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INTRAVENTRICULAR HEMORRHAGE IN PREMATURES: NEUROBEHAVIORAL FINDINGS AND NATURAL HISTORY. David T Scott and Jacqueline R Farwell, Dept. of Pediatrics, Yale Univ. School of Medicine, New Haven. (Sponsored by Joseph B Warshaw.)

Twenty-seven cases of intraventricular hemorrhage (IVH) in premature infants in the Newborn Unit of Yale-New Haven Hospital were studied. IVH occurred in 30% of admitted infants of ≤32 weeks gestational age (GA). Male to female ratio was 20:7. The GA of the infants was ≤28 weeks in 8 cases, 29-30 weeks in 9, 31-32 weeks in 9, and 34 weeks in 1. Average 1- and 5-minute Apgars (4.3, 5.8) were similar to those of a series of prematures of comparable GA without IVH (4.8, 6.6); however, 30% of infants with IVH had Apgars of <5 at 5 minutes, while only 16% of infants without IVH did. Age at diagnosis ranged from <1 day to 14 days; hemorrhage occurred earlier with decreasing GA. Of 27 infants, 19 had developed respiratory distress syndrome, and 17 required mechanical ventilation. 19 patients died. Of the 8 survivors, 6 have ventriculomegaly, and 4 have had a shunt placed. Neurobehavioral evaluation by a modification of the Brazelton examination was performed on 13 infants after IVH, and scores were compared with scores of well infants of similar GA. It is of interest that, although infants with IVH had somewhat lower scores for tone and reflexes, the most marked differences between the two groups were found in average scores for measures of sophisticated behavior: adaptation to light (2.7 vs. 4.1), orientation to sound (1.9 vs. 3.6), alertness (1.4 vs. 2.7), and irritability (2.4 vs. 4.1).

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ATYPICAL NERVE HISTOLOGY IN A CASE OF FAMILIAL DYSAUTONOMIA TYPE II

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Familial sensory neuropathy with anhydrosis is a clinical complex in the group of conditions characterized by autonomic dysfunction and impaired perception to pain. We had the opportunity to study a patient who presented some features of this syndrome but whose sural nerve histology was strikingly different from that reported to date in this condition. P.R. came to our attention at the age of 6 mos. because of failure to thrive, frequent episodes of unexplained fever and indifference to pain. Intradermal histamine test was abnormal and methacholine test produced a miotic response. Anhydrosis and alacrima were present as were slowing of femoral nerve velocities and lower motor neuron denervation. Indifference to pain was present since birth and was evidenced by multiple scars on his lips and fingers. The fungiform, circumvallate papillae and taste perception were present. Abnormal EEG and a moderate delay in overall development were noted. The sural nerve biopsy showed an extremely small nerve and a virtual absence of unmyelinated nerve fibers with absence of Schwann cells but no evidence of a degenerative process. Clinically this case is best classified as FD type II in which absence of myelinated fibers only has been described. Absence of unmyelinated nerve fibers in a peripheral nerve appears to be a previously unpublished finding in this group of disorders, and might suggest that the process is developmental rather than degenerative.

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MENKES KINKY HAIR SYNDROME: SERIAL CT SCAN FINDINGS IN THREE CASES WITH PATHOLOGICAL CORRELATION IN ONE CASE. Alan R. Seay, Patrick F. Bray, Douglas Wing, Joel A. Thompson, James F. Bale, and Darryl M. Williams. University of Utah College of Medicine, Division of Pediatric Neurology, Departments of Neurology and Pediatrics, Department of Radiology, and the Division of Hematology, Department of Medicine, Salt Lake City.

The clinical courses and C-T scans of 3 patients with Menkes Kinky Hair Syndrome (MKHS) are described. Once the diagnosis was considered and confirmatory evidence of disturbed copper metabolism was collected the overall clinical neurological courses were similar. However, striking variation was noted in the patterns of head growth, varying from early and consistent microcephaly to accelerated head growth and increased intracranial pressure. Similar variation in the C-T scan data was noted, from diffuse cerebral atrophy to cystic encephalomalacia to findings which suggested large bilateral subdural hematomas. An angiogram and autopsy in one case document respectively striking blood vessel abnormalities and old infarcts and ischemic lesions. These case findings illustrate the need to consider MKHS in young infants with retarded growth, delayed development and seizures in whom other clinical and radiographic evidence strongly suggests abuse and neglect.

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DETECTION OF INTRAVENTRICULAR HEMORRHAGE (IVH) BY TRANSCEPHALIC IMPEDANCE (TCZ). Shameem F. Siddiqi, David R. Brown, Donald Dallmann, and Donald Reigel (Spon. by P. M. Taylor). Univ. of Pittsburgh School of Medicine.

TCZ, the resistance to a 100 μamp alternating current passed between two surface scalp electrodes, decreases with hydrocephalus in infants. Since the impedance of blood is greater than the impedance of water, the replacement of CSF by blood (as occurs in IVH) should raise TCZ.

Normal neonatal TCZ values were established from serial measurements during the first 8 days of life in 71 neonates ranging in gestational age (GA) from 26 to 43 wks. At all postnatal ages (PNA) with >15 observations there was a significant correlation between GA and TCZ ($r=0.62-0.85$, $p<0.01$). With the exception of day 5-6 for GA 26-29 wks, there were no detectable day-to-day changes in TCZ within GA groups.

Seven babies with autopsy-proven IVH had serial TCZ measurements throughout their clinical course. TCZ rose from 25.5±0.9 ohms (mean±SEM) before to 35.6±3.7 ($p<0.05$) after clinical deterioration. The latter value was also greater than the mean TCZ obtained from 19 neonates of comparable GA and PNA who did not have IVH (35.6±3.7 ohms vs 28.0±0.7, $p<0.05$). TCZ values rose >6 ohms from the first measurement to the last in 6/7 IVH babies but in only 2/19 normal babies (GA=26-30 wks), $\chi^2=18.80$, $p<0.001$.

TCZ reliably detected IVH. It is safe, inexpensive and easily measured. When obtained serially and prospectively, it has the potential to further define the clinical correlates, timing and pathophysiology of IVH. Since TCZ is related to GA it may also be useful in the clinical assessment of GA.

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INFANTILE SPASMS: EVALUATION OF A STANDARDIZED TREATMENT REGIMEN; William D. Singer, Edward F. Rabe, Jerome S. Haller - Department of Pediatrics (Neurology) Tufts - New England Medical Center Hospital - Boston, MA

Infantile spasms have been considered refractory seizures carrying a poor prognosis for seizure control and intellectual development. Treatment with conventional anticonvulsants, ACTH and corticosteroids, as well as newer agents such as nitrazepam, diazepam and clonazepam have produced variable results for seizure control, but they have all been associated with extremely high recurrence rates. Therefore, they have not been shown to alter the natural history of this seizure disorder.

This study presents an analysis of a standardized treatment regimen using ACTH in treating infantile spasms. Data is presented relating to each patient's neurologic status and seizure incidence on treatment and after a prolonged period of follow-up averaging 50 months. Fifty-five patients initially received ACTH, 40 units qd while hospitalized, followed by 80 units qod for a minimum of 3 months at home. The dosage was maintained at this level while improvement was being shown in seizure frequency and EEG pattern. ACTH was then tapered slowly. Thirty-one patients were treated within 1 month (GpI) and 24 patients more than 1 month after seizure onset (GpII). Complete seizure control of 87% of GpI patients and 56% of GpII patients ($P<0.05$) was achieved on ACTH. The recurrence rate was 3.2% for GpI and 12.5% for GpII patients. The seizure control rate for GpI surpasses previously reported studies in which recurrence rate of 33-67% have resulted in overall seizure control rates of only 15-40%. Our results substantiate the use of early and prolonged high dose ACTH therapy for infantile spasms.

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A STANDARDIZED NEUROLOGICAL EXAMINATION FOR INFANTS: OPTIMALITY SCORES FOR A HIGH RISK GROUP. Uwe Stave, Mailman Center, School of Med., Univ. of Miami, Fl.

The neurological evaluation of high risk infants in multicentered, comprehensive follow-up programs needs to be standardized for methodology and normality. Touwen has established the ranges of optimal responses with well defined methods for infants; from this we have derived a standardized neurological examination (SNE) for ages 3 and 6 mo. We have tabulated the results of 38 appropriately selected items of neuromaturational achievement and applied Prechtl's principle of an optimality score expressed as percent of items achieved. Scores are arbitrarily divided into normal, suboptimal and non-optimal categories. Comparisons are made among full-term, preterm and very small babies, and within these categories between infants who as neonates, had no specific diagnosis (NSD) and those with severe pathology (PATH). All differences are statistically significant

At 3 mo. corrected age: Neurological Optimality Scores in %

CATEGORY:	FULL-TERM		PRETERM		1000 - 1500 g BW	
NEONATAL DIAGNOSIS:	NSD	PATH.	NSD	PATH.	NSD	PATH.
number of patients	24	12	23	20	13	14
100-88=normal	79	17	50	20	46	21
87-75=sub-optimal	21	58	33	65	23	65
<75 =non-optimal	0	25	17	15	31	14

This instrument sensitively indicates neurological immaturity and/or dysfunction in the population of high-risk infants studied. A repeat of the SNE at age 6 mo. revealed marked improvements indicating progressive self-repair and/or catch-up.