1135 NEUROLOGIC DEVELOPMENT OF PREMATURES TREATED FOR NEONATAL HYPOCALCEMIA(HC). Gary M.Chan, Marilyn M. Chan, Reginald C. Tsang, Sharon L. Elsass, U. of Cincinnati.

During the first 72 hrs of life, 30% of premature (≤37wks) develop HC. Possible sequelae of early HC have not been examined. Twelve well AGA prematures were studied in matched pairs (gestation and Apgar) and given prophylactic doses of 1,25 dihydroxyvit D_3 ($1,25(OH)_2D_3$ - the active D metabolite), or vit D_2 400 IU(controls) during the first 3 days. Infants were studied prospectively for degree of tremors on day 1,2,3; Brazelton Behavior Assessment (BBA) at 40wks postconception; and Gesell Developmental Exam at 3 & 6 mos. Exams were blinded to ionized Ca (iCa,Orion SS-20) and treatment group. All prestudy iCa were <3.5mg%. By 48hrs, the $1,25(OH)_2D_3$ group had significantly higher iCa, 3.6±0.1mg% (mean± SEM) vs 3.2±0.1 at 12 hrs (paired t,p<.05); controls did not change. Degree of tremors was not related to iCa. Controls were less responsive to BBA orientation (visual, auditory and combined) items than $1,25(OH)_2D_3$ group, 4.1±.16 vs 5.0±.21(t test,p<.01). The 3 mo Gesell was lower for controls vs $1,25(OH)_2D_3$, 89±3 vs 99± 4 (p<.06) and was 110±1 vs 116±1 at 6 mo (p<.01). Eight of the 12 infants were rematched blindly in pairs for Apgar, gestation and HC at 48 hrs vs normocalcemia (NC). HC infants appeared less responsive than NC in the BBA visual-auditory item, 5±0.6 vs 6±.4 (Wilcoxon-Mann, p<.07), to need more consoling (6.6±0.3 vs 7.6±.4, p<.10) and to be more excitable (5.6±.3 vs 6.3±.4, p<.10). No differences were found between HC and NC in 3&6 mo Gesell. Early neonatal hypocalcemia appears to have an effect on the newborn's ability to respond to social(orientation) stimuli; long term sequelae are uncertain.

1136 CHRONIC KETOSIS AND CEREBRAL METABOLISM. Darryl C. DeVivo, Mary P. Leckie, and James A. Ferrendelli, Wash. U. Dept. Pediat. & Neurol., St. Louis, Mo.

We propose that cerebral ketone body utilization underlies the anticonvulsant mechanism of a ketogenic diet. Blood and forebrain analyses (freeze-blowing technique) performed on adult rats fed an 80% fat diet for 20 days (FF group) were compared to a control group. Blood [glucose] was ↓ and blood [βOHB] and [acetoacetate] were ↑ ($p < .005$) in the FF group. Brain concentrations of Na,K, Cl, glucose, malate, AMP, phosphocreatine, oxaloacetate and water content were similar in the two groups. Brain glycogen, glucose 6-P, pyruvate, lactate, βOHB, citrate, α-ketoglutarate, and ATP concentrations were ↑ ($p < .005$) and brain fructose $1,6\text{-}P_2$, ADP, cyclic AMP, and cyclic GMP concentrations were ↓ ($p < .05$) in the FF group. We conclude from these observations that chronic ketosis effects cerebral metabolism: (1) like certain anticonvulsant drugs by slowing brain phosphofructokinase, pyruvate dehydrogenase, and α-ketoglutarate dehydrogenase; (2) by ↑ the brain/blood glucose ratio by 21% and the cerebral energy reserves by 12%; (3) unlike certain anticonvulsant and sedative drugs, by maintaining the concentrations of citric acid cycle intermediates reflecting the precursor role of ketone bodies for acetyl-CoA synthesis; and (4) by decreasing cyclic nucleotide concentrations 12-21% presumably reflecting a depression in neuronal activity. It is suggested that each of these metabolic effects contributes to the anticonvulsant action of the ketogenic diet.

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1137 INTRACRANIAL NEOPLASMS IN INFANTS. J R Farwell, G J Dohrmann, & J T Flannery. (Spon. by Howard A Pearson) Yale Univ. School of Medicine, Dept of Pediatrics, & Sections of Neurosurgery and Neuropathology, New Haven, CT 06510.

Intracranial neoplasms are an unusual but important cause of large head circumference or neurologic dysfunction in infants. To delineate the presentation, course, and outcome of these tumors, the 54 primary intracranial neoplasms of central nervous system origin diagnosed in infants (≤18 months of age) in Connecticut in a 40-year period (1935 to 1974) were reviewed. 30% were medulloblastomas, 16% were ependymal neoplasms, 13% were meningeal neoplasms; other histopathologic types included astrocytoma, dermoid/ teratoma, choroid plexus papilloma, and glioblastoma multiforme. Equal numbers of males and females were affected. 44% of the tumors were located in the cerebellum, 37% in the cerebral hemispheres, and 17% in the brain stem. 6 infants were diagnosed within the first few days of life. Symptoms included vomiting in 47%, abnormally increasing head circumference in 32%, lethargy or irritability in 19%, and convulsions in 13%. Physical findings included abnormally large head circumference (45%), bulging fontanelle, cranial nerve palsies, papilledema, and nuchal rigidity. 39 infants were treated by operation, radiation, or both; 15 were not treated. Average survival was 43 months. Survival varied relative to histopathologic type (longest with choroid plexus papillomas, shortest with ependymal neoplasms), and location of neoplasm. Over half the infants died within 6 months of diagnosis. Neurologic sequellae were frequent: of 18 infants surviving longer than one year, 8 are severely retarded or handicapped.

1138 DEVELOPMENT OF CORTICAL BEHAVIOR IN VERY PREMATURE INFANTS: ADAPTATION AND ORIENTATION. Jacqueline R Farwell and David T Scott (Spon. by Joseph B Warshaw) Yale Univ, School of Medicine, Dept of Pediatrics, New Haven, CT.

Tone, reflexes, and gross motor activity do not always reflect neural maturation with advancing gestational age (GA). To improve assessment of neurobehavioral development, the Brazelton Scale was adapted for use with very premature infants. 15 well infants of ≤32 weeks GA, whose courses were benign (ie, without disease or complication), were examined at ≤3 days of age. Preliminary results suggest that adaptation and orientation scores (cortical measures) correlate well with GA:

GA	N	ORIENTATION ITEMS				ADAPTATION ITEMS		
		Light	Rattle	Face	Voice	Light	Rattle	Pinprick
28	4	1.67	2.75	1.00	2.00	2.75	0.75	1.50
30	7	2.20	3.60	2.50	3.40	4.80	5.00	2.17
32	4	2.50	4.50	2.33	2.75	5.00	4.25	2.50

Four items (adaptation to light and pinprick, and orientation to light and rattle) showed rising linear trends across age groups. Three remaining items (adaptation to rattle and orientation to face and voice) clearly differentiated between 28- and 30-week gestation infants, but did not distinguish between 30- and 32-week gestation groups. Average scores on other measures (alertness, irritability, level of activity, reaction to a cloth on the face) were not reliably different among the three groups of prematures. Tone and standard reflexes, similarly, did not change uniformly with advancing gestational age.

1139 RESPIRATORY DISTRESS SYNDROME IN ≤32-WEEK GESTATION PREMATURES: NEUROBEHAVIORAL PROFILE. Jacqueline R Farwell and David T Scott (Spon. by Joseph B Warshaw). Yale Univ, School of Medicine, Dept of Pediatrics, New Haven, CT.

To investigate the effects of respiratory distress syndrome (RDS) on the neurobehavioral function of premature infants, 26 infants of ≤32 weeks gestation were evaluated by a modification of the Brazelton Neonatal Scale within the first four days of life. 17 were infants with uncomplicated prematurity (PBLCs). 9 had RDS, uncomplicated by depression at birth, sepsis, seizures, hyperbilirubinemia, or intracranial hemorrhage. When examined, 3 were on respirators and 3 had nasal prongs in place. Infants with RDS showed decreased ability to adapt to repeated stimuli and to attend to and orient toward light, sound, face, and voice, compared to PBLCs. Infants with RDS were also less alert. A higher level of irritability and a higher peak of arousal were noted in infants with RDS. The two groups did not differ in level of activity or in response to a cloth on the face. Grasp reflexes were diminished in infants with RDS, but Moro and deep tendon reflexes were not. Resting muscle tone was similar in the two groups. Since in our study tone, reflexes, and activity were not affected by even severe uncomplicated RDS, the development of hypotonia, hyporeflexia, or hypoactivity in an infant with RDS therefore points toward a complication such as sepsis or intracranial hemorrhage. However, our results suggest that cortical functions such as adaptation, orientation, and alertness are impaired in infants with RDS.

1140 CHILDHOOD CENTRAL NERVOUS SYSTEM TUMORS AND FAMILIAL CANCER. J R Farwell, J W Meigs, L D Marrett, & J T Flannery. (Spon. by Howard A Pearson.) Yale Univ. School of Medicine, Depts of Pediatrics & Epidemiology, New Haven.

The occurrence of cancer among parents, siblings, and offspring of 725 children with central nervous system (CNS) tumors was compared with occurrence of cancer among such relatives of 360 birth certificate controls, using the Connecticut Tumor Registry. 84 (12%) of the children with CNS tumors (cases), none of whom had neurofibromatosis, and 42 controls (12%) had at least one first-degree relative with cancer. However, certain types of cancer were much more frequent among relatives of cases. There were 9 CNS tumors and 3 other nervous system tumors among relatives of cases, but no nervous system tumors among relatives of controls. Leukemia occurred in 6 relatives of cases but in no relatives of controls. Disproportionately many patients with cancer of the bladder, ovaries, and uterus were similarly found among relatives of cases. 12 siblings of cases developed cancer before age 20, while only 1 sibling of a control did. In each of 3 families, 3 siblings developed various cancers, confirming previously reported associations of CNS tumors, leukemia, and colonic cancer. Children with more malignant tumors (eg, medulloblastoma) were more likely to have a relative with a CNS tumor than children with better-differentiated tumors (eg, astrocytoma). These data suggest that there is a familial occurrence of CNS tumors, and that the occurrence of a CNS tumor in a child may be associated with an increased incidence of CNS tumors, leukemia, and childhood tumors in general among his relatives.