1159 LONG-TERM SEQUELAE IN REYE'S SYNDROME Irwin Redlener, Jose Leon, John MacDonald(intro. by W.W.Cleveland)Un. of Miami School of Medicine Depts. of Ped. and Neuro. Miami, Florida

Sixteen cases of Reye's Syndrome were seen in this institution from 2/74 to 9/76. Ages ranged from 5 mos. to 14 years; 3 pts. were black. The diagnosis was made clinically and with supporting lab. data(all pts.)and by liver tissue confirmation(11 pts.) The length of followup ranged from 3 to 34 months. Four deaths occured (25%). Eight children(50%)had significant neurologic sequelae leaving only 4 of the children(25%)without mortality or apparent morbidity. Of the six children under 18 mos., there were no deaths although all but one had significant sequelae. Major problems in this group include varying degrees of psychomotor retardation and seizures. Older children exhibited significant long-term problems in speech, behavior and learning. In children older than 3 years, mortality was associated with Stage IV coma(Hutenlocher); moderate sequelae were seen with Stage III although two of these children recovered without apparent residuals. Children less than 18 mos. had very serious neurologic sequelae even though they did not progress beyond Stage III coma. Retrospective review of hospital charts revealed no apparent correlation between outcome and specific laboratory abnormalities or theraputic modalities in our patients with Reye's Syndrome. Conclusion: poor outcome in both mortality as well as long-term sequelae in Reye's Syndrome is related to severity of coma as well as age, with younger infants being particularly prone to the development of significant neurologic deficits.

1160 NERVOUS SYSTEM LESIONS IN INFANTS OF OPIATE DEPENDENT MOTHERS. Lucy B. Rorke, Dian S. Reeser, Loretta P. Finnegan (Spon. by M. Delivoria-Papadopoulos), Univ. of Pa. Sch. of Med., Thomas Jefferson Univ.,Phila. Gen. Hosp. (PGH), Dept. of Pediatrics, Philadelphia, Pa.

Widespread illegal use of heroin and legal distribution of methadone have had vast effects on our society, and particularly on the pregnant woman and her newborn infant. Since 1969, 330 infants born of drug dependent women at PGH have been studied in conjunction with a comprehensive program of medical, addictive and social services. Neonatal mortality rate in this population was 5%; the highest rate was amongst those whose mothers received inadequate or no prenatal and addictive care (8%, 6%). Detailed neuropathological studies of ten of these infants, nine of whom died during the neonatal period, revealed eight categories of lesions, three of which were thought to bear some relationship to the maternal drug dependence period. These included: gliosis (5/10), foci of old infarction (4/10) and developmental brain retardation (3/10). Only minor microscopic brain malformations were found in three cases. Other lesions identified included those common to high risk neonates: germinal plate hemorrhages (7/10), acute brain necrosis with and without hemorrhage (5/10),germinal plate cysts (4/10) and focal subarachnoid hemorrhages (3/10). Two isolated examples of unique lesions consisting of vascular proliferation and arachnoidal proliferation (1/10), and a posterior poliomyelitis (1/10) completed the spectrum of abnormalities.

1161 NEUROBEHAVIORAL ASSESSMENT OF WELL & ILL PREMATURES OF ≤32 WEEKS GESTATION. David T Scott and Jacqueline R Farwell (Spon. by Joseph B Warshaw) Yale Univ. School of Medicine, Depts of Pediatrics & Psychology, New Haven.

The Brazelton Neonatal Behavioral Assessment Scale, adapted for very premature infants, was used to assess the neurobehavioral status of 46 infants of ≤32 weeks gestation at age ≤3 days. There were 14 cases of uncomplicated prematurity (PBLC). The remaining 32 sustained complications such as respiratory distress syndrome, intracranial hemorrhage, hyperbilirubinemia, or sepsis; of these 13 died. The exam comprised standard reflex items, asessments of tone, color, posture, and Brazelton's behavioral items, eg, orientation and adaptation to repeated stimuli. PBLCs differed from the ill survivor group by displaying greater ability to adapt to repeated stimuli. PBLCs showed better orientation to, and following of, rattle, face, and voice. Their level of spontaneous and elicited activity was higher than that of ill survivors. PBLCs demonstrated more sustained alertness and were more easily brought to an alert state. Nonsurvivors attained lowest scores on items assessing orientation to light, rattle, and face. Nonsurvivors also exhibited lowest scores on the activity and alertness scales, were more difficult to bring to an alert state, were relatively hypoirritable, and tended to achieve a lower peak of arousal than the surviving prematures. Gross muscle tone did not distinguish reliably between the PBLCs and the nonsurvivor group. These results help define normal neurobehavioral function in very premature infants and thus identify abnormal findings with implications for clinical management.

1162 ELEVATED CONCENTRATIONS OF CSF HOMOVANILLIC ACID IN REYE SYNDROME-A POSSIBLE RELATION TO CEREBRAL HYPOPERFUSION. Bennett A. Shaywitz, Joan L. Venes, Donald J. Cohen, Malcolm B. Bowers, Jr., Dept. of Ped., Neurosurg., and Psychiatry, Yale Univ. Sch. of Med., New Haven.

Recent investigations suggest a relationship between brain dopamine release and subsequent hypoperfusion. Such an association has important therapeutic implications since abundant pharmacological agents are available that could be utilized to modify central monoaminergic mechanisms. We have examined cerebral ventricular fluid concentrations of homovanillic acid(HVA) and 5-hydroxyindoleacetic acid (5-HIAA), the principal metabolites of dopamine and serotonin respectively,in 7 children with Reye syndrome. In these children, CSF was obtained during continuous intracranial pressure (ICP) monitoring and compared with ventricular CSF from 6 children with communicating hydrocephalus. Maximum HVA (ng/ml, mean ± SEM) was significantly elevated in Reye syndrome averaging 1140 ± 313 compared to 267 ± 64.5 in controls ($p < 0.005$) while 5-HIAA was similar, averaging 232 ± 32.5 and 187 ± 33.9 in Reye syndrome and controls respectively ($p > 0.10$). Peak HVA concentration occurred 1-2 days after the maximum blood NH_3; however, there was no significant correlation between the maximum CSF concentration of either HVA or 5-HIAA and maximum blood NH_3. ICP was increased in all children with Reye syndrome but there was no consistent pattern between HVA concentration and degree of ICP elevation. Our findings suggest a sequence of dopamine release (as reflected by elevations in HVA), cerebral vasoconstriction, and a resultant hypoperfusion in Reye syndrome.

1163 CONGENITAL CYTOMEGALOVIRUS INFECTION (C-CMV) A CAUSE OF SENSORINEURAL HEARING LOSS (SNHL). Sergio Stagno, David W. Reynolds, Rufino Ermocilla, Charles A. Alford, Univ. of Alabama in Birmingham, Department of Pediatrics, Birmingham, Alabama.

C-CMV is exceedingly common in our low socioeconomic population (2.3% of all live births). Over 90% of the cases however, are asymptomatic at birth. Nevertheless, the chronic nature of this infection may lead in time to low grade morbidity in particular SNHL. To define the incidence and significance of this process, the hearing acuity of 59 patients (mean age 54 months, range 2-102) born with C-CMV and of 41 uninfected controls (mean age 46 months, range 7-124) was tested by pure tone audiometry in a sound field (<2 years of age) and/or under earphones. Significant SNHL (50 decibel or over) occurred in 10 infected patients (17%) and had a progressive course in 2 of them. In contrast only 1 uninfected subject had SNHL of borderline significance at 12 months of age.

To better define the CMV-SNHL etiologic relationship we performed histopathologic studies of the inner ear of 3 patients who died with symptomatic C-CMV. Typical inclusion bodies were found in the stria vascularis and the Reissner's membranes. By means of an anticomplementary immunofluorescent assay, CMV antigens were in addition detected in the cells of the organ of Corti and the neurones of the spiral ganglia. These findings clearly suggest that CMV has the potential to infect cells of the most vital structures of the inner ear and thus give further credence to the pathogenic role of this virus in SNHL.

1164 FOLLOW-UP STUDIES OF INFANTS WITH BIRTH WEIGHT (B.WT.) ≤1500 G.: COMPARISON OF RESULTS WITH AND WITHOUT VENTILATORY THERAPY. Annabel Teberg, Paul Y.K. Wu, and Joan E. Hodgman. Univ. of So. Calif. Sch. of Med., LAC-USC Medical Center, Dept. of Pediatrics, Los Angeles.

Currently, there is a paucity of follow-up reports of infants of B.Wt. ≤1500 g and still less of these infants surviving neonatal mechanical ventilatory therapy (VT). The physical (Phy.), neurologic (Neuro.), developmental (Dev.), audiologic (Audio.) and ophthalmologic (Ophthal.) status of 74 infants (58 without respirators and 16 with respirators) of B.Wt. ≤1500 g, born in 1973, 1974 and 1975, were evaluated at ages ranging from 1 to 3 years. Incidence of abnormal findings are listed below:

	Phy.		Neuro.		Dev.		Audio.		Ophthal.	
	No.	%	No.	%	No.	%	No.	%	No.	%
Without VT	9	16	7	12	7	12	5	9	4	7
With VT	3	19	3	19	4	25	2	13	1	6

The data indicate that with the possible exception of developmental tests, infants of B. Wt. ≤1500 g, who were treated with ventilatory therapy had comparable follow-up results to those who did not require ventilatory therapy.