726 PANCREATIC AND BILIARY TRACT DISEASE IN CHILDREN: THE BENEFIT OF ERCP. Steven Werlin, Mark Allendorph, Joseph Geenen, Walter Hogan, Edward Stewart. Departments of Pediatrics, Medicine and Radiology, The Medical College of Wisconsin, Milwaukee, WI.

Although ERCP has become a widely used diagnostic and therapeutic modality in adults for evaluation and treatment of pancreatic and biliary tract disease, there have been few reports of this technique in children. Consequently we reviewed 40 ERCP procedures performed in 32 children and adolescents (26 F, 6 M, age 15 mos-18 yrs). General anesthesia was used in 12, sedation in 20. In the 30 patients who were successfully cannulated, abnormalities were found in 12 of 18 with suspected pancreatic disease, 4 of 7 with suspected biliary tract disease, 3 of 5 with abdominal pain, and 2 of 2 with suspected sphincter of oddi (SO) dysfunction. Findings in the 19 patients with abnormal ERCPs included pancreas divisum (3), pancreatitis (3), common bile duct abnormality (3) pseudocyst (2), biliary tract stones (2), choledochal cyst (2), SO dysfunction (2), choledochocoele (1), and sclerosing cholangitis (1). Minor complications in 6 patients included pancreatitis (3), intramural injection (2), and vomitting (1). The surgical approach was influenced in 8 patients including excision of choledochal cyst (2), drainage of pseudocyst (2), sphincteroplasty (2), excision of choledochocoele (1), and choleystectomy (1). Endoscopic sphincterotomy and stone extraction was performed in 1 patient.

Conclusions: ERCP is a safe and valuable diagnostic and therapeutic tool in children and in appropriate patients may guide operative therapy.

●727 ONTOGENY OF EXOCRINE PANCREATIC FUNCTION IN ISOLATED ACINI FROM IMMATURE PANCREAS. Steven L. Werlin, Joseph M. Harb, Medical College of Wisconsin, Department of Pediatrics, Milwaukee, WI

Morphological maturation and developmental increases in enzyme content of rodent pancreas have been well characterized. In contrast ontogeny of the secretory process has not been extensively evaluated. Consequently we evaluated the secretory process and ultrastructure of isolated acini (ISA) prepared from immature rats by digestion with collagenase and mechanical shearing. ISA prepared from 48 hr old rats responded to both carbachol (C) and CCK-8 with increased secretion of amylase. Characteristic biphasic dose response curves peaked at 3×10^{-6} M for C (636% increase) and at 10^{-7} M for CCK-8 (576% increase). Atropine inhibited C induced secretion. ISA prepared from rats age 4 hr failed to respond to C and CCK-8, but did respond to the calcium ionophore, A23187 (200% increase). Digestion and shearing used to prepare the acini did not alter the ultrastructural appearance of the acinar cells. Cells from acini incubated with C and CCK-8 had increased numbers and greater variation in size of zymogen granules than did the controls. Conclusions: 1) Although ISA prepared from 48 hr rats respond to secretagogues similar to mature rats, responsiveness to secretagogues is not found in ISA prepared from newborn rats. 2) Responsiveness to A23187 in the newborn suggests that the immature step is prior to calcium mobilization. 3) Since physiological and morphological integrity is maintained in ISA prepared from immature pancreas, this procedure can be a useful tool in further elucidation of the ontogeny of the secretory process.

† 728 GLYCOPROTEINS OF HUMAN JEJUNAL CELLS: LOCATION, MATURATIONAL CHANGES AND INDIVIDUAL VARIATION. Milton Westphal, Bradley A. Schulte and Samuel S. Spicer. Medical University of South Carolina, Departments of Pediatrics and Pathology, Charleston, SC 29425.

A battery of 12 lectins conjugated to horseradish peroxidase was used to characterize secretory and cell surface glycoproteins (GP) in secretions from small bowel biopsies of 9 children. The glycoproteins found differed from site to site, among individuals at a given site and from crypt to villar tip. In goblet cells stored secretions showed individual variation in intensity of staining and increasing fucose content from crypt to villus. Terminal N-acetylglucosamine was seen in the Golgi cisternae but not in stored secretion, suggesting capping of this sugar by other residues during GP biosynthesis. Terminal α-N-acetylgalactosamine was present in Golgi cisternae of all and in the stored secretion of two-thirds of the specimens. Immature cryptal columnar cells had fucose-rich GP presumably in secretory granules. Mature villar cells did not. Columnar cells showed mannose-rich GP, presumably lysosomal enzymes, in apical punctate bodies. In some specimens the basolateral plasmalemma of columnar cells stained selectively for fucose and ß-galactose. Brush border staining generally paralleled that of goblet cell secretions with similar maturational changes. Staining for terminal galactose highlighted cells, presumably leukocytes, infiltrating the epithelium and lamina propria. The diversity and variability of the GP in human jejunal mucosa suggest a relationship between specific chemical structures and biologic function at the cytologic loci.

† 729 ELECTROPHYSIOLOGIC MEASUREMENTS OF RECTAL MUCOSA IN CYSTIC FIBROSIS - ABNORMAL RESPONSE TO AMILORIDE. Milton Westphal, Stanley Trojanowski, John G. Blackburn, Samuel S. Spicer and Donald A. Sens. Medical University of South Carolina, Departments of Pediatrics, Physiology and Pathology, Charleston, SC 29425.

The negative potential difference (PD) between rectal mucosa and subcutaneous (SC) space is mainly due to transmucosal sodium transport from the lumen. In order to examine the electrolyte transport defect in Cystic Fibrosis (CF) we measured change in rectal PD in response to normal saline, 10^{-4}M amiloride, 10^{-4}M furosemide and 0.102 M Na_2SO_4 applied topically. Seven CF Patients and seven Controls were in Na balance on diets containing 120 mleq Na per day. We perfused the rectum through a double lumen, double balloon catheter.

Mean Negative PD between Rectal Mucosa and SC Space

	NaCl	Amiloride	NaCl	Furosemide	NaCl	Na_2SO_4
Normal	31.1	21.9*	24.0	30.9	23.9	40.0
CF	29.1	1.6*	31.0	38.6	26.2	38.3

* $p < 0.001$, other normal vs CF differences N.S.

During amiloride perfusion CF patients had a significantly smaller negative rectal-SC PD than normal controls. Thus, amiloride blocked rectal Na transport more completely in CF than in normals. Rectal PDs during perfusion with normal saline and with furosemide and Na_2SO_4, both CF blockers, were similar for CF and normals. These data are compatible with defective facilitation of sodium transport in CF or with the presence in the normal of a non-amiloride sensative Na^+ transport mechanism which is absent in CF.

730 AMINO ACIDS IN INFANTS WITH NON-CORRECTED EXTRAHEPATIC BILIARY ATRESIA (EBA). Sally A. Weisdorf, John Fath, Deborah K.Freese, Frank B.Cerra. Univ. of Minnesota, Depts. of Pediatrics and Surgery, Minneapolis, MN.

Feeding formulations which are enriched in branch chain amino acids (BCAA) and depleted in aromatic amino acids (AAA) and methionine (Met) are used in therapy of adults with hepatic encephalopathy (HE). These formulations are also being used in children with severe liver disease. In order to document a rationale for such therapy we examined amino acid profiles in 20 infants with EBA and cirrhosis who were candidates for liver transplantation, none of whom had clinical signs of HE. Similar to reports in adult patients, we found that our patients had a low BCAA/AAA. While this ratio is lower (2.83 vs. 3.45) for normal children than for normal adults our patients mean BCAA/AAA was 1.02 ± 0.61). This was due both to decreases in BCAA (mean total BCAA was 24.57 um/dl for EBA vs. 42.16 for normals) and increases in AAA (24.51 vs. 14.93). Mean Met levels were moderately elevated in 16 patients (5.27 vs. 2.17) with striking increases in 4 patients considered endstage on the basis of bilirubin level and coagulopathy. While proline and threonine have been reported to be elevated in adults, this was not true for these children. Taurine levels were also normal. We conclude that there is justification for use of BCAA enriched, AAA and Met depleted formulations in children with EBA in the absence of HE, but that because of individual variation in disease-related disorders of amino acid metabolism such therapy requires close monitoring of the amino acid profile in these patients.

●731 AN ACOUSTIC METHOD FOR MEASURING INFANT BODY VOLUME. Dean Winter, William Deskins, Hwai-Ping Sheng, and Cutberto Garza (Spon/B.L. Nichols). Baylor College of Medicine, USDA/ARS Children's Nutr.Res.Ctr., Dept. of Pediatr., University of Houston, Dept. of Mech. Eng., Houston, TX.

An acoustic plethysmograph that measures body volume has been developed for body composition studies on premature infants. It is based on the principle that the resonance frequency of a Helmholtz resonator depends, in part, on the volume of the resonator chamber. Thus, when an object is placed inside the chamber, the resonance frequency changes in proportion to the volume of the object. The prototype system consists of a 20 x 20 x 45 cm plexiglass chamber with a hinged front for access. A loudspeaker, driven by a variable frequency tone generator, is aimed at a 7.5 cm diameter opening in the top surface of the chamber. The frequency of this tone is monitored by a frequency counter. The resonance frequency corresponds to the frequency at which the sound level inside the chamber, as detected by a microphone in the chamber, reaches a maximum. Chamber temperature was maintained at 35°C and the sound pressure level below 75 dBa. The device was calibrated by measurement of change in resonance frequency when known volumes were placed in the chamber. The volumes of a number of inanimate objects of unknown volume were measured, first by the acoustic technique, then by water displacement. The two methods differed by less than 2% for volumes between 250 and 1500 ml. Eight anesthetized minipigs were measured acoustically, and after sacrifice, measured by water displacement. The two measurements differed by less than 5%. When the sacrificed pigs were measured acoustically, the acoustic and water displacement measures differed by less than 1%.