1606 MITRAL VALVE PROLAPSE (MVP) SYNDROME IN DUCHENNE'S MUSCULAR DYSTROPHY (DMD): PREVALENCE, SPECTRUM AND ULTRASTRUCTURAL BASIS. Shyamal K. Sanyal, Warren W. Johnson. St. Jude Children's Research Hospital. Memphis, TN

The prevalence of MVP syndrome was determined in 20 boys with clinical, biochemical and electromyographic, and muscle biopsy evidence of DMD. Auscultatory evidence of a non-ejection systolic click, documented by phono, suggested MVP in 7 patients. Echocardiography confirmed MVP in these 7 and in 4 others. Pansystolic, anteriorly concave, posterior motion (> 3 mm beyond CD line) was seen in all patients, abrupt mid-systolic posterior motion in 5, and multiple sequence echoes in 6.

Hearts from 3 DMD patients with MVP were perfused for 4 hours with 2.5% glutaraldehyde at 4°C and ultrastructure, histology and gross anatomic features of mitral valve annulus, anterior and posterior leaflets, chordae tendinae, right and left ventricular myocardium and papillary muscles were studied and compared with hearts from age and sex-matched normal controls.

Gross, histologic and ultrastructural features of mitral valve leaflets, annulus and chordae tendinae in DMD patients were entirely normal. Myocardium, by contrast, showed multifocal areas of myofiber degeneration with fibrosis and loss of contractile elements – thick and thin myofilaments – producing a "moth-eaten" appearance of myofiber. These abnormal findings predominantly involved posterior papillary muscle and posterobasal segment of left ventricle.

Our observations establish: (i) a high prevalence of MVP syndrome in children with DMD and (ii) that MVP syndrome in DMD is an expression of underlying cardiomyopathy and is not due to dystrophic changes of the mitral valve leaflets, annulus or chordae tendinae.

1607 PROSPECTIVE LONG-TERM FOLLOW-UP OF PREMATURES WITH SUBEPENDYMAL/INTRAVENTRICULAR HEMORRHAGE (SEH/IVH). Howard S. Schub, Peter A. Ahmann, Francine D. Dykes, Anthony Lazzara, Brent Blumenstein (Spon. by James F. Schwartz), Emory University School of Medicine, Dept. of Pediatrics, Atlanta.

Since 1977, an ongoing study has assessed neurodevelopmental outcome of CT-documented SEH/IVH in infants <35 weeks gestation requiring intensive care. Scans were graded: normal, SEH, mild, moderate, or marked IVH. Follow-up status, at mean corrected age of 34 months, was assessed by neurologic exams, Bayley and Stanford-Binet tests. Outcome was designated: Good-no neurologic deficit and Developmental Index (D.I.) > 90; Intermediate-no or minor neurologic deficit and D.I. = 70-90: Poor-significant neurologic deficit or D.I. < 70. The following groups were compared: a) 33/41 surviving SEH/IVH infants with 30/49 non-IVH; b) 22 SEH/IVH infants paired with controls, matched for Apgar, gestation and birth weight c) intragroup, according to degree of hemorrhage. Of the 33 SEH/IVH infants, 21 had good outcomes, 8 intermediate, 4 poor. Of controls, outcome was good in 19, intermediate in 8, poor in 3. Among match-control pairs, there was a balanced distribution in outcome. Intragroup comparison showed: 13 had marked IVH with 8 good outcomes, 3 intermediate, 2 poor; 10 had moderate IVH with 5 good, 3 intermediate, 2 poor, 10 had mild IVH or SEH with 8 good, 2 intermediate, 0 poor. By all methods of comparison, outcome in SEH/IVH infants was not significantly different from controls. Marked IVH did not preclude good outcome (60% good). Other neonatal disease may affect outcome more than hemorrhage.

1608 PROLONGED AMBULATORY EEG (AEEG) RECORDINGS IN CHILDREN S. S. Seshia*, E. Shwedyk**, R. Thorne**, J. Hodges*, E. Moore*, J. Patrick*, (Spon. J. C. Haworth) Departments of Pediatrics* and Electrical Engineering**, University of Manitoba, Winnipeg, Manitoba, Canada.

Twenty-four hour AEEGs were done on 12 children (ages 4 months to 16 years) on a Medilog 4 channel cassette recorder (Ives and Woods, 1975); the tapes were analysed on a PDP 11/40 computer system and the data compared to that obtained from conventional EEGs (CEEGs).

Results: Group I: History of frequent daily seizures (N=8): 6/8 had several paroxysms on AEEGs similar to those on CEEGs; two of these showed a greater than 60% improvement in the AEEG after treatment. Paroxysms were not seen in 2/8 and frequent seizures were considered unlikely. Group II: (N=2): these two children with 2 and 5 normal CEEGs and a history of akinetic seizures had abnormal AEEGs. Group III: ?functional? complex seizures (N=2): one child (2 normal CEEGs) had a normal AEEG during episodes. A focal abnormality was identified in the CEEG and AEEG, in the other.

Problems: two AEEGs failed, presumably due to a faulty plug. A faulty channel in one case resulted from a damaged electrode.

Conclusions: (1) The EEG rhythms on AEEGs were comparable to those on CEEGs. (2) The AEEG provided clinically useful information in 6/12 children and has a practical application in pediatric practice.

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1609 REAL TIME ULTRASONIC SECTOR SCANNING IN DIAGNOSING INTRACRANIAL PATHOLOGY: Seetha Shankaran, Thomas L. Slovis, Mary P. Bedard, Ronald L. Poland. Depts.of Pediatrics and Radiology, Wayne State University and Children's Hospital of Michigan, Detroit, Michigan.

Real time (RT) ultrasonic sector scanning of the head was performed through the anterior fontanelle in 130 neonates for the following indications: birth weight <1500 g (60), suspected hypoxic ischemic injury secondary to perinatal asphyxia (16), or severe respiratory disease (12), neural tube defects (16), multiple congenital anomalies (12), abnormal head size (6), seizures (5) and cranial bruit (3). Additional diagnostic data was obtained from computerized tomographic (CT) scanning and post-mortem evaluation.

RT sonography detected 2/5 Grade I intracranial hemorrhage (ICH) 2/2 Grade II ICH, 11/11 Grade III ICH and 9/9 Grade IV ICH, as well as 1/2 vascular malformations and all 34 ventricular and 1 paraventricular abnormality. RT sonography failed to detect 3/5 Grade I ICH, 4/4 subarachnoid hemorrhage and 1/2 vascular malformations that were confirmed by CT scanning and/or autopsy. Sequential RT evaluations revealed 15/31 neonates developed post-hemorrhagic hydrocephalus 2-4 weeks following hemorrhage. Blood was seen in the ventricles from 1-9 weeks. Follow up evaluations at 2-10 months revealed that 10 of 15 had enlarged but stable ventricles, 2 had shunt insertions and 3 had normal ventricular size.

The sensitivity of RT sector scanning in diagnosing moderate and severe ICH and ventricular and paraventricular abnormalities was 100%. This should be the primary modality in evaluating neonates with intracranial pathology.

1610 THE ASYMMETRICAL TONIC NECK REFLEX (ATNR): INFANT INDICATOR OF LATER DEVELOPMENT. Bruce K. Shapiro, Frederick B. Palmer, Renee C. Wachtel, Pasquale J. Accardo, Alan Ross, Arnold J. Capute, (Spon. by Mark Batshaw), Johns Hopkins Medical Inst., J.F. Kennedy Inst., Dept. of Peds., Baltimore.

The ATNR of 249 subjects was scored as absent or present at serial well baby examinations (2,4,6,9,12 months). The ATNR was present at 2 months in 89% of subjects, 42% at 6 months, and persisted in 8% at 12 months. The Bayley Scales of Infant Development, Mental (Me) and Motor (Mo), were administered at one year. Data obtained were grouped by reflex activity--absent (-) and present (+)--and compared to outcome measures. Mean score for Me and Mo by ATNR activity for 2, 6, and 12 months are displayed below. Significant differences ($p<.05$) are noted (*).

	2 mos		6 mos		12 mos	
	(+)	(-)	(+)	(-)	(+)	(-)
Me	102.6	112.6*	103.0	104.2	97.5	104.7*
Mo	102.4	108.5*	103.2	103.1	96.8	103.7*

Absent ATNR activity at either 2 months or 12 months was associated with higher mean outcome scores. This finding reflects two populations--one with lower ATNR activity at two months and higher mean outcome scores and one with higher ATNR activity at 12 months and lower mean outcome scores. Similar findings were noted for the tonic labyrinthine and positive support reflexes. The differential disappearance of brainstem mediated motor reflex patterns reflects differential central nervous system maturation as measured in both cognitive and motor areas.

1611 ULTRASOUND-AUTOPSY CORRELATION OF THE INFANT BRAIN Michael W. Stannard, Jorge F. Jimenez (Spon. by M. Joycelyn Elders), University of Arkansas, Medical Sciences, Arkansas Children's Hospital, Departments of Radiology and Pathology, Little Rock, Arkansas.

Cranial ultrasound examinations were performed in five newborns and five infants under a year of age in the period immediately after death. Scans were made in multiple coronal planes angled from the anterior fontanelle.

After autopsy, the brains were fixed in formalin and then sectioned in planes corresponding to the ultrasound scans. These anatomic projections are not standard and are unfamiliar even in computed tomography. Comparison of brain sections with scans revealed the anatomic basis for the echoes.

Diagnostic ultrasound of the central nervous system is a noninvasive and relatively inexpensive new technique which is especially valuable during infancy while the anterior fontanelle is unossified and can act as a sonolucent window. It can be used to diagnose a variety of solid and cystic lesions and is ideally suited to follow-up examinations. Illustrative examples include a choroid plexus papilloma, a vein of Galen malformation and intracranial hemorrhage. Anatomy, pathology, and the ultrasound scans are correlated.