**694** EFFECTS OF FAMILY CHARACTERISTICS ON THE RELATIONSHIP BETWEEN THE FUNCTIONAL STATUS OF CHRONICALLY ILL CHILDREN AND THEIR PSYCHOLOGICAL ADJUSTMENT. Ruth E. Stein and Dorothy J. Jessop (Spon. by Michael I. Cohen). Albert Einstein College of Medicine, Department of Pediatrics, Bronx, NY

The extent to which children's psychological adjustment is effected by the presence of chronic illness is a subject of much controversy. Data obtained upon entry to a study of children with chronic illness show that among the 81 children ≥5 years there is little relationship between adjustment and some traditional morbidity measures such as days hospitalized and days in bed. There is, however, a relationship between the child's functional status (FS) and psychological adjustment (PA). While this relationship is not dramatic for the sample as a whole (tau-b=.23), it varies from moderately strong to nonexistent among subgroups defined by social, family, and demographic characteristics. A characteristic such as the family composition alters the relationship between FS and PA from tau-b=.89 for mothers and another adult to tau-b=.03 where both parents are present in the home. The relationship between FS and PA is minimal where the mothers or fathers are at least high school graduates, or when the mother is presently employed, or has social supports in her life. These variables may be important buffers of the impact of illness on the child's adjustment. Such interactional effects may help to explain current controversies over the existence and importance of the relationship between the child's chronic condition and adjustment.

**695** ULTRASOUND (US) IN THE EVALUATION OF AN ABDOMINAL MASS Rita L Teele, Claudia I Henschke (spon. by John A Kirkpatrick) Harvard Med. Sch. Children's Hosp. Med. Ctr. Dept. Radiology Boston

From 1976 to 1980, 482 consecutive patients with the clinical suspicion of an abdominal mass were evaluated with US. 119 (25%) had no mass or an anatomic variant felt as a mass; 363 with a mass are displayed below as to area and type of mass.

| AREA | MALIGNANT TUMOR | BENIGN TUMOR | INFLAMMATORY | OTHER | TOTAL |
|---|---|---|---|---|---|
| INTRAPERIT. | 6.6 | 3.3 | 6.6 | 8.3 | 24.8% |
| RETROPERIT. | 25.9 | 5.5 | .8 | 12.1 | 44.3% |
| PELVIC | 5.5 | 9.4 | 4.4 | 5.5 | 24.8% |
| SOFT TISSUE | 3.9 | 1.4 | 0 | .8 | 6.1% |
| TOTAL | 41.9% | 19.6% | 11.8% | 26.7% | 100% |

Based on the data and results of other radiographic studies, an approach to the diagnosis was developed. Plain abdominal radiograph followed by intravenous urography (IVU) is appropriate when a mass is thought to be retroperitoneal in origin. If a tumor is diagnosed on the radiographs, US is used in its characterization and staging. A pelvic mass, if of gynecologic origin, is evaluated primarily with ultrasound. In young children and adolescent boys with a pelvic mass, US is coordinated with IVU. If the mass is anterior or associated with gastrointestinal bleeding or obstruction, plain radiographs followed by US and studies with barium are appropriate. An hepatic mass is evaluated with plain radiographs, US and, if necessary, nuclear scans and angiography.

**696** QUALITY OF PERINATAL THERAPEUTIC STUDIES. Jon E. Tyson, Jaime A. Furzan, Joan S. Reisch and Susan G. Mize (spon. by Charles R. Rosenfeld). Univ. of Tex. Health Science Center at Dallas, Depts. of Pediatrics and Medical Computer Science, Dallas.

To assess studies which compare or recommend perinatal methods of treatment or management, all such studies (86) published in 1979 in J. Pediatrics, Pediatrics, Am. J. Ob.Gyn, and Ob.Gyn.were reviewed independently by one of two neonatologists and one of two biostatisticians. A 5 page evaluation form was completed by each reviewer. Items noted by both reviewers included prospective design (48%), predetermined criteria for study completion (3%), adequate number of subjects (15%), adequate description of subjects (39%), clear indications for treatment (47%), use of blinding when feasible (36%), evaluation of treatment hazards (45%), and use of a randomized trial to support treatment recommendations (22%). Both reviewers considered the treatment recommendations fully justified in 10% of articles and partially justified in 69%. An overall score was calculated based on the 34 items considered most important to a well executed study. Despite disagreement on individual items, the total score for important items was highly correlated (r=.99). The mean score for all studies was 16.9 for biostatisticians (SD=6.5) and 17.2 for neonatologists (SD=6.5). The range was 3 to 32. Despite tragic past errors in the use of diethylstilbestrol, oxygen, sulfonamide, and chloramphenicol, poorly designed and executed studies are often the basis for therapeutic recommendations in perinatal medicine.

**697** ANALYSIS OF PERINATAL VARIABLES AFFECTING COMPLIANCE TO A FOLLOW-UP PROGRAM. B. Vohr, P. Daniel, W. Oh. Brown Univ. Program in Medicine, Women & Infants Hosp., Dept. of Ped., Providence, RI.

The primary goal of a neonatal Follow-up Program is the early identification of children with neurodevelopmental problems. A high attrition rate for Follow-up is a common problem and may reduce the effectiveness of the program. In this study, we attempted to identify the variables characteristic of the noncompliant family. 168 infants(birth weight 501-1500 grams)born between 1/1/75 and 4/1/77 and cared for in our Intensive Care Nursery were enrolled for Follow-up. 25 infants (Group I) made none of the 4 scheduled visits during the first year of life and 143 (Group II) made 1-4 visits. Group I included 1 institutionalized infant, 7 deaths during the first year, and 3 infants lost to Follow-up. The remaining 14 infants still live in the region but failed to visit the Follow-up Clinic. Significant variables distinguishing Group I from Group II were adolescent pregnancy (maternal age ≤18 years $p<.05$), unwed mothers ($p<.005$), and low socioeconomic score (SES) ($p<.05$). Analysis of multiple neonatal variables indicated no differences between the 2 groups. Neurological and psychological testing at 2-4 years of age showed that the abnormal infants in Group II had similar perinatal variables (adolescent and unwed mothers, low SES) that characterized the Group I (noncompliant) families. Thus, we conclude that the noncompliant population is at risk for neurodevelopmental morbidity and incentive programs should be instituted to lower the attrition rate for their follow up.

# GENETICS

**698** THERAPY OF NEONATAL ONSET UREA CYCLE ENZYMOPATHIES, (UCE). Mark Batshaw, George Sproul, Peter Mamunes, Wim Blom, Ruben Matalon, Richard Koch, Barbara Burton, Irwin Schafer, Virginia Michels, Saul Brusilow. Johns Hopkins Med. Inst. Balto. MD. and other inst.

We treated 12 children with neonatal onset UCE: CPS 1, OTC 3, AS 4, AL 4. CPS and OTC were treated with protein restriction (PR) 0.5-1g/kg/d + essential amino acids (EAA) 1g/kg/d + arginine 1mmol/kg/d + benzoate (B) 1.75 mmol/kg/d. AS was treated with PR + EAA + Arg (3-4mmol/kg/d) + B. AL was treated with PR + Arg (3-4mmol/kg/d). All patients are alive (mean age 12mo., range 1-32 mo.). Plasma NH4 levels were normal (< 35μM) or near normal except when dietary therapy was interrupted by illness or non-compliance. Then hyperammonemia (150-380μM) responded to intravenous Arg and/or B within 5 hours. Weight gain is normal; linear growth delayed. Intellectual development has been normal in 7, mildly delayed in 4 and severely delayed in one. Fasting plasma levels on therapy are: Arg 50-150μM; Gly 120-300μM, B 1-5mg%, hippurate 1-5mg%. There was a transient increase in SGOT in one case each of AS and AL. Two AL patients developed hyperlipemia. While receiving Arg one patient, inadvertently given 6mmol/kg B, developed vomiting and irritability with plasma Gly of 64uM and benzoate of 124mg%. SGOT was normal and the child recovered in 12 hours. Thus reduction of the requirement for waste nitrogen excretion (WNE) and promoting WNE as hippurate, Cit or argininosuccinic acid has been effective in permitting survival in these previously fatal diseases.

**699** INCREASED CHOLESTEROL SULFATE IN PLASMA AND RED BLOOD CELL MEMBRANES OF STEROID SULFATASE (STS) DEFICIENT PATIENTS. E. Anne Bergner and Larry J. Shapiro, Division of Medical Genetics, Harbor-UCLA Medical Center, Torrance,CA.

STS deficiency is an inborn error of metabolism due to a relatively common mutation on the short arm of the X chromosome. The phenotype of affected individuals includes decreased estriol production during fetal life and ichthyosis postnatally. Although increased levels of several steroid sulfates have been observed in amniotic fluid, maternal urine, and cord blood from such pregnancies, no consistent substrate abnormalities have been found beyond the perinatal period. Specifically, normal radioimmunoassayable plasma pregnenolone sulfate (PS), dehydroepiandrosterone sulfate (DS), and androstenediol sulfate (AS) have been reported. Utilizing gas chromatography (GC), we have found cholesterol sulfate (CS) to be strikingly elevated in plasma and red blood cell membranes of patients with STS deficiency. 80% methanol extracts were purified by solvent partition and TLC, subjected to solvolysis and quantitated by GC. Recoveries were monitored with $^3$H-CS.

| Group | Plasma CS (μg/dl) | RBC CS (μg/dl) |
|---|---|---|
| Controls | n=6 178 (115-220) | n=4 216 (161-293) |
| STS deficiency | n=4 3300(2200-4300) | n=4 6900(5000-9200) |

High levels of CS with normal levels of PS, DHEAS, and AS may indicate that desulfation is quantitatively more important for CS than for the other steroid sulfates. CS is known to affect membrane stability and the present observations may help to explain the pathogenesis of STS deficiency and X-linked ichthyosis.