1297 EARLY NEURODEVELOPMENT IN INFANTS WITH SLIT LIKE VENTRICLES, Karol Kaltenbach, Matthew Pasto, Leonard Graziani and Loretta P. Finnegan, Jefferson Medical College of the Thomas Jefferson University, Departments of Pediatrics and Radiology, Philadelphia, PA.

Infants born to drug-dependent women usually undergo neonatal abstinence syndrome characterized by symptoms of central nervous system irritability, hypertonia, and diminution of the sucking reflex. The effect of fetal exposure to narcotics on the developing brain is not yet fully understood. Preliminary studies have shown that infants born to drug-dependent women tend to have ventricular configurations defined as small or slit-like during the first few days of life. The purpose of this study was to determine if a resumption of normal ventricular configuration occurs after abstinence is no longer evident and whether any developmental sequalae are evident. Ultrasound examinations, using a high resolution real-time sector scanner, were conducted on 31 infants at 24 hours and 6 months of age. All infants were born to drug dependent women and all exhibited symptoms of neonatal abstinence. Infants were assessed with the Bayley Scales of Mental Development (MDI) at 6 months of age. Results showed the presence of slit-like ventricles in 97% of the infants during the first 24 hours of life. By 6 months of age, normal ventricular configurations were present in the majority of infants. Nine infants (29%) continued to have slit-like ventricles. However, differences in ventricular configuration were not reflected in developmental status at 6 months of age. No differences were found in the Bayley Mental Development Index scores between infants with slit-like ventricles and those with normal ventricles ($t=.98$, $p>.20$). Furthermore, developmental scores for both groups were well within the normal range of development. These results suggest that the presence of slit-like ventricles at birth do not have an adverse effect on development, at least by 6 months of age, even if resumption of normal ventricular configuration has not occurred.

1298 ZINC DEFICIENCY POTENTIATES ETHANOL EMBRYOPATHY. *Laura Keppen, Theodore Pysher, Owen M. Rennert. (Spon. by M. Joycelyn Elders). University of Oklahoma Health Sciences Center, Department of Pediatrics and Pathology, Oklahoma City, Oklahoma. *University of Arkansas for Medical Sciences, Department of Pediatrics, Little Rock, Arkansas.

Alcoholism has adverse effects on zinc (Z) nutrition. We postulate that Z deficiency is synergistic with ethanol (E) in the fetal alcohol syndrome (FAS). We compared the effects of E on progeny of pregnant mice fed a Z deficient diet compared to a diet with high Z. Pregnant CBA mice (n=66) were fed the Lieber-DeCarli liquid diet with 0%, 15%, or 20% E containing .3 (L) or 8 (H)µg/cc Z. Dams were sacrificed on day 18 of gestation. Number of implantations (Imp), resorptions (Res), fetal (F) wt., and external malformations were recorded. Fetal Z and Z metalloenzymes, soft tissue anomalies, and skeletal malformations were assessed.

DIET	0%E HZ	15%E HZ	20%E HZ	0%E LZ	15%E LZ	20%E LZ
F Wt. $\bar{X}$ gm $\pm$SD	.63±.07	.62±.04	.46±.04	.45±.11	.43±.24	.26±.05
#Imp/#Res	23/0	56/1	15/0	26/0	56/21	27/14
F Z $\bar{X}$ µg/gm$\pm$SD	18±1.0	18±1.2	18±3.6	16±2.4	18±3.0	16±2.0

Fetal weights were lower in the groups fed the Z deficient diet for each concentration of E ($p<.005$). The groups fed the combination of low Z plus E had 37-52% res, while the animals on the Z deficient diet without E or the high Z diet with E had 0-2% res. Skeletal malformations were related to E concentration but not Z intake, while soft tissue anomalies were higher in those maintained on the low Z-E diet. These results suggest that Z deficiency potentiated the teratogenic effects of E and that nutritional intervention might reduce the incidence or severity of FAS.

† 1299 FACIOSCAPULOHUMERAL DYSTROPHY WITH COCHLEAR HEARING LOSS AND TORTUOSITY OF RETINAL VESSELS. PLEIOTROPISM OR HETEROGENEITY? Boris G. Kousseff, Maria Gieron, Jan Korthals, and Pat Nichols. (Spon. by John I. Malone), College of Medicine, University of South Florida, Department of Pediatrics, Tampa.

Facioscapulohumeral dystrophy (FSHD) is usually mild, has slow progression and lacks cardiac involvement and mental regression. The mode of inheritance is autosomal dominant. We studied 3 siblings and their mother who showed:

	Proposita	Brother	Sister	Mother
Age	12 y	8 y	5 y	35 y
Hypomimic face	+	+	+	+
Inability to pucker	+	+	+	+
Eyelid closure weakness	+	+	+	+
Scapular winging	+	-	-	+
Proximal arm weakness	-	-	-	+
Scoliosis	+	+	-	-
Elevated CPK	+	+	+	+
Cochlear hearing loss	+	+	+	+
Tortuosity of retinal vessels	+	+	+	+
Others		Hypospadias		

DTR and sensorium were intact. EKG and NCV studies were normal. EMG revealed myopathy. Muscle biopsy of deltoids showed type I fiber atrophy. No one else in the family had isolated hearing loss, muscle weakness or tortuous vessels.

The phenotype in this family suggests pleiotropism or a new type of autosomal dominant FSHD. Audiologic and ophthalmologic evaluations appear warranted for patients with FSHD.

● 1300 THE INCIDENCE OF CONGENITAL ANOMALIES AMONG INFANTS OF DRUG-DEPENDENT PREGNANT WOMEN, Linda L. Lai, Joyce Diodati, Saundra M. Ehrlich and Loretta P. Finnegan, Jefferson Medical College of the Thomas Jefferson University, Department of Pediatrics, Philadelphia, PA.

There is concern about infants born to drug-dependent women with regard to whether illicit drug use during pregnancy adversely affects structural fetal development. To assess the risk of fetal exposure to psychoactive drugs in-utero, a study of infants born to drug-dependent women enrolled in Family Center, a program providing prenatal care, counseling and methadone maintenance for these women, was conducted. From 1979 through 1983, 201 infants were examined for evidence of congenital anomalies. One-hundred and twelve of the infants (56%) were treated for neonatal abstinence; 89 infants (44%) required no treatment. Overall, the incidence of congenital anomalies in both groups fell within that of the general population (2-3%), suggesting that methadone maintenance, with or without accompanying illicit drug use during pregnancy, does not place the fetus at higher risk for anatomic defects. Of the anomalies seen, the cardiovascular system was most frequently affected. Structural congenital heart disease was found in 3.4% of the infants, (four times the incidence reported in the general population-.6%-.8%). Septal defects and pulmonic stenosis were the lesions identified. No explanation is available at present for these findings; however, the following should be considered: 1) Cardiac development occurs between weeks 2 and 8 of gestation, 2) The drug seeking behaviors of the addicted pregnant woman place her at risk for early intrauterine infection, 3) Exposure to viral teratogens (primarily coxsackie and rubella) has been implicated in abnormal cardiac development. These data suggest that, although the drug-dependent women may experience greater exposure to environmental teratogens early in pregnancy, the incidence of congenital malformations are not increased over that expected in the general population. However, the fetal cardiovascular system does appear more sensitive to environmental stressors in a pregnancy complicated by drug-dependency.

1301 CAMPTODACTYLY-ARTHROPATHY-PERICARDITIS (CAP) SYNDROME Ronald M. Laxer, Bonnie J. Cameron, Leonard D. Stein (Spons. By Richard Rowe), Division of Immunology/Rheumatology. The Hospital For Sick Children, Toronto, Canada.

Juvenile rheumatoid arthritis (JRA), the most common chronic childhood arthropathy, is a diagnosis of exclusion. We present a congenital syndrome which may be confused with JRA but is distinct on a clinical and histopathologic basis. A 5 year old boy was born with camptodactyly involving the 2nd to 5th digits of both hands. Surgical repair was required at 1 year. Polyarticular swelling, noted at 18 months, progressed to generalized decreased range of motion. X-rays showed only soft tissue swelling and osteoporosis, but no joint space loss or erosions. CBC, ESR, ANA, RF, C3, C4, CH50 and IgG, A, M were normal or negative. An asymptomatic pericardial effusion was detected at 4½ years of age, which progressed despite NSAID therapy. Symptomatic relief eventually required pericardiocentesis. A non-inflammatory fluid was drained. Pericardial biopsy showed minimal inflammation but extensive fibrosis. Synovial fluid from the right knee was non-inflammatory. Synovial biopsy showed multiple synovial giant cells (in the absence of a lymphocyte/PMN infiltrate) and extensive deposition of fibrin like material. Camptodactyly with subsequent arthropathy has been reported in multiple members of only 6 families, associated with pericarditis in only 1. This boy's HLA identical sister has no evidence of these manifestations. Camptodactyly beginning in utero may be an earlier and more severe manifestation of the same pathologic process involving joints and pericardium. In summary the CAP syndrome is clinically and histopathologically distinct from JRA. This is the first reported sporadic case of this association.

1302 SPECTRUM OF FINDINGS IN PREGNANCIES WITH SUSPECTED FETAL ANOMALIES. D. Manchester, M. Manco-Johnson, W. Clewell, D. Pretorius, P. Meier, E. Sujansky, J. Scott and C. Avery. Departments of Pediatrics, Radiology and Obstetrics, University of Colorado School of Medicine, University Hospital, Denver, Colorado 80262.

Ultrasound imaging can identify congenital anomalies prior to birth, but the sensitivity and specificity of this diagnostic technique in pregnancy are unknown. We have prospectively followed 172 pregnancies referred on the basis of abnormal ultrasound findings to a multidisciplinary program for evaluation and management of fetal anomalies. Our evaluation included repeat ultrasound examinations by sonologists specializing in fetal diagnosis. Our interpretation disagreed with the referring diagnosis in 41 cases (23%); we felt 27 were normal and 14 were abnormal but with a different diagnosis. Ultrasound revealed additional abnormalities in 46 cases (27%). Ninety-five percent of the diagnoses made by the program were confirmed at delivery. However, in 48 cases (28%), additional, unanticipated anomalies affecting either prognosis or therapy were also present. These included abnormalities previously reported as recognizable on ultrasound (i.e., congenital heart defects) and abnormalities unlikely to be seen (i.e., ambiguous genitalia). These results indicate that ultrasound diagnosis of fetal anomalies by experienced sonologists can be highly accurate in a referred population. Because this technique may be insensitive to some associated anomalies, management of abnormal pregnancies requires utilization of all available diagnostic modalities in both prenatal and postnatal periods.