415 DIAGNOSIS OF EXOCRINE PANCREATIC INSUFFICIENCY IN CYSTIC FIBROSIS (CF) BY A SYNTHETIC PEPTIDE (Bz-Ty-PABA). S. Arvanitakis, C. Arvanitakis, N.J. Greenberger, (Spon. by C.T. Cho), Univ. of Kansas Medical Center, Depts. of Pediatrics and Medicine, Kansas City, Kansas.

Documentation of pancreatic insufficiency in children with CF is important since 20% have normal function and do not require specific treatment. Currently, there are no simple tests to determine pancreatic function in children. We have evaluated a new approach for the diagnosis of pancreatic disease in CF. The synthetic peptide N-Benzoyl-L-tyrosyl-p-amino-benzoic acid (Bz-Ty-PABA), administered orally, is specifically cleaved by the endopeptidase chymotrypsin to Bz-Ty and PABA; the latter is absorbed and excreted in the urine. We studied: a) a control group of children and b) children with CF and steatorrhea (fecal fat > 18 gm/72 hr). Bz-Ty-PABA, 14 mg/kg, equivalent to 5 mg/kg of PABA, was given orally and urine was collected over a 6 hour period. Results were as follows (mean $\pm$ SD):

Groups	Urine PABA (% per 6 hr)	Fecal chymotrypsin (μ g/gm)	Fecal fat (gm/72 hr)
a. Control (n=12)	73 $\pm$ 10	260 $\pm$ 123	16 $\pm$ 6
b. CF (n=10)	34 $\pm$ 9*	61 $\pm$ 49**	77 $\pm$ 31*

*p < 0.001 **p < 0.005

There was a significant correlation of PABA recovery with fecal chymotrypsin (r=0.60, p<0.05) and fecal fat (r=-0.68, p<0.05). Urinary recovery of PABA of the peptide Bz-Ty-PABA 1) correlates with other parameters of exocrine pancreatic function, and 2) may prove a practical test of pancreatic function in CF.

416 ELEVATED CHOLESTEROL(CH)/PHOSPHOLIPID(PL) MOLAR RATIO IN RED CELL MEMBRANES-AN ACQUIRED DEFECT IN CIRRHOSIS WITH ARTERIOHEPATIC DYSPLASIA. William F. Balistreri, Elfrid Cifuentes, Philip G. Holtzapfel, Mary Leslie, and Richard A. Cooper. Depts. of Ped. and Med., Univ. of Penn., Phila.

Hemolytic anemia with cholesterol-rich, spur-shaped red cells has been described in adults with severe hepatocellular disease due to alcoholic cirrhosis. The red cell defect (elevated membrane CH content, without changes in PL content) is acquired from serum, and causes an increase in the microviscosity of membrane lipids. A 4½ year old girl with arteriohepatic dysplasia, intrahepatic cholestasis and cirrhosis developed progressive anemia (Hgb=5.8), reticulocytosis (21%), increasing jaundice (41 mgm/dl; 23 direct), and splenomegaly. Serum levels of iron, folate, Vit. B₁₂ and E were normal. Peripheral smear showed fragmented cells and spur cells. Analysis of patient's fresh red cells revealed elevated CH (22.3% /10⁸ cells; N=14.5), normal PL (33.0% /10⁸ cells; N=31.8), an increased CH/PL molar ratio (1.35; N=0.92), and an increase in microviscosity at 37° (4.1 poise; N=3.0). Following incubation with the patient's plasma at 37° for 24 hours, normal cells acquired the spur-shaped morphology with an associated decrease in osmotic fragility and a 25% increase in cholesterol content. Patient's cells incubated with normal plasma acquired morphological features of spiculated spherocytes, with an increase in osmotic fragility and a 21% decrease in cholesterol content.

CONCLUSION: These studies demonstrate that a process identical to spur-cell anemia in adults with alcoholic cirrhosis may accompany cirrhosis in children with intrahepatic cholestasis.

417 INTESTINAL NECROSIS AND PERFORATION CAUSED BY INTRA-ARTERIAL CALCIUM GLUCONATE INFUSION. Linda S. Book and John J. Herbst. Univ. of Utah, College of Medicine, Department of Pediatrics, Salt Lake City, Utah.

The umbilical artery catheter is frequently utilized as a route for administration of medication in the premature infant. The effect of intra-arterially administered calcium gluconate (Ca Gluc) on the intestine was studied in the rabbit by injecting 1 cc of 10% Ca Gluc, 1% Ca Gluc, or saline into a first order arterial arcade supplying approximately a 10 cm segment of ileum. All animals receiving Ca Gluc demonstrated varying periods of vasodilatation following the injection. Rabbits were sacrificed between 3 and 6 days and the gross and histologic findings compared for saline and Ca Gluc injected rabbits:

	N	Gross Lesion	Mucosal Thickness $\pm$ 1 SD	Mitotic Index $\pm$ 1 SD
Saline (Control)	4	0	.75mm $\pm$.07	3.52 $\pm$.12
10% calcium gluconate	6	6	.45mm $\pm$.17	1.42 $\pm$.43
1% calcium gluconate	5	3	.48mm $\pm$.03	1.63 $\pm$.32

All animals receiving 10% Ca Gluc showed mucosal atrophy with 3/6 having perforation and 1/6 with mucosal hemorrhage. 3/5 animals receiving 1% Ca Gluc showed mucosal atrophy. Intra-arterial infusion of Ca Gluc in the usual therapeutic doses and concentrations administered to premature infants produced serious intestinal injury characterized by mucosal atrophy, depressed mitotic index and intestinal perforation.

418 PSEUDOMEMBRANOUS ENTEROCOLITIS (PMEC) IN CHILDREN. J.P. Butts, A.M. Weber, C.C. Roy, and C.L. Morin. Univ. of Montreal, Ste. Justine Hospital, Dept. of Pediatrics, Montreal, Canada.

Between 1970 and 1975, PMEC was documented in 5 children (5½ - 12 yrs of age). The diagnosis was based on proctoscopic and histological findings of characteristic pseudomembranes. In all patients the disease occurred during or immediately following a course of antibiotics: ampicillin (3), penicillin (1), clindamycin (1). Two children had been recently operated on (tonsillectomy, mastoidectomy). The sudden onset of profuse diarrhea was invariably present. Bloody stools, fever and abdominal distension were inconsistent findings. The clinical course was severe in 4 with hypoproteinemia, complicated by edema, ascites, pleural effusion, sepsis and/or shock. Two children eventually died despite intensive supportive therapy and surgery in the only case with small bowel involvement. In the two patients with exudative enteropathy who survived parenteral alimentation was used in addition to plasma infusions, steroids and I.V. antibiotics.

The following conclusions are warranted: 1) There is a risk of antibiotic associated PMEC in children even with commonly prescribed drugs such as: ampicillin and penicillin. 2) When profuse diarrhea does not respond promptly to discontinuation of antibiotic therapy proctoscopy should be performed before alarming symptoms of PMEC develop.

419 ESOPHAGEAL MOTOR DYSFUNCTION IN 11 PEDIATRIC PATIENTS WITH CHRONIC IDIOPATHIC INTESTINAL PSEUDO-OBSTRUCTION SYNDROME (CIIPS). William J. Byrne, Arthur R. Euler, Andrew F. Ippoliti, Marvin E. Ament, University of California, Center for the Health Sciences, Departments of Pediatrics and Medicine, Los Angeles.

Asymptomatic esophageal motor abnormalities may be part of CIIPS. To identify these abnormalities, barium studies of the esophagus (BSE) and esophageal manometrics (EM) were done on 11 children, ages 2 to 18 years, with CIIPS but without esophageal symptoms. The results are presented in the table below. Radiographic abnormalities, limited to poor motility (PM), were present in 6 and correlated with EM abnormalities which were present in 9. Lower esophageal sphincter pressure (LESP) was decreased (+) in 6. Seven had + amplitude of contractions (AC) and in 5 this correlated with a + LESP. Peristalsis (P) was absent in 6 and correlated with a + LESP in 4 and + AC in 5.

CASE	1	2	3	4	5	6	7	8	9	10	11
BSE	-	-	-	PM	PM	PM	-	PM	PM	PM	-
LESP	-	-	-	-	-	+	+	+	+	+	+
AC	-	-	-	DD	DT	DD	DT	-	DD	DD	DD
P	AT	-	-	AD	-	AD	AT	-	AD	AD	-

- normal DD-decreased distally DT-decreased throughout AD-absent distally AT-absent throughout
Esophageal motor abnormalities in CIIPS are characterized by + LESP, + AC, and absent P. EM, a more sensitive technique than BSE, should be done to identify their presence and serves as a useful adjunct in the diagnosis of this syndrome.

420 CHOICE OF TISSUE TO ESTABLISH THE DIAGNOSIS OF MAGNESIUM DEFICIENCY IN AN ANIMAL MODEL FOR THE HUMAN INFANT. Joan L. Caddell and Rita Schepner (Spon. by Arthur E. McElfresh), Dept. Pediat. and Path., St. Louis U. Sch. Med., St. Louis, Mo.

To find the best tissue to assess the magnesium (Mg) status of the sudden infant death victim, male 28-38 g Sprague Dawley rats were fed fed diets containing 100 or 0 mg of Mg/ 100 g and 20 % casein (100-20 or 0-20) for 1 week; others were fed 150 or 5 mg of Mg/ 100 g and 40 % casein for 2 weeks; and selected tissues were analyzed for cations on an atomic absorption spectrophotometer. Mg values are presented here.

Acute: 1 wk.	100-20-fed rats	0-20-fed rats	P-value
Vitreous humor*	57.5 $\pm$ 3.7 (12)*	47.4 $\pm$ 1.7 (23)	0.01
Heart**	99.7 $\pm$ 0.9 (35)	87.0 $\pm$ 0.9 (77)	0.001
Skeletal muscle**	106.5 $\pm$ 0.7 (26)	100.8 $\pm$ 0.5 (57)	0.001
Sternum**	143.5 $\pm$ 2.2 (23)	69.9 $\pm$ 1.0 (58)	0.001
Femur**	268.7 $\pm$ 3.6 (54)	98.0 $\pm$ 1.7 (107)	0.001
Attenuated: 2 wk.	150-40-fed rats	5-40-fed rats	P-value
Vitreous humor*	52.6 $\pm$ 3.1 (4)	43.2 $\pm$ 1.3 (12)	0.01
Heart**	92.2 $\pm$ 1.6 (15)	89.4 $\pm$ 1.2 (28)	NS
Skeletal muscle**	103.0 $\pm$ 3.1 (12)	101.1 $\pm$ 1.0 (21)	NS
Sternum**	168.7 $\pm$ 9.3 (4)	66.9 $\pm$ 1.4 (23)	0.001
Femur**	283.5 $\pm$ 3.9 (21)	77.6 $\pm$ 1.3 (36)	0.001

* mEq of Mg/ Kg dry wt.; ** mEq of Mg/ Kg dry, fat-free wt.

Mean $\pm$ SEM (Number of samples); (V. humor samples pooled)

Individual Mg concentrations in vitreous humor and soft tissues overlapped between control and Mg-deficient groups, but in bone were distinctly separate. One bone should be consistently analyzed. Support: Missouri Heart Assoc. and the Life Seekers, St. Louis.