451 ORGANIC ACIDURIA FOLLOWING EXTENSIVE SMALL BOWEL RESECTION.

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Routine screening has identified two patients with striking organic aciduria after extensive small bowel resection. JK, a 5 1/2 yo male, underwent resection of all but 21 cm of small bowel at 3 days of age because of necrosis secondary to midgut volvulus and subsequently has had recurrent acidemic episodes. TK, a 5 yo female, was left with 15 cm of jejunum following mesenteric artery thrombosis at 15 months. Both have experienced episodes of progressive abdominal distention, colonic ileus, and CNS depression. Urinary lactate (JK only), glutamate, propionate, p-hydroxyphenylacetate and β -phenyllactate were elevated. White blood cell glutaryl-CoA dehydrogenase activity in JK was normal. Serum prostaglandin E and F were significantly elevated in JK. Absorption of organic acids from stool has been reported and these patients illustrate that this may become quantitatively significant following small bowel resection. The data also suggest that the acidemia of short bowel may not be solely due to bicarbonate wasting. Our patients demonstrate that gastroenterologic disorders constitute a previously unrecognized etiology to be differentiated from inborn enzymatic errors associated with glutaric aciduria. Dietary restriction, oral bicarbonate, catharsis, and inhibitors of prostaglandin synthesis may play a role in treatment of this disorder.

452 IRON ABSORPTION FROM HUMAN MILK, SIMULATED HUMAN MILK AND PROPRIETARY FORMULAS.

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Studies from our laboratory have shown that iron is better absorbed from human milk than from cow milk and can provide sufficient iron for infants during their first year. This study was designed to compare iron availability from human milk with other formulas and determine factors responsible for its superiority. Adults were fed 100 ml of the following: human milk (HM), simulated human milk (SHM), simulated human milk containing added lactoferrin (SHM-L) and 2 commercial formulas (CF) containing iron, 12 mg/qt. The SHM resembled HM in concentration of protein, fat, carbohydrate, iron, total minerals, calcium and phosphorus. Fe⁵⁹ was added to each feeding and iron incorporation into red cells was determined 14 days after each feed. Iron absorption was highest from HM (15.7%) and lowest from one of the CF (1.7%). The SHM supported a 9.3% absorption; addition of lactoferrin reduced this to 4.7%. Net iron absorption was 0.12 mg/L from HM and 0.20 and 0.37 mg/L from the iron enriched CF's. This study demonstrates that the enhanced iron absorption from HM is not a simple result of its gross composition or the presence of lactoferrin, and that HM, with only 6% of the iron of a leading CF, can provide as much as 60% of the iron derived from such a proprietary milk.

453 PROTEIN SUPPLEMENTED FASTING IN CHILDREN.

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Protein supplemented fasting allows safe rapid wt reduction in adults. We have studied a protein supplemented fast in 5 male and 2 female obese children aged 9-14 weighing >150% of ideal wt. They received lean meat supplying 1.5-3 g protein and 7-20 CHO-free calories/kg ideal wt/day plus supplemental potassium (K) (25 mEq), multivitamins and calcium. Aside from nausea and hypotensive symptoms in the diuretic phase of the first week, the diet was well tolerated without complaint of hunger. Ketonuria appeared within 3 days. At 7-10 days, blood β -Hydroxybutyrate was $3.37 \pm .63$ mM (mean $\pm$ SEM), plasma free fatty acids were 1.75 ± 1.8 mM, and fasting blood glucose fell significantly from $5.81 \pm .25$ mM to $4.06 \pm .35$ mM. During the 4 weeks there was significant elevation of uric acid and depression of total lymphocyte count. Total wt loss was 7.0 ± 0.2 kg (mean $\pm$ SEM). There was a loss of 1.1 ± 0.3 kg lean body mass (LBM) as estimated from daily N-balance. Total body K by K^{40} counting declined $6.0 \pm 0.5\%$. Weekly N-balance improved from -17.5 ± 2.3 g in week 1 to 0.1 ± 4.0 g in week 4. Serum albumin was unchanged. Short term protein supplemented fasting results in rapid wt reduction with less loss of LBM than reported in fasting children, but more than that in adults on a protein supplemented fast. This loss of LBM in children may reflect increased K losses, decreased utilization of non-glucose substrates, lower insulin levels or higher essential amino acid requirements during the modified fast.

454 NUTRITION IN SMALL FOR GESTATIONAL AGE INFANTS (SGA): IN VIVO MEASUREMENTS OF BONE MINERALIZATION.

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In vivo quantitation of bone mineral content (BMC, gm/cm) by direct photon absorptiometry was used to evaluate the bone nutritional status in 24 SGA (26-42 wks, 880-2360 gms). Formula (1 batch) was fed and Vit D was standardized at 300-500 IU/day. Measurements were made at birth, 2,4,6,8 and 12 wks. BMC at birth in SGA did not correlate with gestational age, weight, length, or head circumference, whereas in appropriate for gestational age infants (AGA) there was a significant correlation of BMC at birth with gestational age and birth weight ($p < .001$). BMC at birth in 34-36 wk SGA ($.054 \pm .005$ gm/cm, mean $\pm$ SE) were comparable to 34-36 wk AGA. Postnatal bone mineralization in SGA was significantly less than intrauterine mineralization (covariance analysis) reaching $.092 \pm .004$ at 12 wks, equivalent to term AGA birth values, $.090 \pm .009$. In term SGA, BMC at birth was decreased $.067 \pm .008$ vs term AGA (t test $p < .01$). 12 wks of postnatal life were needed to reach term AGA birth BMC ($.090 \pm .005$). Thus, the postnatal increase of BMC for term SGA infants was less than for 34-36 wk SGA, $.023$ vs $.038$ gm/cm over a 12-wk period. Delayed bone mineralization in SGA occurred in spite of excellent weight gain. Mean Ca intakes in 34-36 wk and term SGA averaged 110-130 mg/kg/day, comparable to term AGA. Thus postnatal bone mineralization in SGA infants is decreased. Term SGA infants have decreased bone mineralization at birth, and require a prolonged period to achieve normal bone mineralization.

455 IDIOPATHIC INTESTINAL PSEUDO-OBSTRUCTION (IIP): MANOMETRIC STUDIES IN 2 CHILDREN.

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IIP is a functional bowel obstruction without known cause. Esophageal (EMS) and anorectal (RMS) pressure studies were performed on 2 boys with presumed IIP at age 9 yrs. (Case 1) and 11 yrs. (Case 2). Case 1 had chronic constipation from early infancy, bilateral hydronephrosis, recurrent obstructive episodes from age 8 and hemiplegia and aphasia later. EMS showed reduced lower esophageal sphincter (LES) tone unresponsive to bethanechol chloride (B) 0.1 mg/kg SC and metoclopramide (M) 0.15 mg/kg IV and a non-contractile esophageal body (EB). Case 2 had cerebellar ataxia and diarrhea from age 9 and obstructive episodes from age 10. Heller myotomy was done at 10 yrs. for presumed achalasia. EMS showed reduced LES tone and weakly contractile EB. B 0.1 mg/kg SC, Pentagastrin 1.0 and 0.5 ug/kg IV but not M 0.15 mg/kg IV increased LES and EB tone, and caused abdominal pain, vomiting and increased small bowel contractility. In both patients the anal sphincter did not relax with balloon distension of the rectum. In Case 1, rectal biopsies and bowel at autopsy at 9 yrs. were normal. In Case 2 autopsy at 11 yrs. showed cerebellar degeneration and reduced intramural nerves and ganglia especially at the LES. Although both patients seemed to have IIP with similar EMS and RMS, pathology and responses to pharmacologic stimulation were different. IIP may be a common manifestation of different physiologic disorders of the g.i. tract.

456 NARCOTIC DEPRESSION, FEEDING TECHNIQUES, AND LOWER ESOPHAGEAL SPHINCTER (LES) PRESSURES IN NEWBORN INFANTS.

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Previous reports suggest that neonates subjected to maternal narcotic depression (MND) and/or fed by conventional orogastric feeding techniques are at high risk for aspiration pneumonia. We have evaluated this risk by determining LES pressure in 16 neonates of 30-42 weeks gestational age (GA). Neonates were fed 20 cal/oz. commercial simlac in consecutive 24 hour time periods by intermittent gastric gavage, continuous drip gastric gavage, and by continuous drip duodenal gavage. Before termination of each feeding period, LES pressures were determined using a single lumen side opening perfused catheter system currently being investigated at our institution.

Narcotic depression was mimicked with 50 mg/kg body weight chloral hydrate given orally and LES pressure measurements were repeated in 90 min. After completion of LES studies at termination of the continuous drip duodenal period, the duodenal side opening feeding catheter was perfused with water at a rate of 1.3 ml/min. and pyloric sphincter (PS) pressures were recorded.

The results obtained revealed that LES pressures are not affected by GA, mimicked MND, or the type of feeding technique utilized. Although we have demonstrated a functionally competent PS in the preterm infant as immature as 30 weeks GA, all preterm infants studied exhibited a competent LES and no advantages of nasoduodenal drip feedings are apparent.