

1141 INTRAUTERINE GROWTH RETARDATION (IUGR): AN ADDED RISK TO THE PRETERM INFANT. Pamela M. Fitzhardinge, Karen E. Pape. Univ. of Toronto, Research Inst., Hosp. for Sick Children, Toronto, Canada.

In order to determine the dual effects of IUGR and premature birth, a prospective study of growth and development was done in a group of preterm small-for-gestational age infants (SGA). All were \leq 32 weeks gestation with birth weights more than 2 standard deviations below the mean for gestation. During 1974, 60 infants meeting these criteria were referred to our neonatal intensive care unit. Twenty-six of the 29 survivors were followed to age 18 months post-term. Mean birth weight was 988 ± 152 g, gestation 30.5 ± 1.7 weeks. Each infant was randomly paired with a surviving infant of appropriate weight for gestation (AGA) who matched for birth weight, sex, and type of ventilatory support (birth weight 999 ± 129 g, gestation 27.8 ± 1.5 weeks). The SGA infants were significantly smaller at 18 months post-term than the AGA controls with a mean difference in weight of 0.8 kg, length 2.2 cm, head circumference 1.3 cm ($p < 0.005$). Eight of the 26 SGA children had major neurological defects: 1 with microcephaly, 2-hydrocephaly, 4-cerebral palsy, 1-seizure disorder. None of the controls were so affected ($p < 0.005$). The Bayley developmental indices (corrected for gestation) were significantly lower than those of the controls: mental 88 ± 15 , mean difference 10 ($p < 0.02$); motor 79 ± 21 , mean difference 11 ($p < 0.05$).

The results suggest that the complication of IUGR significantly increases the risk of serious sequelae in the tiny premature infants.

1142 ACUTE ENCEPHALOPATHY WITH LIVER DYSFUNCTION, CHYLOUS ASCITES, AND CYTOMEGALOVIRUS INFECTION. A CASE REPORT. Donald E. Greydanus, Thomas F. Smith, and Gunnar B. Stickler. Mayo Clinic and Mayo Foundation, Rochester, Minnesota.

A patient with acute encephalopathy and liver dysfunction, followed by acute chylous ascites, is described. Titers of cytomegalovirus (CMV) increased from less than 1:2 to 1:32 during the illness, and CMV was isolated from the urine by use of preparations of WI-38 tissue culture. The CMV infection probably was not a hospital-acquired infection because the titers were significantly increasing when the abdominal distention, due to the chylous ascites, was seen. This is the first recorded case possibly linking CMV with acute encephalopathy and liver dysfunction. Thus, CMV should be included in the growing list of viruses that can produce this disorder. A liver biopsy was not done. The clinical findings suggested the diagnosis of a variant of Reye-Johnson syndrome. Also, the enlargement of the patient's abdominal lymph nodes, as seen on lymphangiogram, could have resulted in severe obstruction to the abdominal lymphatic flow, producing a transudation of lymph into the peritoneal cavity. This is the first recorded case of acute chylous ascites associated with mesenteric lymphadenitis in which the causative virus may have been isolated.

1143 CT SCANNING AND NEURO-DEGENERATIVE DISEASES IN CHILDREN. Charles A. Haenggell, E. Ralph Heinz, and Michael J. Painter. (Spon. by Thomas P. Foley, Jr.) Presbyterian University Hospital, Children's Hospital of Pittsburgh, Departments of Pediatrics, Neurology, and Neuroradiology, Pittsburgh.

CT scanning has for the first time given an objective means of assessing the changes in gray, and particularly white matter degenerative diseases in infants and children. Patients with adrenoleukodystrophy, metachromatic leukodystrophy, juvenile ceroid-lipofuscinosis, progressive multifocal leukoencephalopathy, and disseminated necrotizing leukoencephalopathy all have been studied. Some of these patients have had serial scanning to follow the evolution of the cerebral lesion. Correlation with enzymes, brain biopsy, skin biopsy, as well as clinical and electroencephalographic examinations have been made. CT scanning has given objective quantitative confirmation of white and gray matter degenerative changes, and in the appropriate clinical context, has allowed distinction of specific progressive neurodegenerative diseases in childhood. In several instances initial evaluation with CT scanning has determined the proper subsequent biopsy and enzyme studies.

1144 HYDROXYLYSINE DEFICIENT COLLAGEN IN A FLOPPY BABY. Peggy A. Hanson, Albany Medical College, Depts. of Peds. and Neurology, Albany, N.Y.; and Ruth S. Quinn and Stephen M. Krane, Harvard Medical School, Massachusetts General Hospital, Boston, Mass. (Introduced by I. Porter).

Five disorders of collagen have been characterized. In four of these disorders the patients have had symptoms of Ehlers Danlos. Three families with lysyl hydroxylase deficiency have previously been reported. We are reporting an additional case diagnosed in infancy. This infant was investigated as a floppy baby but nerve and muscle were normal. Lysyl hydroxylase activity measured in sonicates of dermal fibroblasts was 10% of normal.

	Patient	Controls
hydroxylysine/4-hydroxyproline	0.0054, 0.0062	0.048 \pm 0.005
hydroxylysine(residues/1000 AA)	0.37	4.1 \pm 0.4
hydroxylysine/lysine	0.010	0.15 \pm 0.02
4-hydroxyproline/proline	0.64	0.67 \pm 0.04

The deficiency of hydroxylysine results in abnormal cross-linking of collagen molecules which may account for the symptomatology. The patient we are reporting eventually developed symptoms of Ehlers Danlos. This type of Ehlers Danlos is an autosomal recessive disorder in which one might expect to make an intrauterine diagnosis.

Collagen abnormalities may thus be a cause of floppy baby syndrome and the delineation of a specific defect should be the goal of diagnostic studies.

1145 OTITIS MEDIA IN HYPERACTIVE CHILDREN WITH LEARNING DISORDERS. Leonard Hersher. (Spon. by William Bergstrom). Dept. of Pediatrics, SUNY, Upstate Medical Center, Syracuse, New York.

The frequency of otitis media among hyperactive children with learning disorders was compared with the frequency of otitis media in a large sample of normal children, using the same criteria for the diagnosis of otitis media in both groups. The groups were matched for social class and age when studied (7 to 13 years of age).

A significantly higher percentage of hyperactive children (54 percent) had more than six episodes of otitis media than was found in the normal group (15 percent). Thirty-six percent of hyperactive children had more than ten episodes, compared to five percent in the normal sample. There was no difference in the percentage of children with no episodes of otitis media (18 percent).

Several alternative hypotheses will be offered as possible mechanisms to account for the data.

1146 CACOGRAPHY IN THE MOTHERS OF CHILDREN WITH LEARNING DISORDERS. Leonard Hersher. (Spon. by William Bergstrom). Dept. of Pediatrics, SUNY, Upstate Medical Center, Syracuse, New York.

Mothers of children with learning disorders completed questionnaires concerning the histories and current problems of their children. All the children had average or above average IQ's, but below average academic achievement. Compared with a control group matched for years of education, the mothers of children with learning disorders made significantly more spelling errors per 100 words.

The differences in number of spelling errors between learning disorder and control groups was greater for mothers with twelve or less years of education than for mothers with more than twelve years of education.