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INAPPROPRIATE ADH SYNDROME (IADH) AND PERSISTENT METABOLIC ACIDOSIS (PMA) IN NEONATAL PERIVENTRICULAR HEMORRHAGE (PVH).

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PVH is the most common cause of death of LBW infants, accounting for 45% of deaths of infants < 1500 gm admitted to our ICU in 1976. To delineate the signs of PVH, we have analyzed the clinical findings of infants who died in 1976 with suspected PVH. Mean birth weight $\pm$ SD = 1070 ± 273 gm; gestational age = 31 ± 2.7 wks. A clinical diagnosis of PVH was made at 1-15 days (mean = 3.1) and confirmed by autopsy in 13 (Group I). 13 had no autopsy, but had bloody CSF and severe neurologic signs (Group II). Significant changes in hematocrit, blood pressure, temperature, and blood glucose each occurred in 20-50% of infants. 12 (46%) required mechanical ventilation despite an $\text{FiO}_2 < 0.3$. 23 (88%; 11 Group I) had PMA with $\text{pH} < 7.25$ for > 8 hrs. despite oxygen and transfusion therapy; 7 (27%) had a $\text{pH} < 7.20 > 8$ hrs. NaHCO_3 was used sparingly and serum $\text{Na}^+ > 145$ mEq/L occurred in only 1 infant. IADH was suspected in 12 (46%; 5 Group I), each having concentrated urine, weight gain, and serum $\text{Na} < 130$ mEq/L (mean = 119 mEq/L). Urine Na^+ recorded in 7 was > 35 mEq/L. 11 (42%) had both IADH and PMA within 24 hrs. of other signs. IADH, rarely reported in neonates, appears to occur commonly with PMA as a sign of PVH. In view of an 88% incidence of PMA, a causal relationship of NaHCO_3 therapy to PVH can be established only if treatment precedes onset of signs.

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ABNORMAL CATECHOLAMINE EXCRETION IN PATIENTS WITH SYDENHAM'S CHOREA.

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The pathogenesis of Sydenham's (rheumatic) chorea is not well understood. Laboratory tests are not helpful in establishing the diagnosis. The chorea can be successfully treated with haloperidol, an agent known to interfere with the binding of dopamine to its receptors. This suggests that dopamine, and its urine catabolite homovanillic acid (HVA), might be increased in Sydenham's chorea. To test this hypothesis the urines of three patients with the clinical diagnosis of Sydenham's chorea were analyzed for HVA and Vanylylmandel acid (VMA) during the acute phase of the disease. Urine HVA was 28.07 ± 6.1 mcg per mg. creatinine (m $\pm$ SEM). Seven age matched control children had urine HVA levels of 8.8 ± 0.7 mcg per mg creatinine (m $\pm$ SEM) ($p < 0.01$). Urinary VMA was 8.7 ± 3.2 mcg per mg creatinine in the patients with chorea, and 6.4 ± 0.6 in the control group ($p > 0.1$). The ratio HVA:VMA was 3.7 ± 0.7 (m $\pm$ SEM) in the 3 patients, with chorea and 1.4 ± 0.2 in the control group ($p < 0.005$). In conclusion, urine HVA is increased in patients with Sydenham's chorea, suggesting increased dopamine, the metabolic precursor of HVA, in the disease. The determination of HVA and HVA:VMA ratio may be helpful in establishing this diagnosis.

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SYSTEMIC CARNITINE DEFICIENCY: A CAUSE OF RECURRENT "REYE'S SYNDROME."

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A white female had two episodes of protracted vomiting followed by an acute encephalopathy after a "viral" illness at 11 months and 4 9/12 years of age diagnosed as Reye's syndrome on the basis of laboratory data (most abnormal value given; value at 11 months given first) serum glucose-15,12 mg/dl; serum ammonia-97,144 ug/dl (nl<48); SGOT-105,103; prothrombin time-47%, 49%; CSF-normal and at 4 9/12 years a liver biopsy showing extensive small vacuole fatty degeneration. A third mild episode occurred at age 5 years. Evaluation following recovery revealed minimal proximal muscle weakness, a non-specific EMG abnormality and no evidence of a urea cycle disorder. A fast had to be terminated after 18 hours when she vomited several times and became very lethargic. A muscle biopsy, obtained at 5 1/2 years at a time of clinical remission, contained excess lipid; a liver biopsy was histologically normal. Free carnitine levels were: serum 8.66 nm/ml (nl range 27.9-67.2) muscle 4.22 nm/mg non-collagen protein (NCP) (nl 7.96-22.86) and liver 0.4 nm/mg NCP (nl 3.3-10.4). In 7 children with single episodes of Reye's syndrome serum (n=4), muscle (n=3) and liver (n=1) free carnitine levels were normal except for one slightly low serum level. Systemic carnitine deficiency may mimic "Reye's syndrome"; the episodes, some initiated by fasting, may be preventable by carnitine therapy.

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NONINVASIVE MEASUREMENT OF INTRACRANIAL PRESSURE: EVALUATION OF A TECHNIQUE

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In 1976 a new technique for measuring intracranial pressure transcutaneously across the anterior fontanel using an aplanation fiberoptic methodology was introduced. The present study attempted to duplicate the previous work. Materials and methods: 4 infants with open fontanels and suspected increased intracranial pressure were studied. The Ladd Intracranial Pressure Monitoring Device (model 1700, Roche Medical Electronics) was applied to the anterior fontanel as per the original authors. Continuous digital and graphic displays of measured pressures were obtained. Results: A pressure range of 10-26 cm H_2O was obtained by altering tension applied to the straps which hold the transducer in place. Manipulation of the straps did not result in a reproducible baseline. Pressures measured represented the sum of intracranial (transfontanel) pressure and externally applied pressure and interpretation of data became impossible. Conclusion: Previous results using this technique could not be duplicated. The application of an aplanation transducer to measure pressure across a deformable membrane necessitates establishment of co-planarity of transducer and membrane and the introduction of external pressure into the system. The technique under study does not quantify the amount of external pressure required to achieve co-planarity and thus cannot give meaningful data.

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ISOLATED HYPERMETHIONINEMIA WITH BILATERAL OPTIC NERVE HYPOPLASIA.

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Persistent hypermethioninemia unassociated with homocystinuria, cystathioninuria, or liver disease has been reported in a clinically normal infant (Science 186:59, 1974). We know of two unreported cases and have recently encountered a fourth case in a 9-month-old infant evaluated for apparent blindness and developmental delay.

Pregnancy was complicated in the first trimester by excessive alcohol intake, ten-pound weight loss, and ingestion of diethyl-propion HCl (Tenuate). Birth weight was 2460 gm., small for gestational age of 39 weeks, with APGAR's of 71 and 105 following fetal distress. Family history was not contributory. At nine months the baby was unable to fixate or follow and had roving, nystagmoid eye movements with poor pupillary reactions to light. Optic discs were less than one-half the normal size for age. A 3-4 month lag was seen in motor and language development. EEG showed occipital spike discharges. CT scan demonstrated enlarged occipital horns of the lateral ventricles. Serum methionine value was 164 $\mu\text{moles/dl}$ (normal 2.7 ± 0.5). All other serum amino acid concentrations were normal. Urine amino acids and organic acids were normal except for elevated methionine. Spinal fluid methionine was 14.5 $\mu\text{moles/dl}$ (normal 0.26 ± 0.16). The ratio of serum to spinal fluid methionine was 11.3 (normal 9-13).

A link between the findings of isolated hypermethioninemia and optic nerve hypoplasia is suggested by the association of hyperornithinemia and gyrate atrophy of the retina as well as homocystinuria and ectopia lentis.

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SLEEP STUDIES IN INFANTS BETWEEN BIRTH AND SIX MONTHS OF AGE.

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Altered sleep state patterns have been associated with various clinical entities in the perinatal period, such as infants of diabetic mothers, infants of addicted mothers and premature infants with an abnormal neonatal course. The objective of this study was to provide normative laboratory sleep state values during the first six months of life. Twelve-hour polygraphic measures were obtained in 10 full term infants during the first week of life and at 1, 2, 3, 4 and 6 months of age. Each minute of the recording was coded as Awake (AW), Active Sleep (AS) or REM, Quiet Sleep (QS) and Indeterminate (IN). A computer program calculated sleep variables. The entire study was replicated in another group (N=10). Results were assessed with an analysis of variance. The total percent of time spent in AS decreased with age. This was due to the decrease in the number of AS episodes. Both the duration of AS episodes and intervals between them remained the same. The percent of time in QS increased with age. This was due only to an increase in the duration of QS episodes. The average percent of time spent in AW and IN remained the same. The mean number of awakenings (16) at one and two months was twice as high than at four and six months. Although variability among infants was high, the age effects described were robust. Only relatively large departures from normative values are indicative of abnormal developmental patterns.