**1582** AUTONOMIC DYSFUNCTION ASSOCIATED WITH SECRETORY DIARRHEA: A NEW SYNDROME. Paul E. Hyman, Melvin B. Heyman, Edward J. Feldman, William E. Berquist, and Marvin E. Ament, UCLA Medical Center, Department of Pediatrics, Los Angeles.

A 1 year old male had features of autonomic dysfunction including lack of axon flare in response to intradermal histamine, abnormal pupillary response to conjunctival instillation of methacholine, absent fungiform papillae of the tongue, lack of overflow tears, decreased response to deep pain, and a history of recurrent aspiration pneumonias and failure to thrive. The patient differed from individuals with familial dysautonomia (FD) in that 1) he was not of Jewish heritage, 2) sweating was absent, 3) corneal reflexes were present, 4) temperature and vasomotor instability were absent. The patient differed from individuals with congenital sensory neuropathy with anhidrosis (CSN) in that 1) overflow tearing and fungiform papillae of the tongue were absent, 2) deep tendon reflexes were present, and 3) methacholine caused prompt pupillary miosis. True secretory diarrhea is a feature of neither FD or CSN, but was present in our patient. During a 9-day fast stool volume averaged 20 gm/kg/day. Stool electrolytes were Na 111 mEq/L, K 26 mEq/L, Cl 55 mEq/L. There was no evidence of malabsorption. Enteropathic bacteria and parasites were absent from the stool. No histologic abnormalities were evident in biopsies of the small bowel or rectum. The diarrhea improved during treatment with tincture of opium. Studies in animals and man have demonstrated that the autonomic nervous system contributes to the control of secretion from the small bowel and colon. This is the first report of a syndrome of a congenital autonomic dysfunction associated with secretory diarrhea.

**1583** PROGNOSTIC VALUE OF PROMINENT SUBARACHNOID SPACES IN MACROCEPHALY. Suresh Kotagal, St. Louis University, Cardinal Glennon Memorial Hospital for Children, Department of Pediatrics & Neurology, St. Louis, MO.

Diameters of the extracerebral frontal space (hereinafter referred to as the frontal subarachnoid space) were measured on the CAT scans of 80 children, their ages ranging from 0-24 months. The maximal diameters from the outer surface of the cortex to the inner table of the skull were measured and corrected, using the minification factor for the Siemens SIRETOM Scanner.

Ten of these infants (8 males, 2 females) underwent CAT scans for evaluation of macrocephaly with head circumference above the 98th percentile. All had normal CAT scans except for a mean frontal subarachnoid space diameter of 2.24 mm. This differed significantly from 0.69 mm, the mean for 70 other children with miscellaneous neurologic disorders ($p<0.05$). Specifically, the frontal subarachnoid space diameter in children with macrocephaly and normal neurologic examinations differed significantly ($p<.05$) from the measurement in 22 children with hydrocephalus (mean 0.77 mm), 7 children with idiopathic seizures (mean 0.45) and 6 children with spastic diplegia (mean 0.27). Six of these 10 infants upon follow up for a mean period of 13.6 months, continued to demonstrate normal development.

Macrocephaly in infants, when associated with prominent frontal subarachnoid space may indicate an increased cerebrospinal pool and a relatively benign prognosis.

**1584** INTRAVENOUS (I.V.) FLUID ADMINISTRATION RATES: POSSIBLE FACTOR IN CAUSATION OF CEREBRAL INTRAVENTRICULAR HEMORRHAGE (IVH) IN NEONATES? Venkatesan Krishnan, Malini Satish, Gerald Katzman, Jose Urrutia, Irwin Weinfeld, Sidney Kripke, P.L.S. Amma. (Spon. by M.G. Robinson) Medical College of Ohio, The Toledo Hospital, Dept. of Ped., Toledo, Ohio.

Hourly I.V. fluid administration rates (including all medication, catheter flush, etc.) were analyzed in all 19 neonates admitted between 1/80-10/80 who had CT Scans done in the first 16 days of life to confirm or R/O IVH. 12 infants had analyses of IV rates until suspicion of IVH by bloody CSF; confirmed by CT Scan. The 7 neonates who had normal CT scans were used as controls. (3 were twin siblings, same sex, of neonates with IVH). These controls had analysis of IV rates for the first 120 hours of life. Birth weights; gestational ages; Apgar Scores at 1 and 5 minutes; initial mean blood pressure; and resuscitative measures at birth were similar in both groups.

| | IVH (12) | | | CONTROL (7) | | |
|---|---|---|---|---|---|---|
| | RANGE | MEAN | S.D. | RANGE | MEAN | S.D. |
| B.W. (Gms.) | 720-1360 | 1064 | 226 | 920-1490 | 1117 | 183 |
| Hrs. of Study | 24-240 | 90 | 75.7 | -------- | 120 | --- |
| # Episodes of I.V. Fluids >9 ml/kg/hr. | 1-12 | 5.4 | 3.9 | 0-11 | 3.4 | 3.5 |

There was a significant correlation ($p<0.05$ Mann Whitney U Test) between the # of episodes of IV fluid rates over 9 ml/kg/hr and occurrence of IVH. It is conceivable that bolus administrations of IV fluids contribute to causation of IVH in preterm neonates.

**1585** AUTOREGULATION OF BRAIN BLOOD FLOW (BBF): REGIONAL DIFFERENCES IN REDUCED BBF WITH HYPOTENSION. A. Laptook, B.S. Stonestreet, W.Oh, Brown Univ. Program in Medicine, Women & Infants Hosp., Dept. of Ped., Providence, RI

The potential use of a piglet as a model for investigation of brain blood flow (BBF) was evaluated by assessing the presence of autoregulation in 11 spontaneously breathing newborn piglets. MABP was altered by phlebotomy. Within the normotensive range (MABP=51-105 mmHg) no significant change in BBF (measured by the microsphere technique)occurred ($r=0.03$) indicating the presence of autoregulation. During hypotension (MABP=15-50 mmHg), a pressure passive relation exists between BBF and MABP ($r=0.59$). However, since the piglets breathed spontaneously and hyperventilated during hypotension, both the MABP and $PaCO_2$ fell and both correlated with BBF during hypotension. Thus, it is uncertain which factor is responsible for the loss of autoregulation. The blood flow (BF) to three specific regions of the brain (cerebrum, cerebellum, brainstem) and MABP also showed no correlation during normotension. During hypotension each region demonstrates significant pressure passive relationships, but the reduction in BF is most pronounced in the cerebrum, less in the cerebellum, and least in the brainstem, (M±SE, 58±7%, 34±12%, 27±11% reduction from control respectively, $p<.05$). The study indicates that a newborn piglet may serve as an appropriate model for the study of brain hemodynamics particularly with regard to autoregulation. Furthermore, during hypotension, preferential protection of vital regions of the brain (cerebellum and brainstem) occur which may have important implication in interpreting the effect of hypotension on the newborn central nervous system.

**1586** EFFECTS OF DOPAMINE ON CEREBRAL BLOOD FLOW (CBF) AND OXYGEN METABOLISM ($CMRO_2$) IN DOGS UNDER THE INFLUENCE OF E COLI ENDOTOXIN (ET). Lars E. Larsson, Barbro Ekström-Jodal and Egil Häggendal. Spon. by Hakan Sundell. Gothenburg University, Dept of Anesthesiology, Östra Sjukhuset, Gothenburg, Sweden.

In normocapnic dogs ET causes a decreased CBF, an increased $CMRO_2$ and decreased cardiac output (CO) and mean aortic pressure (MAP). With a defective blood-brain barrier during endotoxemia a direct influence of monoamines on the cerebral metabolism is possible. Dopamine (DA), a neurotransmittor, was used to test this hypothesis.

In four dogs ET 1.0-1.5 mg/kg was injected over five minutes. CO and MAP were followed. CBF was measured using a radioactive gas elimination method ($Xenon^{133}$). $CMRO_2$ was calculated from CBF and the arteriovenous oxygen difference. Venous blood was sampled from the superior sagittal sinus. During the established shock phase dopamine HCl 10 µg/kg min was infused. Effects after ET and ET+DA are compared.

| | CBF ml/100 gr min | $CMRO_2$ ml/100 gr min | CO ml/kg min | MAP mm Hg |
|---|---|---|---|---|
| Control | 43±4 | 4.6±0.2 | 161±4 | 127±9 |
| E T | 37±3 | 4.9±0.2 | 91±19 | 87±18 |
| ET+DA | 49±5* | 6.2±0.4* | 106±24* | 74±13 |

*=$p<0.05$ (Student's paired t-test). Mean values±SEM.

The same dose of DA without ET causes an increased CBF without affecting $CMRO_2$. These results indicate a direct effect of DA on cerebral metabolism. The effects of ET can thus be influenced by treatments that normally would not affect the brain metabolism.

**1587** THIN RIBS AND NEONATAL MUSCULAR DISORDERS. Raul A. Lazarte, Jacinto A. Hernandez, Daniel M. Hall, and Frank L. Quattromani (Spon. by James K. Todd), The Children's Hospital, Department of Perinatology, Denver, Colorado.

Newborn infants suffering from severe generalized neuromuscular disorders at birth usually represent a diagnostic dilemma for the clinician. Not uncommonly they are given low Apgar scores and develop early signs of respiratory failure requiring ventilatory assistance. Frequently, the floppiness and muscle weakness is attributed initially to birth asphyxia.

In this study we report our experience with 7 neonates diagnosed as having generalized muscle disorder - Congenital Muscular Dystrophy (3), Congenital Myotonic Dystrophy (1), Myotubular Myopathy (2), Undefined Congenital Myopathy (1) - in whom the chest x-ray revealed markedly thin ribs as a striking and consistent finding. This finding was not present in term infants with severe hypotonia as the result of acute hypoxia at birth. Normal values for rib width were established for infants of different birth weights in an attempt to provide an objective means of defining thin ribs.

Based on the consistency of this finding in severe cases of neuromuscular disorder we consider this as a useful radiological sign in differentiating muscle disease from other causes of neonatal hyptonia and respiratory insufficiency while waiting for more definitive studies.