937 MATERNAL SERUM ALPHA-FETOPROTEIN (AFP) SCREENING FOR NEURAL TUBE DEFECTS (NTDS). Aubrey Milunsky, Elliot Alpert, Frederic D. Frigoletto. Harvard Med. Sch., Eunice Kennedy Shriver Cntr., Mass. General Hosp., Boston Hosp. for Women, Depts. Pediat., Med. and Obstet. Gynecol., Boston.

About 90% of NTDs occur without a history of an affected sibling or parent. From available data, routine screening of all 2nd trimester maternal sera has the potential of early prenatal detection of 60-80% of anencephaly and 30-50% of spina bifida. We have done radioimmunoassays on 1608 maternal sera collected mainly from patients attending for routine antenatal care in early-mid pregnancy. Among 1060 women (pregnancy outcome known in 399 thus far) between 10-24 weeks gestation, there were 48 with fetal loss or major malformation. Of these, 16 had AFP levels $\geq +3$ S.D. above the mean (anencephaly (8), fetal loss (4), encephalocele (1), cong. nephrosis (1), fetal growth retard. (1), myelomeningocele (1). Raised AFP levels were noted in twins (2) and 5 normal pregnancies. Pregnancies with an untoward outcome which were not signaled by a raised serum AFP conc. included an open myelomeningocele (1), fetal death (1), Siamese twins (10), spontaneous abortion (18), fetal growth retard. (1), chromosomal abnormalities (2), and others. Assays were done on a research and not a clinical basis, while normal ranges were established. Even in retrospect the number of unnecessary amniocenteses because of falsely elevated serum AFP appears small. Maternal serum AFP screening warrants continuing evaluation.

938 FETO-PLACENTAL GROWTH AND THE OUTCOME OF PREGNANCY, Richard L. Naeye, Pennsylvania State University College of Medicine, M. S. Hershey Medical Center, Dept. of Pathology, Hershey, Pennsylvania 17033.

The study attempted to determine if widely used standards of fetal growth have been distorted by the inclusion of infants with unrecognized growth disorders. 4505 infants of various gestational ages who survived were compared with 1034 of similar gestational ages who died in the perinatal period with disorders not known to affect growth (group A) and with 51 who died after intrapartum accidents (group B). There were enough infants in the surviving group to compute mean body and placenta measurements starting at 28 weeks of gestation. Group A infants were undergrown at each gestational age compared with the surviving controls. Mean body weights in % of control values were: placenta previa 70%, abruptio placentae 85%, amniotic fluid infection syndrome 82%, premature rupture of the membranes 83%, fetal hypoxia of unknown cause 72%. There were similar reductions in placental weights. Body lengths and head circumferences were much less abnormal. By contrast, infants in group B had body and placental measurements close to those of the surviving controls. Mean body weights in % of control values were: acute compression of umbilical cord 89%, birth trauma 95%. Measurements of body length and head circumference were even closer to the control values. Widely used standards of fetal growth undoubtedly include many infants with growth disorders. (Supported by U.S.P.H.S. contract NO1-NS-3-2311).

939 DIFFERING POLYAMINE RESPONSES DURING TERATOGENESIS WITH DIPHENYLHYDANTOIN AND A FOLIC ACID ANTAGONIST. Michael L. Netzloff, Jaime L. Frias, Owen M. Rennert. Univ. Florida College of Medicine, Dept. Pediatrics, Gainesville.

Polyamine content is correlated with nucleic acid synthesis in rapidly growing tissue and may serve as an indicator of damage by toxins,including teratogens. This report concerns the alterations in embryonic polyamine content after exposure to teratogens. Primagravid Swiss albino mice were injected intraperitoneally on day 9 with diphenylhydantoin (DPH), 88 mg/kg body weight; embryos were recovered on day 11. Pregnant Long-Evans rats were intubated on day 10 and a modified diet was supplemented with 9-methyl pteroylglutamic acid (9-methyl PGA) on days 10-13. Spermidine, spermine and putrescine were quantitated on a Durrum D-500 amino acid analyzer, and are reported as nmoles/mg of embryonic protein. Rat embryos from pregnancies treated with 9-methyl PGA had unaltered polyamines. Mouse embryos whose mothers were exposed to DPH had significant reductions in spermidine and spermine, but putrescine levels remained comparable. Growth during polyamine accumulation is attributed to their stimulation of dihydrofolate reductase, the enzyme antagonized by 9-methyl PGA. Since polyamines were unchanged during this antagonism, they may play no direct role in the reactivation of this enzyme system.

Following DPH treatment embryonic DNA was lowered proportionally more than protein content, suggesting a block in DNA synthesis.

Folate deficiency has been suggested as the mechanism of DPH teratogenesis. However, the differing polyamine responses to the 9-methyl PGA and DPH teratogenic regimens indicate different mechanisms may exist.

940 THE GENETIC DIVERSITY OF SYNDROMIC ANAL ATRESIA. Leonard Pinsky. Lady Davis Inst. Jewish Gen. Hosp. and Dept. of Pediatrics, McGill University, Montreal.

It is accepted that vertebral, anal, tracheo-esophageal, renal and radial defects concur nonrandomly (the VATER "association"). Recently, the association has been "extended" to include cardiac, external genital and "other" (usually preaxial) limb anomalies. Despite repeated allusion to the etiologic heterogeneity that must underlie an inconstant association of such breadth, the universal appeal of classificatory labels has resulted in a growing tendency to exploit the VATER association as a definitive diagnosis for any child with an anorectal anomaly and at least one other defect typical of the association. Since VATER patients are mostly sporadic, indiscriminate use of the VATER diagnosis can lead to faulty genetic counseling. The purpose of this communication is to publicise six individually-rare patterns of malformation that include anorectal anomalies and have a high risk of recurrence. These patterns extend from familial expression of typical VATER association anomalies to those in which an anorectal anomaly and at least one other VATER defect coexist as components of familial syndromes that are otherwise distinctive. The latter include: (1) the Johanson-Blizzard syndrome (s); (2) the esophageal-facial-genital ("G") s.; (3) the s. of sacrum defects, anterior sacral meningocoele and anal malformation; (4) the cat eye s.; (5) the s. of imperforate anus, hand, foot and ear anomalies; (6) the s. of covered (anteriorly displaced) anus, preauricular skin tags and broad (bifid) thumbs. The VATER label is helpful for genetic prognosis only as a diagnosis of exclusion.

941 FAMILIAL PYLORIC ATRESIA AND EPIDERMOLYSIS BULLOSA DYSTROPHICA; REPORT OF 2 CASES. Ray Postuma, William G. de Groot, Thomas K. Goodhand. (Sponsored by James C. Haworth). University of Manitoba, Children's Centre, Departments of Pediatrics and Pediatric Surgery, Winnipeg, Canada.

Pyloric atresia, particularly the familial type, is rare. We present 2 cases where it was associated with epidermolysis bullosa dystrophica. To our knowledge, this association has not been reported elsewhere.

The first patient presented with non-bile stained vomiting since birth. Multiple, fluid-filled, bullous lesions of the skin and mucous membranes appeared at 2 days. She underwent a gastrojejunostomy for pyloric atresia but failed to thrive. The skin lesions spread and she developed severe hypoproteinemia, hyponatremia, anemia, septicemia and died at 9 weeks.

Her first cousin also presented with vomiting and underwent gastrojejunostomy for pyloric atresia at 2 days. He developed similar skin lesions that day; biopsy was characteristic of epidermolysis bullosa dystrophica. Persistent diarrhea contained casts of intestinal mucosa. He failed to respond to steroid, cholestyramine, antibiotic and parenteral nutrition therapy. Severe hypoproteinemia, anemia, and septicemia developed and he died at 16 weeks.

Autopsy showed extensive epidermolysis bullosa lesions of skin, scalp, nails and esophagus; focal ulcerations were found in the small intestine. The pyloric atresia measured 1.7 cm.

942 DERMATOGLYPHICS IN FETAL ALCOHOL SYNDROME Qutub H. Qazi, Akiko Masakawa, Barbara McGann, and James Woods State Univ. of New York, Departments of Pediatrics and Child Psychiatry, Downstate Med. Ctr., Brooklyn, N.Y.

An exposure of the fetus to deleterious agents during the period of formation of epidermal patterns may result in abnormalities of dermatoglyphics. To test the validity of this assertion the dermatoglyphics of 4 male and 13 female unrelated black children with fetal alcohol syndrome were studied. In all cases the diagnosis was based on characteristic clinical stigmata and a history of maternal heavy drinking during pregnancy. Since the number of male patients in the study was inadequate for any group comparisons, only the dermatoglyphics of female patients were compared to those of 100 black female controls from the New York area.

Significant differences between the patients and normal subjects were observed. The patients had (1) a lower frequency of whorls (13.1 versus 31.3) and a higher frequency of ulnar (73.8 versus 58.5) and radial (6.2 versus 1.7) loops ($P<.005$), and (2) a higher a-b ridge count (82.3 ± 3.01 versus 74.27 ± 1.10). No differences were found in the total finger ridge count, the width of the atd angle, and palmar and hallucal patterns. One patient had simian lines and another had sidney lines on both hands.

Because of the relatively small number of patients in the study, it is impossible to be definitive about the observed differences. However, if these observations could be confirmed in a larger sample of patients, the dermatoglyphics may serve as a useful marker in the fetal alcohol syndrome.