1714 NEUROLOGIC INVESTIGATIONS IN PATIENTS WITH FRAGILE X SYNDROME. Siegfried M. Pueschel and Pasquale V. Finelli. Brown University Program in Medicine, Rhode Island Hospital, Department of Pediatrics, Providence, Rhode Island, and Division of Neurology, Department of Medicine, The Memorial Hospital, Pawtucket, Rhode Island.

We have identified 22 patients with the fragile X syndrome. The majority of these patients have come to our attention because several family members had significant mental retardation and/or dysmorphic facial features suggestive of fragile X syndrome. All patients underwent extensive examinations including determination of phenotypic abnormalities, measurement of testicular size and a thorough neurologic assessment. The Stanford-Binet intelligence test was used to assess their intellectual functioning. One hundred metaphase spreads of each patient were examined for fragile X chromosomes.

The results of this investigation revealed that 82% of patients displayed brisk deep tendon reflexes and 41% had extensor plantar responses. Gaze eversion was observed in 77% of patients. Almost half of the subjects (47%) exhibited hyperactive behavior and had a stooped posture and gait. Incoordination was noted in 59% of the patients and blepharospasm was present in 38%. All patients were mentally retarded, ranging from mild to profound retardation. The subjects' language development was also retarded and they had marked difficulties with syntax, intonation and fluency. All patients had macro-orchidism. Their main dysmorphic features included coarse facies, large ears, prominent glabella, synophrys, large nose, prognathism, and abnormal dermatoglypics.

†1715 CERVICAL CORD COMPRESSION IN ACHONDROPLASIA. C Reid, H Wang, R McPherson, B Maria, C Francomano, J Phillips, III, A Rosenbaum and R Pyeritz, Johns Hopkins Univ Sch of Med, Depts of Peds, Radiol, Anesth and Neurol, Baltimore.

Cervicomedullary cord compression due to an abnormally small foramen magnum (FM) has been thought to be rare in achondroplasia during childhood. Since autopsy-proven cases of cord compression have had apnea or other serious respiratory complications in the absence of specific neurologic signs, evaluation of patients with respiratory symptoms might reveal occult cord compression early in its course. With this in mind, we evaluated 10 children with achondroplasia (6 males, 4 females; ages 4 mos - 6yrs) for potential cervical cord compression; using CT, somatosensory evoked potentials (SEP) and brainstem auditory evoked responses (BAER). 2/10 patients were asymptomatic and 8/10 had respiratory problems. All 10 were hypotonic, but only 3 had focal neurologic findings. In 9/10, sagittal diameter of the FM was > 3 S.D. smaller than normal and an abnormal keyhole shape was seen. Moreover, in 4/10, sagittal CT reconstructions showed virtually no subarachnoid space posterior to the cord at the FM. The same 4/10 had prolonged latency on SEP; when contrasted with normal BAER in 10/10, this suggested cord dysfunction at the level of the FM. Metrizamide CT myelography confirmed and delineated the extent of cord compression in these 4. All 4 had respiratory abnormalities but only 2 had specific neurologic findings. This suggests that cervicomedullary cord compression may occur without neurologic findings and lead to respiratory problems in a significant number of achondroplastic children.

1716 THE USE OF ANTERIOR CEREBRAL ARTERY PULSATILITY INDEX TO PREDICT NEUROLOGIC OUTCOME. Rita Saldanha, Grant Somes, John Wimmer, Stephen Engelke, Lillian Ruckman and Arthur Kopelman (Spon. by Jean Kenny). East Carolina Univ. Med. Sch., Pitt Mem. Hosp., Dept. of Peds., Greenville, NC.

Decreased blood flow velocity (increased pulsatility index, PI) in the anterior cerebral arteries (ACA) has been reported in preterm infants with intraventricular hemorrhage. From Feb. 1982 to Aug. 1983, we prospectively evaluated PI, cranial ultrasound and neurologic status (Sarnat Score), in 60 infants ≤32 weeks gestation on days 1, 3 and 7 and then weekly until discharge, to determine their correlation with PI. PI was calculated from the tracings obtained by doppler ultrasound by the method described by Bada et al. Survival, neurologic status (Sarnat Score), cranial ultrasound, occurrence of seizures, and discharge neurologic examination (Parmalee exam) were assessed.

Mean B.Wt. was 1151 grams (640-1750) and mean GA was 30.1 weeks (26-32). There were 31 males and 29 females. Mean Apgar Score at 1 minute was 4.7 and at 5 minutes was 6.6. Forty-two of the infants were inborn.

The maximum PI recorded on day 1 correlated negatively with B.Wt. ($p<0.05$) and positively with mortality ($p<0.06$), occurrence of seizures ($p<0.05$) and development of hydrocephalus ($p<0.06$). There was no correlation between PI done at any time and GA, neurologic status (Sarnat Score), cranial ultrasound or discharge neurologic examination.

Decreased cerebral blood flow velocity (high PI) is correlated with mortality, occurrence of seizures and later development of hydrocephalus. Follow-up of these infants is pending.

1717 PERINATAL ASPHYXIA AND PLASMA β ENDORPHINS. Koravangattu Sankaran, K. Wayne Hindmarsh and Valerie Watson, Perinatal Research Laboratory, Department of Pediatrics and College of Pharmacy, University of Saskatchewan, Saskatoon.

In an attempt to determine whether plasma β endorphin (β-ED) concentrations correlate with perinatal asphyxia, measurements were made in three groups of term infants. Group 1 (control) consisted of eight infants with a mean (± SE) gestation of 38.6 ± 0.4 weeks, a mean birthweight of 3420 ± 150 g, and a mean postnatal age of 1.4 ± 0.7 days. Group 2 consisted of ten infants with perinatal asphyxia with a mean gestational age, birthweight and postnatal age of 40.1 ± 0.5 weeks, 3310 ± 80 g and 3.9 ± 1.1 days, and Group 3 included six infants with a mean gestational age, birthweight and postnatal age of 40.4 ± 1 week, 3650 ± 310 g, and 2.8 ± 1 day, respectively. Group 2 and 3 infants suffered clinical and neurological evidence of hypoxic-ischemic brain injury, however, infants in Group 2 suffered meconium aspiration, persistent fetal circulation with on-going hypoxemia as measured by transcutaneous or umbilical arterial oxygen monitoring. Group 3 infants were normoxemic after resuscitation. The mean (± SE) plasma β-ED concentrations were 19 ± 2.7 pg/ml, 103 ± 35.7 pg/ml and 25 ± 4.5 pg/ml in Groups 1, 2 and 3, respectively. A significant elevation of plasma β-ED concentration was observed in Group 2 when compared to Groups 1 and 3. The association of increased plasma β-ED concentration in infants with hypoxic-ischemic encephalopathy associated with on-going hypoxemia suggests that hypoxemia may act as a strong stimulus for plasma β-ED release in term infants.

1718 CAN ULTRASOUND PREDICT DEVELOPMENTAL OUTCOME IN INFANTS WITH GRADE III AND IV IVH? P. Sasidharan, E. Marquez, E. Dizon and C. Sridhar, Porter Memorial Hospital, Valparaiso, IN (Spon. by R. Schreiner)

We studied the usefulness of realtime ultrasound scan (US) in predicting developmental outcome in infants with Grade III and IV IVH. US were done prior to discharge from NICU in the coronal plane only. Infants were classified into 5 groups depending on size of ventricles, visible dilated temporal horns and asymmetry in size of ventricles or presence of porencephalic cysts. Developmental assessment was made using Denver Developmental and Bayley Scales of Infant Assessment. 36 infants were studied.

	A	B	C	D	E
#n	1	15	3	8	9
GA(wks)	29	29.7±3.3	28.6±3.05	28.8±5.2	28.6±2.34
PN Age(mos)	11.5	19.5±8	11.6±6	13.7±3.4	15±9.5
DQ	100	99.3±8.2	85.6±12.9	57.1±22.7	58.6±23.9

The mean birthweight of the infants is 1240±498gms. and gestational age is 29.1 weeks (range 24 to 40). The mean postnatal age at follow-up is 16.25±7.7 months. All infants with a DQ > 85 are considered normal, 75-85 suspicious and < 75 abnormal. Statistically significant differences are noted in the DQ between infants in the classes of A and B to infants in the classes of D and F ($P = <.001$). Our results indicate that US scans done prior to discharge are useful in predicting developmental outcome in infants with severe IVH using our classification.

1719 A WEIGHTED APGAR SCORE FOR LOW-BIRTHWEIGHT INFANTS. David T Scott & Laura R Ment (Spon. by Norman Siegel). Yale Sch of Med, Dept of Ped, New Haven, CT.

The Apgar score is a simple arithmetic sum of the five areas of neonatal functioning (heart rate, respirations, tone, irritability, and color). However, some of these functions may have more prognostic significance than others and should therefore have greater weight in deriving a prognostic summary score.

In order to investigate the relative significance of the five Apgar constituents, we retrospectively reviewed the delivery records of the preterm infants from our Newborn Follow-Up Program. We then employed linear regression methods to assess the extent to which the Apgar scores and their constituents could predict outcome as measured by the Bayley Scales of Infant Development.

We first evaluated the prognostic significance of the traditional one- and five-minute Apgar scores. Each of these scores accounted for less than 1% of the variation in the Bayley Mental Index at 6 months corrected age; the two summary scores together accounted for about 1.5%. In contrast, the five-minute heart-rate constituent score, taken alone, accounted for 14% of the variation of the Bayley Index. The addition of other Apgar constituents to the regression model increased the proportion of the variance that appeared to be accounted for. Preliminary cross-validation analyses suggest that somewhat less of the variance will be accounted for in separate samples--but still considerably more than that accounted for by the traditional summary scores. We conclude that the traditional Apgar score may mean less than the array of its constituents.