1153 ELEVATED PLASMA GLUTAMATE LEVELS WITHOUT HYPOTHALAMIC LESIONS. Shirley M. Mueller, Lewis D. Stegink and W. Ann Reynolds (Spon. by L. J. Filer, Jr.) University of Iowa College of Medicine, Dept. Pediatrics, Iowa City, and University of Illinois Medical Center, Dept. Anatomy, Chicago.

Three male infants have been reported with elevated plasma glutamate levels, mental retardation and brain atrophy. We have observed a fourth male infant with elevated plasma glutamate (10 X normal). CSF glutamate was normal. The infant was hypotonic without Moro, suck or grasp. His clinical course was complicated by seizures (not the "shuddering syndrome" type), intolerance to feedings, hypocalcemia, and hypernatremia. EEG was abnormal. Chromosomal analysis was normal. At death (53 days), moderately severe atrophy of the cerebral hemispheres with greater atrophy of the cerebellum and mildly dilated ventricles were noted. The hypothalamus was embedded in celloidin and serially sectioned at 40 u. Cellular morphology was normal throughout; no evidence of dendritic swelling, pyknotic nuclei or other manifestations of the lesion induced by glutamate in the rodent were noted. The elevated plasma glutamate may represent a rare enzymatic abnormality of glutamate metabolism either directly or indirectly associated with mental retardation and cerebral atrophy. However, the infant's chronically elevated plasma glutamate levels were not associated with the type of gross hypothalamic lesions reported in the infant rodent with high glutamate levels. An oral glutamate challenge to the child's parents was given. The father showed an abnormal tolerance curve and reported the symptoms of the Chinese Restaurant Syndrome.

1154 THE 1000 GRAM INFANT AT 18 MONTHS: Karen E. Pape, Pamela M. Fitzhardinge, Univ. of Toronto, Research Inst., Hosp. for Sick Children, Toronto, Canada.

During 1974, 97 infants with birth weights $\leq$1001 g. were referred to our neonatal intensive care unit from outlying hospitals. Forty-five (46%) survived. Forty-two (93%), 14 of whom were small for gestational age, have been studied prospectively and the results analyzed at age 18 months post-term. Eleven of the 42 had major developmental sequelae. Nine had a developmental index (D.I.) less than 70; 4 of these and 2 others had major neurological defects (cerebral palsy, hydrocephaly, microcephaly). The median D.I. for these 11 with severe handicap was 64 mental, 57 motor. The remaining 31 infants (74%) were free of major handicap although 19 had evidence of minor dystonia or speech delay. The median D.I. for these 31 children was 98 mental, 90 motor. The average D.I. for the entire sample was 89$\pm$25 for mental, 81$\pm$22 for psychomotor.

Retrolental fibroplasia was diagnosed in 6/42; 3 had less than 10% vision. None of the survivors had impaired hearing. Wilson-Mikity Syndrome or bronchopulmonary dysplasia occurred in 7; 4 were still symptomatic.

Major developmental handicap at follow-up was most closely related to the complication of severe intrauterine growth retardation ($p < 0.005$) or to the occurrence of neonatal intracranial hemorrhage or seizures ($p < 0.005$).

1155 CEREBRAL INTRAVENTRICULAR HEMORRHAGE (CVH) IN INFANTS $\leq$ 1500 GRAMS. Lu-Ann Papile, Jerome Burstein, Rochelle Burstein. (Sponsored by Robert Greenberg) UNM School of Medicine, Department of Pediatrics, Albuquerque, New Mexico.

A prospective study using Computed Tomography (CT) was initiated to determine the incidence of sub-ependymal (SEH) and intraventricular hemorrhage (IVH) in infants $\leq$1500 grams. All 38 infants $\leq$1500 grams who were admitted to the Newborn Intensive Care Unit during a five month period have been evaluated.

The initial CT was performed between the third and seventh postnatal day. If a CVH was noted, follow-up CT was done at one and three weeks after the initial CT. Nine of the 23 infants who survived had SEH and/or IVH present on the initial CT. Only one of these nine infants was suspected of having a CVH on clinical findings. Of these nine, five who had a small SEH and/or IVH with normal ventricular size, had complete resolution of the hemorrhage at three weeks. The other four infants who demonstrated some degree of ventricular dilatation with the hemorrhage on the initial CT, required medical intervention to prevent the development of clinical hydrocephalus.

This study documents, for the first time, the relatively high incidence of CVH in infants $\leq$1500 grams and indicates that there are at least two types of CVH: 1) CVH with no ventricular dilatation which resolves spontaneously and 2) CVH with ventricular dilatation which requires medical intervention to prevent the development of clinical hydrocephalus.

1156 MUSCLE ULTRASTRUCTURE IN REYE'S SYNDROME (RS): EVIDENCE FOR A MYOPATHY. J.C. Partin, J.S. Partin*, and W.K. Schubert. Department of Pediatrics, University of Cincinnati, Cincinnati, Ohio.

Biochemical studies have shown elevation of muscle creatine phosphokinase (Roe et al., Ped. 55:119, 1975) and excessive urinary nitrogen losses up to 0.64 g/kg/day (Snodgrass and DeLong, N. Eng. J. Med. 294:855, 1976) in RS, suggesting skeletal muscle injury. We report electronmicroscopy of muscle biopsies obtained from 15 RS cases on the day of admission and at followup 4 days to 23 weeks later. A myopathy of variable severity was present in each case acutely. In severely affected specimens about 1/40 muscle cells were necrotic with dissolution of the sarcolemma, loss of striations and dispersion of myofilaments, producing a homogenous matrix which contained vesicular fragments of sarcoplasmic reticulum, autophagic vacuoles and spherical mitochondria with expanded matrix and frequently ruptured outer membranes. Surrounding muscle cells were abnormal, displaying glycogen loss, ribosome disorganization, dilation of sarcoplasmic reticulum and intramyofibrillar edema. There were 2 populations of mitochondria in muscle cells: Individual mitochondria demonstrated extreme matrix expansion suggestive of that seen in brain and liver in RS, but the majority of muscle cell mitochondria demonstrated only slight matrix expansion. Muscle cell triglyceride was increased and many cells contained myelin figures. Inflammation was absent but influenza virus was isolated from 3 of 5 biopsies in which cultivation was attempted. These studies confirm the existence of a myopathy in RS. The relationship between the myopathy of RS and post-influenzal myopathy of childhood should be investigated.

1157 STROKE SYNDROME IN SICKLE CELL DISEASE (SCD) Darleen R. Powars, Charles H. Pegelow, Brian J. Wilson, John P. Allen, (Spon. by Paul F. Wehrle). University of Southern California School of Medicine, Los Angeles County-University of Southern California Medical Center, Department of Pediatrics, Los Angeles, California.

To delineate the natural progression of cerebral vascular accidents in SCD and to determine the resultant structural and functional defects we investigated 29 patients (27 SS, 2 SC) who sustained strokes. Subarachnoid or intra-cerebral hemorrhage and bone marrow embolism occurred in 9 patients ages 14-35 years. Cerebral infarction occurred in 20 (19 SS, 1 SC) with a modal age of onset of 5 years. We were able to evaluate 14 long term survivors who were observed for a mean period of 9.4 years following the initial stroke. 10 (71%) had 1 or more recurrences with a mean interstroke interval of 28 months. Residual functional impairment was noted in all 14. Computerized axial tomography on 9 showed areas of low density and ventricular dilatation indicating structural brain damage. These patients were not treated with a transfusion program. Their clinical course indicates that evaluation of such therapy requires long periods of observation.

1158 FETAL INTRACRANIAL HEMORRHAGE(ICH): T.N.K.Raju(by invitation) O.T.Bailey(by invitation) and D.Vidyasagar, Dept. Ped. & Neuropath. Uni. Ill. ALSM, Chicago, IL

ICH in the newborn is usually recognized as a postnatal event. Prematurity, respiratory distress, hyperosmolarity, hypoxia and acidosis are known predisposing factors. Intrauterine occurence of ICH is not widely recognized. Because of paucity of information in this regard autopsy records of 60 consecutive still births were reviewed. Detailed neuropathological study was possible in only 12 instances because of excessive maceration and autolysis in the rest. 4/12 had massive ICH in varying combinations and degree at following sites: subarachnoid(SA), germinal plate(GP), choroid plexus(CP), & intraventricle(IV). Clinical & autopsy date of these 4 cases is shown below.

G.A.(Wks):	Wt.(gm):	Prenatal history:	Site of ICH:
40	2400	Fetal death: footling & cord presentation.	Massive Sub ependend.
42	3425	Fetal death before labor	CP
31	1300	Maternal Anticonvulscents: placenta previa: 20% abruption.	Massive SA, IV, GP
28	610	Prem. rupture of membrane: beta strept. infection.	Massive SA, IV, GP

In 3/4 a significant fetal result could be identified. All had normal labor: one was a product of C-section and others were delivered non-operatively. We conclude: (1): multitudes of prenatal factors can cause fetal ICH and hence prenatal etiology must be considered in neonates with ICH. (2): Detailed neuropathological study of stillborn infant's brain should be attempted.