618 ENZYMATIC MARKERS OF GASTROINTESTINAL MATURATION IN AMNIOTIC FLUID OF HIGH AND LOW RISK PREGNANCIES. Leo A. Heitlinger, William P. Dillon, and Emanuel Lebenthal. SUNY at Buffalo, Children's Hospital of Buffalo, Departments of Pediatrics and Obstetrics, Buffalo, New York.

Amniotic fluid samples were prospectively collected from women undergoing amniocentesis early in pregnancy (15-19 weeks) for genetic reasons (G; n=49), or later in pregnancy (≥ 35 weeks) prior to repeat C-section (C; n=52) or in the management of high risk conditions (≥ 35 weeks), ie., gestational diabetes (D; n=12) hypertension (H; n=6) and premature labor (P; n=8). Protein, lactase, sucrase, maltase, and amylase concentrations were determined in each sample. Maturational changes were detected going from groups G to C: disaccharidases decreased greater than ten-fold, while amylase increased more than four-fold. Protein concentrations did not vary with age. Sucrase was elevated in D compared to C (10.33±6.53 U/L vs. 2.94±4.55 U/L, P<0.001). Amylase was low in H compared to C (365±115 U/L vs. 679±481 U/L, P< 0.001). In contrast, sucrase (7.25±7.13 U/L, P<0.05) was high and amylase (158±196 U/L, P<0.05) was low in P compared to C. These data suggest that in normal pregnancies, amniotic fluid enzyme profiles are dependent upon gestational age and a mature pattern is attained prior to 35 weeks. In high risk pregnancies, attainment of a mature enzyme profile was not seen even at term. The mechanisms responsible for these apparently specific profiles remain to be elucidated. Supported in part by grants from NIH (HD 12586) and BRSG (150 E267K).

619 GRADE 1 REYE'S SYNDROME (RS) - OUTCOME AND PREDICTORS OF PROGRESSION TO DEEPER COMA GRADES. James E. Heubi, Cynthia C. Daugherty, Jacqueline S. Partin, William K. Schubert, John C. Partin, Dept. Pediatrics, Univ. Cinti., Child. Hosp., Cincinnati, OH, and SUNY, Stony Brook, NY.

Recent studies (N Engl J Med 309:133, 1983) suggest that non-comatose RS (so-called Grade 1) may comprise as many as 75% of all cases. We sought to determine the outcome of all cases of hospitalized Grade 1 RS and determine if any clinical or laboratory characteristics might predict progression to deeper coma grades. Since 1969, 84 patients had liver biopsy proven Grade 1 RS. Group A included 79 patients with no coma grade change and in Group B, 5 (5.9%) progressed to deeper coma grades (1 to Grade 2, 3 to Grade 3 and 1 to Grade 4). All patients survived without sequelae except one Group B patient with severe brain damage. There were no differences between groups in age at presentation, sex, admitting SGOT, glucose, CPK, BUN, salicylate level, uric acid, or hours of emesis prior to admission; however, admitting NH_3 was significantly higher ($p = .005$) in Group B (291 ± 42 µg/dl, $\bar{x}$ ± SEM) vs. Group A (52 ± 5 µg/dl) and the prothrombin time was significantly more prolonged ($p = .003$) in Group B (3.86 ± .49 seconds) vs. Group A (1.63 ± .16 seconds). Liver ultrastructural changes were severe in all Group B subjects, and varied from mild to severe in Group A. Conclusions: 1) The prognosis is excellent for survival without sequelae in Grade 1 Reye's Syndrome (98.8%) when management includes hospital surveillance and intravenous glucose-electrolyte infusion. 2) A small number (6%) progress to deeper coma grades and the admitting NH_3 and prothrombin time may predict progression.

● 620 EFFECTS OF TOTAL PARENTERAL NUTRITION (TPN) ON BILIARY FUNCTION IN RAPIDLY GROWING RATS. Melvin B. Heyman, Victor Ling, and M. Michael Thaler. University of California, Department of Pediatrics, and Liver Center, San Francisco.

The cholestatic effects of TPN in early infancy are well documented, whereas little is known about the influence of TPN on bile flow and bile acid output in growing children. Unrestrained actively growing rats weighing 50±4g (Y) and non-growing mature rats weighing 225±8g (A) were kept on TPN for 8 days with fluids containing 4.0% amino acids, 27.5% dextrose, minerals, vitamins, and trace metals. Controls were on standard chow or fed via gastrotomy using the nutrient solution. In each subgroup (3-5 rats), the study was terminated with a 150 min collection of bile obtained in 15 min aliquots from the common bile duct. All Y gained at least 20% B.W. during the 8 days; weight in all A was stable. Bile flow (µl/h/100g), biliary bile acid concentrations (mM), and bile acid output (mM/h/100g) were comparable in control and TPN-treated A. In Y controls compared with A, bile flow was greater (773±90 vs 298±45) and bile acid concentrations higher (45±7 vs 29±5), resulting in 3-4 fold greater bile acid output in Y. In striking contrast with A, bile flow was reduced 38% ($p<0.004$) and biliary bile acid concentration decreased 33% ($p<0.002$) in Y on TPN, causing bile acid output to decline from 35.6±11.4 to 14.8± 2.6. Conclusions: Bile flow and bile acid output are greater in Y than in A. TPN has no effect on these functions in A detectable after 8 days, while both bile flow and biliary bile acid concentrations are reduced in Y. Thus, growing rats secrete more bile and bile acids, and are susceptible to cholestasis from TPN.

621 A SCORING SYSTEM TO ASSESS NUTRITION AT BIRTH. Reba M. Hill, Linda M. Tennyson, Russell L. Deter, Baylor Col of Med, St.Luke's Episcopal & Woman's Hospital of Texas, Dept's. of Ped, & OBGyn, Houston.

357 infants delivered consecutively from a high socio-economic population were given a clinical score of 0-2 if well nourished (WN) or 3-4 if intrauterine malnourished (IM). 46 infants (14%) were identified as IM. To confirm the clinical impression of IM, the amount of subcutaneous tissue (SCT) at the face, neck, chest, lateral abdominal contour, anterior biceps, anterior thigh, back, buttocks & Wharton's jelly was scored 0-3, ie 3 representing abundance and 0 a decrease.
Infants with a clinical score of 3-4 had lower scores for SCT, weight, length, weight/length^2 X 100, FOC, chest, abdominal girth, & Wharton's jelly ($p<.01$) than infants with scores 0-2.

Clinical Score	SCT Score	WT Grams	Length cm	100 x Wt/L^2	FOC cm	Chest cm	Abd umb.
0	24±2	3661±436	51±2	142±11	35±1	33±2	32±2
1	21±2	3437±350	50±2	137±10	35±1	33±1	31±2
2	18±3	3180±274	51±2	128±8	34±1	32±1	30±1
3	16±3	3058±284	49±2	125±8	35±2	31±1	30±1
4	11±4	2758±376	48±3	117±8	34±1	30±2	28±2

The SCT score differed between the 5 categories (0-4) of infants ($p<.01$). Length, FOC & Wharton's jelly were least helpful in identifying an IM infant. Different patterns of SCT deposition were seen with scores of ≤ 2 & 3 or 4. The Kappa score to measure agreement between examiners was significant ($p < .02$)

622 SELECTIVE DENERVATION SUPERSENSITIVITY IN A HIRSCH-SPRUNG MODEL. Craig Hillemeier, Mark Evens, Jose Behar, Piero Biancani. (Sponsored by Robert Schwartz) Brown University and Rhode Island Hospital, Department of Pediatrics and Internal Medicine, Providence, Rhode Island.

In the mouse the Ls trait is an autosomal recessive allele, that when present in the homozygous Ls/Ls condition, causes a segment of distal colonic aganglionosis and obstruction similar to Hirschsprung's disease. Muscle rings were removed from the aganglionic colon (AGC) and ganglionic colon (GC) of Ls/Ls mice and from corresponding segments of control mice. Muscle rings tested in vitro exhibit spontaneous tone. Electrical field stimulation with parameters known to stimulate nerve fibers causes relaxation followed by contraction in GC of both Ls/Ls and control mice while the AGC did not respond. This neurally mediated relaxation is not affected by cholinergic or adrenergic block, while excitation is blocked by cholinergic antagonism. The AGC however is affected by agents that act directly on the muscle and in some instances responds at doses that have no effect on GC. In comparison to GC, the AGC is supersensitive to: 1) VIP, a putative neurotransmitter which causes relaxation 2) bethanechol, a cholinergic agonist which causes contraction 3) phenylephrine, an alpha adrenergic agonist which causes contraction. However, the AGC segment is not supersensitive to all direct acting muscle agonists; isoproterenol, a beta adrenergic agonist, causes relaxation without showing supersensitivity. We conclude that AGC may show denervation supersensitivity to VIPergic, cholinergic, and alpha adrenergic agonists, but not beta adrenergic agonists.

623 MECONIUM FROM INFANTS WITH CYSTIC FIBROSIS (CF) CONTAINS IMMUNOREATIVE TRYPSINOGEN. Monica C. Hsieh, Helen K. Berry (Spon. by Clark D. West). Children's Hospital Medical Center, Cincinnati, Ohio.

To understand the defect that leads to accumulation of proteins in CF meconium we measured immunoreative trypsinogen (IRT) by radioimmunoassay and trypsin catalytic activity (CA) by fluorimetric method. IRT concentration was normal in CF meconium based on total protein content, but was elevated based on wet weight, as was total protein and albumin. Trypsin CA was reduced in CF meconium compared to normal meconium:

	CF n = 22	Normal n = 26	Probability
IRT[a]	7.1 ± 4.2	3.1 ± 2.7	<.001
IRT[b]	38 ± 24	54 ± 44	n.s.
Total prot.[c]	205 ± 81	57 ± 12	<.001
Albumin[d]	72 ± 40	<10	Not tested
Trypsin CA[e]	.25 ± .27	1.03 ± .99	<.001
Trypsin CA[b]	42 ± 48	357 ± 196	<.001

[a] µg/g meconium (wet weight); [b] µg/g protein; [c] mg/g meconium; [d] µmol substrate hydrolyzed/g meconium; [e] µmole/mg IRT

Trypsin inhibitory activity. measured by incubating porcine trypsin with meconium was: CF, 33 ± 27; normal, 1 ± 3 µg porcine trypsin inactivated/g meconium. Trypsinogen, an inactive precursor of trypsin, is synthesized by pancreatic acinar cells and secreted into intestinal lumen. The results show this process is normal in infants with CF. Presence of trypsin inhibitor(s) rather than absence of IRT accounts for accumulation of proteins in CF meconium.