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CENTRAL NERVOUS SYSTEM (CNS) LUPUS ERYTHEMATOSUS (LE) IN CHILDREN. I.G. Dresner, N.K. Dysart, A.F. Michael, R.L. Vernier, A.J. Fish, Department of Peds., Univ. of Minnesota Medical School, Minneapolis, Mn.

In 67 unselected patients under the age of 20 years with LE, evidence of CNS disease was present in 18% (3M,9F). Abnormal neurologic findings included: chorea-7, upper motor neuron signs-3, cranial nerve paresis-3, cerebellar dysfunction-2, focal seizures and coma-1, stroke associated with hypertension-2. Chorea is the major abnormality in this group; in 3 patients it presented as long as three years prior to the diagnosis of LE. Significant renal disease was present in 8 patients. Laboratory tests were not helpful in diagnosis. Lumbar puncture and brain scan were normal unless stroke was present. EEG was always abnormal but correlated with neurologic findings and improvement. Serologic parameters were only mildly abnormal during recurrences of CNS disease. Standard doses of prednisone and Imuran used to treat LE exacerbations corrected all major CNS abnormalities except neurologic deficits secondary to strokes. Three patients had recurrences of CNS disease which responded to retreatment with high-dose therapy. This study provides the first comprehensive view of the high incidence of CNS involvement in a non-selective series of childhood LE patients and emphasizes the need to recognize subtle abnormalities and initiate adequate therapy.

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POST-HEMORRHAGIC ENCEPHALOPATHY IN PREMATURE INFANTS. Orest Dubynsky, Robert Vannucci & M Jeffrey Maisels, Penn State Univ-M S Hershey Med Ctr, Dept Ped, Hershey

Post-hemorrhagic encephalopathy is a distinct clinical condition resulting from intraventricular hemorrhage (IVH) in premature infants. Nine neonates with IVH were studied to detect possible chronic encephalopathy. Serial CSF analyses revealed persistent xanthochromia but variable RBC (679-132xK/mm^3) & WBC (0-3780/mm^3) counts. Protein contents were elevated (>100 mg/dl) in 7 patients (61-2420mg/dl). Hypoglycorrhachia (CSF/blood glucose ratio< 0.35) occurred in 7 cases & CSF glucose was< 10 mg/dl in 5. Hypoglycorrhachia was not secondary to persisting cellular elements in CSF, since no correlation existed between CSF glucose level & total cell count. In addition, in vitro incubation of cellular CSF with added glucose at 37°C led to negligible rates of glucose consumption over 24 hr. Three infants manifested gross hydrocephalus documented by CAT scanning. CSF lactate in these 3 infants was increased 1-3 fold (2.3-5.5 mmol/l) above levels in non-hydrocephalic IVH neonates (1.1-1.6 mmol/l) & in patients with congenital (non-hemorrhagic) hydrocephalus (1.0-1.4 mmol/l). We speculate that IVH occurs concurrently with periventricular brain ischemia which, in turn, leads to focal anaerobic glycolysis & increased glucose requirement. With inadequate cerebral glucose delivery from blood, glucose diffuses into brain from CSF resulting in hypoglycorrhachia. Cerebral lactate production is enhanced but lactate accumulates in CSF only in the presence of an obstructed ventricular system. These metabolic alterations merit consideration of therapeutic intervention.

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ETHACRYNIC ACID THERAPY OF CEREBRAL EDEMA IN THE NEWBORN. Patricia H. Ellison, Mhairi G. MacDonald, and Robert B. Bourke (Spon. by Ian H. Porter). Albany Medical College, Albany Medical Center Hospital, Departments of Neurology and Pediatrics, Albany, New York.

The effect of ethacrynic acid on cerebral edema of the newborn was evaluated in 9 infants who experienced severe hypoxia-ischemia during delivery. Six expired, without clinical or pathological evidence of cerebral edema at autopsy. The 3 surviving infants are normal developmentally. One has a seizure disorder.

Ethacrynic acid has been used effectively in immature animals to prevent cerebral edema. Bourke et al. have used ethacrynic acid in the immature cat to prevent edema that was K+ dependent, HCO_3-stimulated and appeared following the stage of brain maturation characterized by glial proliferation.

The human infant has significant glial cell proliferation by 35-36 weeks of gestational age and becomes subject to severe cerebral edema at that time. The infant who experiences anoxia is considered to sustain cellular damage initially from hypoxemia-ischemia and secondarily from cerebral edema. The goal of treatment with ethacrynic acid is to change the impact of the secondary cerebral edema. Criteria for initiation of treatment were strict: apgar of 0-1 at 1 minute and less than 3 at 5 minutes, or late or variable deceleration with heart rate under 100 beats per minute for 20 minutes or longer during the delivery. After resuscitation and stabilization, 1 mg/kg per IV was given every 12 hours for a 48 hour period.

Our initial study suggests that ethacrynic acid is a safe and effective drug for treatment of cerebral edema in the newborn.

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CEREBRAL OLIGODENDROGLIOMAS OF CHILDHOOD. J R Farwell, G J Dohrmann (sponsored by H A Pearson). Dept. of Pediatrics and Section of Neurosurgery, Yale University School of Medicine, New Haven.

Oligodendrogliomas are rare in children. In 40 years (1935-1974), 12 histologically verified cases of oligodendroglioma in persons <20 years of age were reported to the Connecticut Tumor Registry, constituting 1-2% of all intracranial neoplasms in Connecticut children. Eleven occurred in the cerebral hemispheres; these cases were reviewed and the available slides were analyzed. The male to female ratio was 2.7:1; the age range was from 2 to 19 years with a mean age of 13.5 years. The chronicity of the process was evidenced by a mean duration of 50 months from the onset of symptoms to diagnosis. Seizures (64%), nausea/vomiting (55%), headache (45%), and cranial nerve palsies (45%) were the most common presenting symptoms while papilledema (45%) and paresis (27%) were the most frequent physical findings. A normal neurological exam was found at diagnosis in 36% of the children. Ten patients were operated upon; 7 of these also received radiation. One patient died before diagnosis. The mean survival with cerebral oligodendrogliomas was 61 months. Comparison of those treated with operation alone and those also treated with radiation revealed no statistically significant difference in survival. Six patients survived longer than 5 years, and the functional status of these longer term survivors was determined: one has no neurologic deficit; one has a quadrantanopsia; one has a hemiparesis; two have seizures; and one has psychiatric disease and seizures.

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FOLLOW-UP STUDIES OF INFANTS WITH ABNORMAL NEONATAL COMPUTED TOMOGRAPHY (CT) RESULTING FROM ASPHYXIA. Pamela M. Fitzhardinge, Charles R. Fitz, Derek C. Harwood-Nash. Univ. of Toronto, Hosp. for Sick Children, Depts. of Pediatrics and Radiology, Toronto.

20 full term infants had abnormal CT during the neonatal period. All had histories of peripartum asphyxia followed by seizures. Bleeding into the ventricles and/or adjoining brain tissue was present in 7 and was extensive in 5. Areas of decreased cortical density secondary to anoxia were present in the remaining 13. In 3, these changes were confined to one hemisphere with a pattern suggesting infarction of the middle cerebral artery. Severe edema obliterating the ventricles was present in 2 others.

All patients have been followed for a minimum of 9 mo with a repeat CT done at 6-12 mo in 18. 12 of the 20 have severe neurological abnormalities: 2 hydrocephalus; 7 cerebral palsy; 3 microcephaly with retardation. All 18 follow-up CT show brain atrophy with localization and extent corresponding to the neonatal CT. The 5 children with extensive intracranial bleeding are severely defective; the other 2 have minimal dystonia and developmental delay. All 3 patients with unilateral changes are hemiplegic. The 2 patients with severe neonatal edema have central atrophy on follow-up CT and spastic quadriplegia with retardation.

These results show that CT in the asphyxiated neonate is highly predictive of the type and severity of later neurologic deficits, especially in cases showing severe edema, major vessel infarction, or extensive intracranial hemorrhage.

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^{64}Cu UPTAKE IN CULTURED, X-CHROMOSOME LINKED COPPER MALABSORPTION (MENKES' DISEASE) CELLS. J. French, I. Spigland, J. Rosen, H. Nitowsky, Albert Einstein Coll.Med., Montefiore Hosp. and Med. Ctr., Dept.Ped., Pathology and Neurology, The Bronx, NY.

The number of cells, DNA, RNA, protein and ^{64}Cu uptake per unit fibroblast culture were determined from skin of x-chromosome linked copper malabsorption (X-cLCM) patients and amniocentesis cells of a pregnancy at risk. Appropriate controls were similarly evaluated. The ^{64}Cu uptake per unit RNA was the most discriminating determinant between the control and X-cLCM populations ($0.005<p<0.01$). The ^{64}Cu uptake per unit protein was the least discriminating determinant ($0.02<p\leq0.05$). Intermediate values ($0.01<p\leq0.02$) were found for the ^{64}Cu uptakes per unit DNA and per 10^6 cells. The data do not reveal any significant differences between cultured X-cLCM and control fibroblasts' rate of replication, transcription or translation. These findings suggest that Horn's description of an increased ^{64}Cu uptake per unit protein in X-cLCM cultured cells (Lancet 1:1156-1158, 1976) is not due to an alteration of protein synthesis. This technique may prove useful for antenatal discrimination of X-cLCM fetuses.