1588 PROGRESSIVE POSTHEMORRHAGIC HYDROCEPHALUS: FACTORS PREDICTING RESPONSE TO LUMBAR PUNCTURE MANAGEMENT
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Eighteen preterm infants with severe progressive post-hemorrhagic hydrocephalus (PPH) following IVH were managed with serial lumbar punctures (LP) according to the following protocol. Daily serial LP with measurement of opening and closing pressure (OP,CP) were performed for no more than four weeks. Sufficient CSF was removed with each LP to lower OP by half. If OP became normal ($\leq$ 80 mm H_2O) and remained so for two successive days, LP was deferred for 48 hours and then repeated. If OP remained $\leq$ 80 mm H_2O LP management was discontinued and the patient observed. If hydrocephalus progressed after discontinuation of LP or symptoms of increased intracranial pressure developed during LP management, shunt was effected.

Of the 18 infants thus far studied, 11 responded to LP management. OP in 10 of the 11 was $\leq$ 80 mm H_2O by three weeks; none of the non-responders demonstrated OP $\leq$ 80 mm H_2O prior to three weeks of therapy (p=0.0002). Initial CSF protein and sugar and response of CSF protein and sugar to LP were not predictive of response.

In conclusion, attainment of OP of $\leq$ 80 mm H_2O for 3 successive days within a 21 day period of serial LP management is highly predictive of PPH response to LP management.

1589 NEUROLOGIC STABILITY AND OLIGOSACCHARIDE EXCRETION IN MANNOSIDOSIS.
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Mannosidosis is noteworthy among lysosomal storage diseases affecting the nervous system for having a low mortality and a stable clinical course. Comparative studies of oligosaccharides were carried out on serum and urine of a 31-year-old man with mannosidosis (Arch Neurol 34:45, 1977) whose neurologic status has been stable since the age of 8 years. A mannose-containing trisaccharide with two mannose and one N-acetyl glucosamine residues was predominant in urine (304 mg/l), in agreement with other reported cases. By use of a sensitive new high pressure chromatographic procedure, a very low level of this trisaccharide was detected in serum (0.1-0.4 nmol/ml), with respective urinary values from 161 to 558 nmol/ml. The urinary clearance of trisaccharide was on average 16.1 times the glomerular filtration rate of creatinine, suggesting that the renal tubular cell is the immediate source of urinary oligosaccharides. The substrate burden in brain is much less in mannosidosis (0.11 µmol/g; J Pediatr 75:360, 1969) than it is in fucosidosis (19.9 µmol/g; J Neurochem 27:733, 1976), despite the fact that both diseases reflect a failure to catabolize glycoproteins. Our data suggest that upon maturation of the kidney a mannose receptor is elaborated which, by removing mannose-rich oligosaccharides from the circulation, favors the elimination of excess oligosaccharides and lowers the substrate burden for brain. (Supported in part by USPHS grants HD 05515 and HD 04147)

1590 DEMYELINATION IN PELIZAEUS-MERZBACHER DISEASE.
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The pathogenesis of Pelizaeus-Merzbacher (P-M) disease is unknown. A 17-year-old male, whose brother died at 12 years with pathological findings of classical P-M disease, had congenital nystagmus, ataxia, dementia, and progressive quadriparesis. Electronystagmographic testing showed prolonged latency and slow phase duration during rotation and caloric stimulation. These findings suggest ongoing involvement of myelinated fibers in brain stem-cerebellar connections which may underlie the abnormal eye movements of P-M disease. Electron microscopic examination of CSF ultrasediment (100,000 g) disclosed scattered cells containing lipid droplets (1-3 µ diameter) with unique satellite aggregation which were not represented in over 200 normal and pathological controls. In the white matter of the deceased sibling, the activities of cholesterol ester esterase (pH 6.6) and 2'3'-cyclic nucleotide phosphohydrolase were decreased to 40% and 20% of controls, respectively. The cholesterol ester fraction was 10X normal, and this fraction contained disproportionately increased amounts of oleic as opposed to arachidonic and other polyunsaturated fatty acids. This pattern is highly similar to that in other demyelinating diseases. These clinical and biochemical findings suggest that P-M disease is characterized by a chronic ongoing demyelinating process. (Supported in part by USPHS grants HD 05515 and HD 04147)

1591 CT EVALUATION OF NEONATES WITH SEIZURES ATTRIBUTABLE TO PERINATAL COMPLICATIONS.
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Although the incidence of neonatal seizures has been reported to be 0.2 - 0.8%, seizures are more prevalent in NICUs. During a 36 month interval, 116 (3%) of all our NICU patients had one or more seizures. In 75 (65%) patients, these seizures were attributable to perinatal complications, either intracranial hemorrhage or asphyxia, defined as a 5 min. Apgar score of $\leq$4, and could be divided by age of occurrence into early seizures (<24 hrs of age) and late seizures (>24 hrs of age). 56 of these 75 infants, 26 preterm (PT) and 30 full term (FT) survived for greater than 72 hrs. 24/26 PT and 24/30 FT infants were evaluated by CT scan. Of those 24 PT infants, 23 had late seizures at an average age of 55 hrs; 22 had intraventricular hemorrhages (IVH) and 1 had a subarachnoid hemorrhage (SAH). The other PT infant was asphyxiated and had an early seizure with a normal CT scan. Of the 24 FT infants, 19 had early seizures at a mean age of 9 hrs. 2 of those infants had subdural hematomas, 3 had focal regions of infarction, and the remaining 15 had normal scans. 5 FT infants had late seizures at a mean age of 56 hrs and had SAH. Thus, although echoencephalography has been found to be extremely useful for the diagnosis of IVH in PT infants, FT neonates with both early and late neonatal seizures should be evaluated by CT scan for the detection of intracranial lesions requiring immediate and/or close follow-up care.

1592 NEURODEVELOPMENTAL FOLLOW-UP OF VLBW NEONATES: EFFECTS OF GMH/IVH.
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During a 12-month period, our NICU admitted 65 VLBW neonates (BW <1250 g) who survived longer than 36 hr. CT scans and/or postmortem examination were performed on 62 (95%). CT scanning revealed germinal-matrix and/or intraventricular hemorrhage (GMH/IVH) in 27 of the 62 (43.5%), and GMH/IVH was found at postmortem in 7 others (11.3%), yielding a total incidence of 54.8%. There were no significant differences in the BW, GA, Apgars, sex, or mode of resuscitation in the survivors with or without GMH/IVH and in the deceased. No infants with Grades III or IV IVH survived. Of the 49 VLBW survivors, 32 (65%) have been followed with neurologic examinations and with the Bayley Scales of Infant Development at 3, 6, and 12 months corrected age. When the results for the survivors with and without IVH were compared at each age, no significant differences were found (see table). The incidence of major neurologic abnormalities was quite low and was similar for both groups. No infant with a Grade II IVH has required shunting for post-hemorrhagic hydrocephalus. We conclude that a Grade I or II GMH/IVH appears to add little neurodevelopmental risk for the early development of the VLBW infant.

	3 mos (n=12)		6 mos (n=10)		12 mos (n=4)	
	Mental	Motor	Mental	Motor	Mental	Motor
GMH/IVH	97.8	100.7	97.0	96.9	92.3	77.2
No GMH/IVH	89.6	95.4	95.5	92.3	102.3	88.0

1593 CEREBRAL SPINAL FLUID (CSF) PROTEIN AS AN INDICATION OF INTRAVENTRICULAR HEMORRHAGE (IVH)
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CSF protein levels performed during the second week of life can predict the presence of IVH of grade II-IV as scaled by Papille, et al. A screening test for IVH would be useful in those nurseries that do not have portable ultrasound capabilities and can obtain CAT-scans only with difficulty. In a previous study, RBC counts in CSF fluid have been determined not to reflect the presence or absence of IVH (Silverboard, et al.).

Twenty-four infants in our NICU had an IVH of grade II-IV diagnosed by CAT-scan. Fifteen of these infants had 33 lumbar punctures (LP) performed during the second week of life, only 4 specimens had CSF protein$<$ 300mg%. All patients had at least one CSF protein value greater than 300mg%. (normal$\leq$140mg%) The presence of many RBCs in CSF ($>$250,000) negates the validity of the test and 7 specimens were excluded for this reason. A group of 7 prematures with normal CAT-scans had CSF proteins of $<$300mg% and only 2 values of$>$200mg% on LPs performed during their stay. CSF protein values on infants with IVH averaged 156mg% during the first week, 672mg% during the second week and 288mg% beyond the second week of life. One 'false positive' occurred in a premature who had a large subarachnoid bleed on CAT-scan. No false negatives have been noted, though they may be expected to occur in small grade II hemorrhages. This is a useful screening tool in nurseries without portable ultrasound capabilities to diagnose IVH.