†1266 CAVUM VERGAE AND NEUROLOGIC ABNORMALITIES. Marvin E. Miller, Frederick Horner, and Daniel Kido. (Spon. by Elizabeth R. McAnarney) University of Rochester School of Medicine and Dentistry, Departments of Pediatrics and Radiology, Rochester, New York.

Cavum vergae is a midline cavity of the brain, posterior to the septum pellucidum which often communicates with a cavum septum pellucidum. While the clinical significance of this anatomic finding is not clear, we have seen 6 pediatric patients with cavum vergae who had neurologic abnormalities. They ranged from 3 weeks to 4 years of age at the time of presentation. The reason for initial evaluation was large head size in 3 patients, dysmorphic facies in 2 patients, and seizure associated with fever in one patient. The diagnosis of cavum vergae was made by CT scan of the brain - 2 of the 6 had only a cavum vergae while 4 had a cavum vergae and cavum septum pellucidum. It is significant that 5 of the 6 patients had developmental delay, and 4 had head circumferences >97th percentile without radiographic evidence of gross hydrocephalus. Two of the patients with large head circumferences were thought to have cerebral gigantism. To determine the background frequency of cavum vergae in children without apparent neurologic abnormality, we reviewed 50 CT scans of the brain in children between 4 months and 5 years of age whose CT scans were done for head trauma and found no cases of cavum vergae. These observations suggest that cavum vergae can be associated with neurologic dysfunction and macrocephaly.

1267 TOENAIL ZINC (Zn) IN MOTHERS OF PROGENY WITH NEURAL TUBE DEFECTS (NTDs), Aubrey Milunsky, J. Steven Morris, Walter C. Willett, Boston Univ. Sch. Med, Dept. of Peds., Research Reactor Facility (Missouri) and Harvard Univ. Sch. Public Health.

Diet-induced Zn deficiency in pregnant rats is known to cause cong. malfs. in 90-100% of their progeny, some 3% having spina bifida(SB).Mothers of children with SB have been reported to have high concentrations of hair Zn. Recent UK claims suggest vitamin A/folic acid periconceptional supplementation dramatically reduces the rate of recurrence of NTDs in women at risk.Because animal data have established a consistent relation between low plasma vitamin A and Zn deficiency, we studied the Zn status of women with a NTD fetus.Toenail clippings obtained mostly between 18-24 wks gestation provides a valuable retrospective assessment of maternal Zn status in the months prior to & around the periconceptional period.Zn analyses of toenails were performed by neutron activation analysis on 35 patients.21 controls (7 matched normal pregnancies, 14 normal females & males) had a normal range for Zn of 79.6-125 ppm.10 mothers with NTD fetuses, 2 with omphalocele, & 2 with with exogenous Zn intake were analyzed. High Zn values were found in 2 with SB fetuses (181;144), 1 with anencephaly (Anen.)(213), & 2 with Zn intake (208.4;290.8).Lowest value of all 35 was 1 with anen.(62.3).2 with SB+other malfs., 2 with Anen, 1 with lone SB all had normal Zn.In Zn deficiency both decreased and paradoxical increased Zn values have been found in hair.The consonance of our findings of high (3) or low(1) toenail Zn values among 10 mothers with NTD fetuses, suggests an abnormality in Zn metabolism in some women who bear children with NTDs.

●1268 FETAL LUNG GROWTH IN EXTRA-UTERINE PREGNANCY. Adrien C. Moessinger (Spon by W.A. Blanc). Dept. of Ped., Coll. of P&S of Columbia Univ. N.Y., N.Y.

Several hundred cases of advanced abdominal pregnancy have been reported. Although 20-40% resulted in livebirths, only 50-70% of those survived the early neonatal period. The major cause of death is unknown and most infants have multiple deformities involving mainly the joints and skin.

Using an animal model, we freed rat fetuses from the uterus without disturbing the placental implantation site. The fetuses were allowed to reside in the peritoneal cavity, either within intact membranes (with amniotic fluid), or outside of the fetal membranes. Untouched littermate fetuses served as controls. The surgery was performed on either days 17, 18 or 19 of gestation and the outcome was analyzed at term (i.e. 2,3, or 4 days later). The fetuses were then killed, photographed and body, lung, liver, brain and placenta weights recorded. Total lung DNA was determined. Results: Extra-uterine fetuses with intact membranes were in all respects indistinguishable from littermate controls. Extra-uterine and extramembranous fetuses weighed slightly less than controls, all had thin, shiny skin with webs and contractures of multiple joints. After correction for body weight, only their lungs weighed significantly less ($p<.01$) and contained less DNA ($p<.02$). Lung DNA per gram of fetal weight was less ($p<.05$). The difference was even more significant following surgery on days 17 and 18.

This animal model highlights the protective role of the amniotic environment and suggests that lung hypoplasia is a major cause of neonatal death in human abdominal pregnancy.

1269 AMBIGUOUS GENITALIA IN A NEWBORN MALE WITH DOWN SYNDROME. Lucinda Dykes, Michael A. Fragoso, Ray E. Helfer, Saroj Kapur, and Michael L. Netzloff. Michigan State University College of Human Medicine, Department of Pediatrics and Human Development, East Lansing, MI.

Males with Down syndrome have been reported with cryptorchidism and hypospadias but not with frankly feminine external genitalia. We report such a patient. The 3020 g. white infant was the product of a full-term pregnancy in a 33-year-old gravida 2 para 1 mother and was noted to have dysmorphic features consistent with Down syndrome.

Newborn examination by a pediatrician revealed a phallus 2 cm in length with urethral opening at its base (apparent clitoromegaly) and separate labial-scrotal folds with palpable masses (apparent testes). Systolic blood pressure was 54 mm Hg and serum sodium and potassium were 139 and 4.8 mEq/L, respectively. All concerned agreed that sex of rearing would be female.

Subsequent data included a karyotype of 47, XY + 21. Plasma cortisol and serum testosterone at 8:00 a.m. were 4.2 mcg/dl and 29 mg/dl, respectively. Following the stress of venipuncture, a p.m. ACTH was 114 pg/ml and cortisol was 5.8, which increased to 43.1 mcg/dl 60 minutes after I.V. infusion of Cortrosyn.

Diagnostic considerations exclude the rare, non-virilizing forms of congenital adrenal hyperplasia, but include a 17,20-desmolase enzyme defect in adrenal and testes. Alternatively, the ambiguous genitalia in this male patient may result from gonadotropin deficiency, or yet another rare congenital malformation association with the Down syndrome. To our knowledge, none of these associations have previously been reported.

●1270 MATERNAL CONNECTIVE TISSUE DISEASE AND CONGENITAL HEART BLOCK: DEMONSTRATION OF TISSUE IMMUNOGLOBULIN DEPOSITION IN CARDIAC INTERSTITIUM. Jacqueline A. Noonan, Stephen E. Litsey, William N. O'Connor, and Carol M. Cottrill, Albert B. Chandler Medical Center, Departments of Pediatrics and Pathology, Lexington, Kentucky

We recently demonstrated by immunofluorence microscopy antibody deposition in atrial interstitial tissue of an infant with fatal congenital heart block (CHB). Light microscopy showed extensive inflammation, calcification and scarring in the atrial septum which accounted for premature closure of the foramen ovale and CHB. By electron microscopy no discrete deposits were found suggesting a diffuse dispersion of interstitial antibody. Mother and infant both had elevated serum levels of antinuclear antibodies. This new observation provides evidence that placental transmitted antibodies may act directly on cardiac tissue to produce CHB and supports the report by Scott et al who found antibodies to ribonucleoprotein antigen in 34 of 41 mothers of infants with isolated CHB.

From our experience with three patients dying with CHB and review of the literature we postulate that maternal connective tissue disease both symptomatic and asymptomatic may result in placental transfer of antibodies which can act directly on the fetal cardiac tissue leading to: (1) CHB in the infant; (2) cardiovascular defects such as atrial septal defect, patent ductus arteriosus, or pulmonary valve dysplasia; and (3) fetal myocardial damage which may result in late onset congestive cardiomyopathy.

●1271 OMPHALOCEPHALY IN CHICK EMBRYOS AS A RESULT OF DELAY IN FORMATION OF THE ANTERIOR INTESTINAL PORTAL. Glenn C. Rosenquist, Children's Hospital National Medical Center, and George Washington U. School of Med, Washington, D. C.

This paper describes omphalocephaly in chick embryos surviving a mechanical insult in ovo. Under direct visualization, a portion of the right side of the endoderm/mesoderm layer of primitive-streak to early head-fold stage embryos was incised with a needle advanced through the yolk into the sub-blastodermal space. The 7-10 incisions were less than .05 mm in diameter. Among the 623 blastoderms re-incubated at 37°C, six omphalocephalic embryos survived to a total of 5-6 days incubation. In these embryos, most of the foregut endoderm had failed to converge at the anterior intestinal portal. As a result, the foregut was shortened and the liver and lung diverticulae failed to develop or were abnormal because the endodermal anlagen had not converged nor met the corresponding organ-specific mesenchyme. The brain assumed its normal curvature, so that the prosencephalon and flanking paraxial mesoderm had herniated ventrally, covered with endodermal rather than ectodermal epithelium. The heart was abnormally positioned cephalic to the head; chambers, curvature and circulation were normal. Embryos sacrificed less than 24 hours after the procedure had developmental delay in formation of the gut in relation to the head fold. Although no counterpart of this syndrome has been described in humans, the study of omphalocephaly in chick embryos is helpful in highlighting the importance of normal sequence in differentiation of embryonic germ layers.