age at delivery varied from 32 to 40 wks with a mean of 36 wks. Four of these children had myelomeningocele, 6 had hydrocephalus without other anomalies, three had intracranial cysts, and one had an encephalocoele. All of them underwent ventriculo-peritoneal shunting. Five had shunt related infections. Six had Bayley mental or Stanford-Binet scores >80 (43%), 2 had scores between 65 and 80 (14%), and 6 had scores <65 (43%). The poorer outcome of this group in comparison to other series of such hydrocephalus may relate both to the early stage at which the diagnosis was made such that children with more severe conditions have survived and the high rate of infection.

**1670** BIOGENIC AMINE METABOLISM IN RETT'S SYNDROME, Huda Y. El-Hibri, Alan K. Percy, Ian J. Butler, Baylor College of Medicine, Texas Children's Hospital, Dept. Pediatrics & Neurology, Houston, Texas.

Rett's Syndrome is a progressive disorder in females characterized by autistic behavior, dementia, ataxia, loss of purposeful use of the hands and seizures. Extensive laboratory investigations are usually normal with the exception of electroencephalography. To date no biochemical abnormalities have been reported. In an attempt to find a lead to the pathogenesis of this syndrome we assayed biogenic amine metabolites in the cerebrospinal fluid (CSF) of six patients, age 2-15 years, with Rett's Syndrome. The following metabolites were assayed and compared to age matched controls: 3-methoxy-4-hydroxyphenylethyleneglycol (MHPG), homovanillic acid (HVA) and 5-hydroxyindole acetic acid (5-HIAA). Results are as follows: Mean  $\pm$  S.D. (ng/ml).

	MHPG	HVA	5-HIAA
Patients	7.81 $\pm$ 1.49	64.70 $\pm$ 26.5	23.83 $\pm$ 6.25
Controls	12.35 $\pm$ 0.64	110.23 $\pm$ 25.83	32.07 $\pm$ 8.26
p-value	0.0005	0.0008	0.004

Thus, norepinephrine, dopamine, and serotonin metabolites are significantly reduced in the CSF of patients with Rett's Syndrome. This may provide new insight into the pathogenesis and future therapeutic intervention in these patients.

**1671** DIAGNOSIS OF CEREBRAL ATROPHY IN INFANTS BY THE NEAR-FIELD. METHOD: A NEW TECHNIQUE IN CRANIAL SONOGRAPHY by Asma Q. Fischer, Medical College of Georgia, Dept. of Neurology, Augusta, Georgia

Cerebral atrophy in infants has been difficult to assess by cranial sonogram (CS) because the subarachnoid space has been a "blind spot" by ultrasound, yet CS is an ideal imaging method for infants as it is portable, non-radiative and unlike computed cranial tomography (CCT), requires no intravenous contrast or sedation. We present a new method of evaluating the subarachnoid space by CS in infants and correlate it with the presence of cerebral atrophy. Material & Methods: Realtime ultrasound equipment with a modified transducer was used for imaging the "nearfield". The nearfield was defined as the first 2-3 cm from the scalp into the brain. The subarachnoid space which can be detected within the nearfield was assessed in 25 consecutive infants who were sonogrammed for various neurologic indications. The results were correlated with CCT findings of cerebral atrophy. Results: On nearfield sonography, 3 of the 25 patients had enlargement of the subarachnoid space. All 3 patients had cerebral atrophy on CCT. 22 of the 25 patients had no enlargement of the subarachnoid space by nearfield sonography. None of the infants who had CCT showed evidence of cerebral atrophy. Enlargement of the subarachnoid space by nearfield sonography was therefore found to be most sensitive and specific for cerebral atrophy when compared to CCT. Summary: Nearfield sonography provides a simple tool for assessing the subarachnoid space and therefore the presence of cerebral atrophy. It opens the way for a non-invasive method of evaluating the degree of cerebral atrophy and its correlation with the clinical state and prognosis. Larger series using this technique seems appropriate.

**1672** NEONATAL NEUROSONOGRAPHIC PREDICTORS OF CEREBRAL PALSY IN PRETERM INFANTS.

L. Graziani, M. Pasto, C. Stanley, F. Pidcock, H. Desai, S. Desai, B. Goldberg, Thomas Jefferson University Hospital, Departments of Pediatrics and Radiology, Philadelphia, PA.

In a prospective study, preterm infants of less than 33 weeks gestation were routinely evaluated by cranial ultrasonography at frequent intervals during their neonatal course. Thirteen survivors were found to have a spastic form of cerebral palsy at 12 to 36 months of age; two of the infants had a hemiparesis, 1 a monoparesis and 10 a diparesis. During the neonatal course of each handicapped infant, either or both of the following neurosonographic abnormalities were present: 1) frontally located periventricular porencephaly secondary to a grade IV intracranial hemorrhage (ICH), 2) extensive periventricular echodensity with subsequent cyst formation extending frontally. Ultrasound abnormalities consisting of ICH, or periventricular echodensity but without frontal porencephaly or persisting periventricular cysts were noted during the neonatal course of 49 other preterm infants who did not have cerebral palsy at age 12 to 36 months. The pathogenesis of the periventricular echodensities and subsequent cyst formation is unknown but the ultrasound studies are consistent with hemorrhagic infarction and ischemic lesions involving pyramidal tracts as they enter the internal capsule.

We conclude that spastic forms of cerebral palsy are likely to develop in preterm infants if neonatal ultrasound studies demonstrate persisting destructive lesions in frontally located periventricular brain structures.

**1673** IN VIVO VARIATION OF BRAIN OXIDATIVE METABOLISM WITH THERAPEUTIC LUMBAR PUNCTURE (LP). Ronnie Guillet, Barry Lawson, Donald Younkin, Eileen Donlon, Britton Chance, and Maria Delivoria-Papadopoulos, Univ. of PA. School of Medicine, Depts. of Pediatrics, Physiology, Biochemistry and Biophysics, Phila., PA.

Sequential cerebral spectra of phosphorus-containing compounds, ATP, phosphocreatine (Pcr), inorganic phosphate (Pi), phosphodiester (PD), and phosphomonoester (PME), by 31-P nuclear magnetic resonance (NMR) were obtained in a severely asphyxiated premature infant with hydrocephalus and seizures. Between days 2-20, 10 therapeutic LPs were done and NMR cerebral spectra were obtained pre (a) and post (p) LP. Pcr/Pi (normal  $\bar{x}$  = 1.01), a measure of bioenergetic reserve, was low at 48h; Left (L) = 0.50, Right (R) = 0.60. After the initial LP, Pcr/Pi rose to L=1.4 and R=0.6. After subsequent LPs, a sharp decrease in Pcr/Pi was followed by a rebound (a LP, L=1.50, R=1.03; p LP, L=0.87, R=0.62; 1 1/2 hrs p LP, L=1.52, R=1.66). Pcr/Pi increased by day 8 (L=1.52, R=1.60) then stabilized during the next month (L=1.0, R=0.8). Tissue pH did not change significantly (day 5: a LP=7.07, p LP=7.02, 1 1/2 hrs p LP=7.08; day 8: a LP=7.12, p LP=7.02, 1 1/2 hrs p LP=1.12). PME/ $\beta$ -ATP, a precursor in membrane biosynthesis, declined (day 2, L=3.15, R=2.53; day 4, L=1.46, R=1.17) but changed inconsistently post LP. Acute changes in Pcr/Pi and PME/ $\beta$ -ATP represent changes in local cerebral metabolism. Removal of 5 cc of CSF may result in acute intracerebral tissue shifts, acute hydrostatic changes with probable effects on CSF and cerebral blood flow, and hence provision of metabolic substrate to brain tissue. These data suggest that acute transient changes in Pcr/Pi and PME/ $\beta$ -ATP, representing changes in local cerebral metabolism, reflect general changes in cerebral metabolic or seizure activity.

**1674** CEREBRAL INFARCTION IN UTERO PRESENTING AS NEONATAL SEIZURES Erik A Hagen (Spon by Willa H. Drummond) Univ of Florida College of Medicine, Gainesville

Neonatal cerebral infarction (CI) is usually associated with risk factors such as birth trauma, asphyxia or coagulopathy, but there are reports of "idiopathic" infarctions without apparent risk factors. We have seen 2 cases of left middle cerebral artery (MCA) infarction presenting with seizures by one day of age in infants with "moderate" risk factors (meconium staining in case #1; variable/late decelerations, vacuum extraction, nuchal cord x 1 in case #2). In both infants, CT scans within 24 hours of age showed CI in the left MCA distribution, and, in both cases, these changes appeared to be subacute (i.e.: infarction occurred in utero). Case #1 demonstrated asymmetric cortical atrophy, ipsilateral optic nerve atrophy and absent nasal retinal vessels. This infant's seizures are well controlled with anticonvulsants, but by 6 months he has major developmental delays, spasticity and decreasing head growth. In case #2, the infant at 15 months has normal development with no physical or neurologic residua. On CT there is minimal residual evidence of CI with "islands" of normal brain tissue in the previous area of encephalomalacia. This infant is seizure free off medication. These cases are similar in presentation but remarkably different in outcome. CI can result in devastating permanent sequelae, but outcome may also be entirely normal. Prognosis in CI may be more dependent on severity of associated risk factors than simply on occurrence of the lesion itself.