1309 ULTRASOUND MEASUREMENTS (USM) OF BRAIN GROWTH IN INFANTS UNDER GOING ABSTINENCE, Matthew Pasto, Leonard Graziani, John Deiling, Saundra Ehrlich and Loretta Finnegan, Jefferson Medical College of the Thomas Jefferson University, Department of Pediatrics, Philadelphia,

Cross-sectional USM of brain structures were done to evaluate the brain growth of infants undergoing abstinence due to narcotic exposure in-utero in comparison to that of control infants. The groups were comparable in sex, race, socioeconomic status, birth weight, gestational age and Apgar scores. Mothers of the drug exposed infants had been maintained on methadone during pregnancy and many were known to use undetermined quantities of other psychoactive drugs. USM were performed at 72 hours, 1 month and 6 months after birth. Transaxial images were done at two levels: 1) the maximal circumference of the thalami and 2) the margins of the lateral ventricles where they parallel the midline. Intracranial hemidiameter (ICHD) was recorded at this level. Head circumferences (HC) were also measured. Results revealed significant differences in the overall pattern of brain growth between the two groups (p=.05). No increase in the incidence of hemorrhage, structural abnormalities or dampened arterial pulsations was noted. However, at 72 hours and 1 month, both right and left thalamic areas were smaller in the drug exposed infants than in the controls. The ICHD's were slightly smaller, but no differences were found in HC. Between 1 and 6 months of age, the rate of thalamic growth for drug exposed infants more than doubled that of the controls, resulting in greater thalamic circumferences at 6 months and HC were also somewhat greater. The data suggest a suppression of brain growth, particularly thalamic growth, in drug exposed infants during the first month of life, with a greater than normal growth rate of the thalami occuring at 6 months. Although growth occurred, the ICHD's of the drug exposed infants continued to remain smaller than the controls at 6 months of age. Additional USM and clinical evaluations are needed to ascertain the disparity in hemispheric and thalamic growth and its long-term effects.

1310 KING SYNDROME WITH FOCAL MYOPATHIC INVOLVEMENT DEMONSTRATED BY C-T SCAN. Qutub H. Qazi, Tariq M. Sheikh, Roger Kula, George Kassner, Downstate Medical Center, State Univ. of New York, Depts. of Pediatrics, Neurology, and Radiology, Brooklyn, New York.

Patients with King syndrome are known to suffer from a non-specific myopathy. By using C-T scan in one case we have found focal myopathy involving only a certain group of muscles.

The proband was a nine year old Mexican boy referred for evaluation of a neuromuscular disease. He tired easily while walking and had much difficulty climbing stairs. At six years of age, while undergoing an operation for undescended testes, he developed muscular rigidity, tachycardia, and acidosis during induction of anesthesia. His other physical features consisting of short stature, micrognathia, pectus carinatum, dorsal kyphoscoliosis, and lumbar lordosis were compatible with the diagnosis of King syndrome.

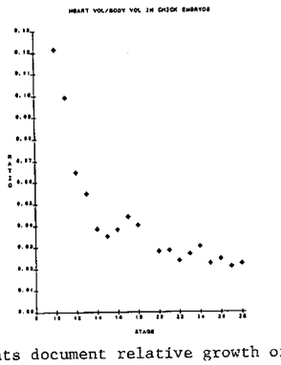
Neurological examination showed mild facial weakness, lordotic gait, equivocal Gower's sign, and proximally diminished DTRs. A needle EMG of selected proximal and distal muscles, and a muscle biopsy of vastus lateralis showed a chronic, morphologically non-specific, but severe myopathy. Fiber type differentiation was indefinite. A C-T scan at various levels through the extremities revealed a patchy involvement of certain muscle groups (e.g. vastus medialis, intermedius, and lateralis) with muscle degeneration and fatty infiltration, while others (e.g. rectus femoris and muscles of the posterior compartment) appeared well preserved. We conclude that myopathy in King syndrome is focal and can be demonstrated by a relatively non-invasive procedure.

●1311 PHENOTYPIC HETEROGENEITY IN THE JEUNE SYNDROME. C.S. Reid, S.J. Metz, & C.B. Whitley. Depts of Peds Johns Hopkins Univ School of Med, Balto MD & Univ of Minnesota, Minneapolis MN. (sponsored by David Valle)

Jeune syndrome (JS) is a rare autosomal recessive dysplasia which may present with asphyxiating thoracic dystrophy in the neonatal period or progressive renal or visual impairment in childhood and adolescence. Pathologic studies have shown cystic renal dysplasia and periportal hepatic fibrosis in JS patients of all ages, but the incidence and clinical significance of these abnormalities is unknown because few surviving patients have been extensively evaluated. We report 7 patients with diagnostic radiologic findings of JS, 6 of whom were extensively evaluated. Patients 1,2 & 3 presented as neonates, patients 4 & 5 in infancy and childhood and patients 6 & 7 as adolescents. Reasons for referral included: short stature (2), respiratory distress (2), renal disease (1), liver dysfunction (1) and incidental finding (1). Pts 4 & 7 were clinically well. Pts 1 & 2 had chronic pulmonary disease without clinical renal disease. Pt 6 had both chronic pulmonary disease and proteinuria. Pt 5 died of chronic renal failure. Both Pt 5 & 6 also had pigmentary retinopathy. Hepatic dysfunction occurred only in Pt 3; liver biopsy revealed fibrosis. These patients illustrate the multisystemic nature of JS and its variability in presentation. Although JS is classified as a skeletal dysplasia, its pleiotropic manifestations indicate a need for comprehensive evaluation in all patients with the disorder.

1312 RELATIVE SIZE OF HEART AND BODY IN THE EARLY CHICK EMBRYO Glenn C. Rosenquist, Pandu Soprey. George Washington University School of Medicine and Health Sciences and Children's Hospital National Med Ctr., Wash. D.C.

The heart begins to beat in the chick at the 10-somite stage and circulates blood into embryonic and extraembryonic regions by the 16-somite stage, even though valves and separate chambers develop later. This paper is an attempt to quantify the larger size of the heart over other organs in the early embryo. H-H stage 10 to 28 chick embryos were dissected free of extraembryonic membranes and the hearts removed. Hearts and bodies were pooled by stage in units of 1-5, minced, aspirated into capillary tubes, sealed, and centrifuged at 50G for 5 min. Tissue volume was calculated as: Vol=length x πr^2/embryo units. Our results indicate that although the heart >10% of body volume at stage 10 (40-44 hrs. incubation) this ratio drops to 2-3% by stage 28 (144 hrs incubation). We conclude that tissue volume measurements document relative growth of embryonic organ systems and show priority for heart growth in early development, with trend reversal at later stages.



1313 A CASE OF MURCS ASSOCIATION AFTER EXPOSURE TO CIMETIDINE IN UTERO. Burhan Say, Nancy Barber, Cindy Chambers. Children's Medical Center, Departments of clinical genetics and pediatrics, Tulsa, OK.

MURCS Association consists of Mullerian Duct, Renal, and Cervical Vertebral defects. A 30-year-old mother took cimetidine, 200 mg 4 times daily, during the first month of gestation and gave birth at term to a daughter with an absent vaginal opening, fusion of the cervical vertebra, and clubfeet. No uterus was palpated on rectal examination. The child is otherwise in good health with normal intelligence. Her chromosome studies were normal as were her parent's. Family history is negative for birth defects but positive for increased fetal wastage. The patient has three normal male siblings.

As this is a single case report and the affected person is a female, the association may be entirely coincidental. However, cimetidine is widely used. It is intriguing that intrauterine exposure to cimetidine in rats followed by neonatal exposure results in hypoandrogenization in adult males. In humans cimetideine in vitro blocks the positive chronotropic and ventricular iontropic effects of histamine on the fetal heart. In a single case report, doses up to 1 gm. per day caused no adverse effect on the fetus. No association between intrauterine cimetidine exposure and abnormal sexual differentiation in man has been reported to our knowledge. Cimetidine should be used with caution in pregnancy.

1314 POTENTIAL IMPACT ON THE FETUS OF GESTATIONAL HYPOGLYCEMIA. Lawrence R. Shapiro, Michael Bergman, Timothy B. Seaton and Carolyn Auerhahn. Departments of Pediatrics and Medicine, New York Medical College, Valhalla.

Tight control of diabetes has led to considerable concern regarding the occurrence of hypoglycemia in the non-gravid state since type I diabetic patients may manifest a defective counter-regulatory response to insulin-hypoglycemia. This concern has not been addressed with regard to the effect of maternal hypoglycemia on the fetus and ultimate childhood development.

We have studied well controlled pregnant diabetics and found that hypoglycemia (60mg/dl) occurs as frequently as 30% of the time. This incidence may be related to the severity of diabetes judged by the White classification. Earlier studies evaluating the effect of maternal hypoglycemia on neurological and psychological development of offspring defined hypoglycemia according to either perinatal blood glucose levels or anectodal symptoms of hypoglycemia during gestation.

Thus, the potential impact of hypoglycemia on the fetus is uncertain but has been inadequately investigated. To assess this problem, the occurrence of gestational hypoglycemia documented by capillary blood glucose monitoring with subsequent neurological and developmental testing is needed.