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REVERSIBILITY OF VITAMIN E DEFICIENCY NEUROLOGIC SYNDROME DURING CHRONIC CHILDHOOD CHOLESTASIS. Ronald J. Sokol, Mary A. Guggenheim, Susan T. Iannaccone, Christopher A. Miller, Arnold Silverman, James E. Heubi. Univ. of Colorado School of Medicine, Department of Pediatrics, Denver, and Children's Hospital Research Fdn., Dept. of Ped., Cincinnati.

To determine the reversibility of neurologic abnormalities due to vitamin E (E) deficiency, we prospectively studied the effects of E repletion on 14 children with chronic cholestasis, well-documented E deficiency, and characteristic sural nerve or muscle lesions: Group A (n=2)- both age 20 mo., no neurologic signs; Group B (n=3)- age <3 yrs., with characteristic neurologic signs; Group C (n=9)- ages 5-17 yrs., with neurologic signs. E repletion was achieved by oral (n=4; 60 to 90 IU/kg/day) or intramuscular (n=10; 1.0-1.5 IU/kg/day) dl- α -tocopherol. Child neurologists performed serial neurologic exams each 6-12 mo., assigning a neurologic score based on rating each of 12 neurologic signs from 0 (normal) to 3+ (severely abnormal). The mean neurologic scores for each group at 6 month time intervals were (Table):

Group	Neurologic Score (# Pts.) --- Duration of E Repletion			
	Baseline	6-12 mo.	12-24 mo.	24-36 mo.
A	0 (2)	0 (2)	0 (1)	--
B	4.7 (3)	1.7 (3)	0 (1)	--
C	15.6 (9)	12.8 (6)	12.8 (9)	8.3 (6)

Conclusion: Vitamin E repletion reverses or prevents neurologic abnormalities when begun prior to age 3 years, whereas only partial improvement occurs when therapy is begun after age 5 years. We recommend surveillance for and treatment of E deficiency prior to age 3 years in children with chronic cholestasis.

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TRANSIENT HYPERPHOSPHATASEMIA OF INFANCY. Robert N. Spady, Bruce B. Grill, John W. Mace, (Spon. by James J. Quilligan); Loma Linda University School of Medicine, Loma Linda University Medical Center, Department of Pediatrics, Loma Linda, CA.

To evaluate the clinical characteristics of transient hyperphosphatemia of infancy (THI), a syndrome of marked benign transient increase of serum alkaline phosphatase (SAP), we retrospectively studied eight patients with the following: 1) SAP > 3x upper limits of normal, 2) no hepatobiliary, metabolic, neoplastic, or osseous abnormality, 3) SAP level returning to normal without therapy. Mean age was 9.5 months, range 3-14 months; five patients were female. Mean maximum SAP was 2,460 IU/dl, range 1115-7276 IU/dl; mean time for SAP to normalize was 4.6 weeks (range 1.5 - 8 weeks); six patients (63%) had gastrointestinal symptoms (gastroesophageal reflux, food allergy, chronic diarrhea). One patient was < 5th percentile for weight. Serum calcium, phosphorus, transaminases, and bilirubin were normal. Skeletal radiographs and bone scans were normal. We conclude: 1) in infants, extreme elevations of SAP are often benign; 2) a minimal workup should include skeletal radiographs and liver function studies; 3) SAP should be repeated biweekly, and if decreasing extensive studies should not be performed. Because of high incidence of gastrointestinal (GI) symptoms we speculate that THI may be secondary to an acute GI injury with trans-mucosal passage of liberated intestinal alkaline phosphatase.

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CHANGES IN BODY COMPOSITION IN GROWING LOW BIRTH WEIGHT INFANTS D.W. Spady, D. Schiff, G. Chan. University of Alberta, Department of Pediatrics, Edmonton, Canada.

Body composition (BC) was estimated in 19 premature infants by measuring total body water (TBW) and extracellular fluid (ECF) using antipyrine and bromide. In 16 infants, measurements of nitrogen (N) balance were also made weekly. The diet was mother's milk (EBM), 8 cases, formula (F), 2 cases, and mixed EBM and F, 6 cases. In 13 cases, initial water measurements were made at a mean age of 19 days (weight (WT) 1226 +/- 254g) and finally at a mean age of 52 days (WT 2023 +/- 238g). In 6 cases only one measurement was made.

This was good correlation of weight with TBW (r=0.97) and ECF (r=0.94) with analysis yielding the equation TBW=43.73+0.742WT. There was a trend to more water retention with F feeding but small sample size precluded analysis. The mean protein (P) accretion was 11.82 +/- 3.17%. In cases with 2 water measurements, fat retention was estimated as follows: Fat-Dryweight-P retained. Mean fat retention was 11.41% +/- 9.4%. In these 'complete' cases there was a mean WT gain of 874g, of which 86.8% (671g) was water, 11.41% (100g) was fat, and 11.7% (103g) was P.

These estimates of nutrient retention are less than those made by others but are derived by more direct methods. This research underscores the need to perform direct BC measurements in growth studies.

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EFFECT OF RACE & NORMAL MATERNAL DIET ON BREAST MILK VITAMIN D CONCENTRATIONS, Bonny Specker, Reginald C. Tsang, Bruce Hollis, John Searcy, & Ron Levine, U. Cincinnati & Case Western U. Cleveland.

Differences in breast milk (BM) vitamin D by race and the influence of maternal vitamin D intake, in ranges normally consumed, on BM D have not been reported. We hypothesized that BM D or 25-hydroxyvitamin D (25OHD) concentrations were decreased in blacks, because of decreased skin production of D vs whites, and correlated with maternal D intake; and infant serum 25OHD is related to BM D content. All infants were less than 6 mos. and exclusively breastfed. BM and serum samples, and 3-d diet diaries, were obtained from 25 mother-infant pairs. D was purified by organic extraction and chromatography; metabolites were quantitated by ligand binding assay and HPLC. Mean total bioactivity from BM D and 25OHD was 51 IU/l with 25OHD contributing 77% (D 25 pg/ml = 1 IU/l, 25OHD 5 pg/ml = 1 IU/l). Mean (95% confidence) BM D concentrations for white and black mothers were 662 (400-1094) and 112 (61-206) pg/ml resp (t-test p<.001); BM 25OHD concentrations were 212 (171-263) and 156 (140-175) pg/ml (p=.03). BM D₂, D₃, and 25OHD₃ metabolites differed by race (p=.002, p<.001, and p=.03 resp); 25OHD₂ did not differ by race. BM D (r=.57, p=.005), but not 25OHD (p=.24) correlated with vitamin D intake (\bar{x} =457, range 0-706 IU/d). BM D₂ was correlated with D intake (r=.55, p=.006) even after control for race by multiple regression; BM D₃ was not correlated with D intake with race controlled. Infant serum 25OHD did not correlate with BM D or 25OHD (p=.11 & p=.33). Thus, black mothers have lower breast milk D₂, D₃, and 25OHD₃ than white mothers; breast milk D, in particular D₂, correlated with D intake in ranges normally consumed; infant serum 25OHD was not correlated with breast milk D or 25OHD.

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CONTRIBUTION OF PASSIVE PROXIMAL BILE SALT (BS) ABSORPTION TO ENTEROHEPATIC CIRCULATION (EHC) OF BS IN THE RAT. G.E. Stahl, Y-F. Shiau, J.B. Watkins. Divs. Neo. & Gastro, Univ. of PA Sch. Med., Philadelphia, Phila., PA.

Although the active transport of BS across the terminal ileum has been well documented, analysis of previous studies shows that ileal active transport cannot account for total BS recovery (Am J Physiol 244:G507,1983).

To evaluate passive BS absorption in the proximal small intestine, weanling male Sprague-Dawley rats (age 39 ± 4 days; wt 146 ± 26 g - mean ± SD) were anesthetized, the common bile duct was cannulated and the jejunum (J) and ileum (I) were catheterized and isolated. 1 ml of taurocholate (TC) solution in concentrations ranging from 0.5 mM to 10 mM and containing ³H-TC and ¹⁴C-polyethylene glycol (PEG - non-absorbable marker) were injected into the J. Bile was collected for 90 min. post-injection and counted. J and I were then ligated (to prevent mixing), divided into 8 segments, homogenized, and counted. TC absorption from the intestine was determined using ¹⁴C-PEG as a reference substance. In all animals, >94% of the ¹⁴C-PEG marker was found in proximal J (segments 1-3).

A linear correlation was found between the conc. of TC administered and the amount of TC recovered in bile (N=20, r=.91, p<.001). 8.4 ± 2.9 % of the dose was excreted in bile in 90 minutes. The rate of TC absorption was linearly related to TC conc. (N=13, r=.97, p<.001) and was non-saturable.

These findings demonstrate that TC is avidly absorbed by a passive process at physiologic concentrations (8-10 mM) in the proximal small bowel of the rat. When calculated for the entire small bowel, passive absorption accounts for between 54 and 69% of total BS secretion and is the major mechanism of BS conservation. These findings suggest that rapid proximal BS absorption rather than fecal BS loss is the mechanism for decreased intraluminal BS concentration in the neonate.

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THE EFFECT OF IMMATURITY ON INTESTINAL HANDLING OF COW'S MILK PROTEINS. Martin Stern, Kam-Yee Pang and W. Allan Walker, Harvard Medical School, Massachusetts General Hospital, Dept. of Pediatrics, Boston, MA. 02114

To investigate postnatal maturational changes of gut barrier function, we used two small intestinal in vitro models. Microvillus membrane (MVM) preparations were used for studying membrane surface binding and the everted gut sac technique was used for studying mucous coat binding and uptake of ¹²⁵I-labelled bovine serum albumin (¹²⁵I-BSA) and betalactoglobulin (¹²⁵I-BLG) in newborn, preweanling and adult rats. Surface binding of BSA and BLG to MVM was greater from newborn and preweanling rats, compared to adults (BSA, newborns: 2.74±0.53%; adults 1.08±0.17%, p<0.001). Binding however was weak and nonspecific. Contrary to surface binding, mucous coat binding of BSA and BLG to preweanling gut sacs was less than to adult ones, as was uptake (BSA binding: 0.94±0.30 vs 3.06±0.74 µg ¹²⁵I-protein equivalents /mg mucosal protein, p<0.001; BSA uptake: 0.35±0.17 vs 0.71±0.20, p<0.01). Gut sac binding and uptake were closely correlated in adult and preweanling animals (r=0.76, p<0.001 for BSA; r=0.85, p<0.001 for BLG in preweanlings). These studies suggest that immature animals are binding more cow's milk proteins to their MVM, but that less protein is bound to the mucous coat and taken up by immature gut sacs. Differential handling, mucous coat and MVM binding of food proteins in immature animals may be controlled by mucus layer factors which act to protect the underlying MVM.