

GASTROENTEROLOGY & NUTRITION

409 TRANSIENT CEREBRAL ATROPHY AND HYPERLIPIDEMIA IN AN INFANT WITH KWASHIORKOR FROM FOOD FADDISM. Raymond D. Adelman & Arthur B. Dublin, Departments of Pediatrics and Radiology, UC Davis (Spon. by Charles Abildgaard).

A case is presented of transient cerebral atrophy and hyperlipidemia in an infant with kwashiorkor. At age 2 1/2 months J.H. developed feeding problems from "milk allergy". Milk intake was reduced to 4 oz/day. Intake of baby fruits and vegetables increased to 14-16 jars/day. Anorexia with weight loss occurred over the next four months. Milk intake fell to only 4 oz/week and intake of fruits and vegetables to 4-6 jars/day. At age 8 1/2 months he developed apathy, vomiting, marked pallor, generalized edema, and hepatomegaly. Weight and height had fallen from the 90th %ile to <3rd %ile. On admission total protein was 3.4 gm%, albumin 1.9 gm%, SGOT 130 mU/ml, alkaline phosphatase 340 mU/ml, LDH 744 mU/ml, bilirubin 1.7 mg%, BUN 16 mg%, and creatinine 0.5 mg%. Serum cholesterol was 334 mg%; triglycerides 1580 mg%. 24 hour urinary protein was 60 mg. Liver biopsy showed hepatocytes filled with neutral fat. Computerized tomography (CT) of the brain showed cerebral atrophy with ventriculomegaly and increases in cortical sulci, sylvian fissures and the interhemispheric fissure. After 10 weeks of feeding with infant milk formula, weight increased from 6.3 kg (<3rd %ile) to 9.3 kg (50th %ile). Hepatomegaly and edema disappeared. Serum albumin rose to 4.5 gm%; serum cholesterol and triglycerides fell to 128 mg% and 60 mg% respectively. All liver function studies returned to normal. A repeat CT scan was normal. The patient at age 13 months has a mild developmental lag.

410 A NUTRITIONAL SURVEY OF CHILDREN WITH CEREBRAL PALSY. Raymond D. Adelman, Andrea Netten, Joan Edelstein, & Ronald Cohen, Dept. of Pediatrics, UC Davis, & Alta California Regional Center, Sacramento (Spon. by Charles Abildgaard).

29 non-institutionalized children under 6 years of age with cerebral palsy underwent nutritional assessment. Nutrient intake was estimated by both 3 day dietary records and 24 hour recall. Anthropomorphic measurements included height/length, weight, mid-arm circumference, and skinfold thickness (S.T.) of triceps and subscapular areas. 57% of children had caloric intakes <80% of the Recommended Dietary Allowance (RDA) for age; 40% had caloric intakes <80% of RDA for height age. Protein intake was generally adequate. 13% of children had protein intakes <80% of RDA for age. 9% had protein intakes <80% of RDA for height age. A significant number of children had subnormal height, weight, midarm circumference, and S.T.

	<50th %ile	<5th %ile
Height/length	27/29	18/29
Weight	29/29	20/29
Triceps S.T.	24/25	17/25
Subscapular S.T.	24/25	21/25
Midarm circ.	24/24	7/25

Mean values for hemoglobin and for serum iron, total protein, albumin, calcium, phosphorus, cholesterol, creatinine, and alkaline phosphatase were normal. Poor growth and undernutrition are common in children with cerebral palsy. Poor caloric intake may be a contributing factor.

411 THE EFFECT OF E. COLI ENDOTOXIN ON THE DEVELOPING RAT LIVER: Possible role in neonatal hepatitis. Joel M. Andres and W. Allan Walker. Harvard Medical School, Mass. Gen. Hosp., Pediatric GI Unit, Boston.

Little is known about toxic intrauterine insults associated with hepatic changes of neonatal liver disease. Attempts to study neonatal rat hepatotoxicity by toxin injections of pregnant rats has resulted in sporadic giant cell induction. This study used a direct route of injection of *E. coli* endotoxin (TOX) to investigate its hepatotoxicity and pathogenesis of neonatal hepatitis (NH). Fetal amniotic cavities of 16 day, timed-pregnant Sprague-Dawley rats were injected with TOX or with saline as controls. Five days later, fetal livers were removed and prepared for histologic evaluation. A significant increase ($p < 0.001$) in giant cell transformation was noted in TOX-injected fetuses (mean \pm SEM, 5.01 ± 0.61 cells/mm²) compared to controls (1.46 ± 0.14). A direct correlation was also noted between the number of giant cells and the concentration of injected TOX. Furthermore, the percent albumin (ALB) synthesis (immunoprecipitation of ¹⁴C-ALB) of total protein synthesis (¹⁴C-leucine incorporation into TCA-precipitated liver homogenates) was significantly lower in TOX-treated fetuses compared to controls ($p < 0.02$). In addition, preliminary studies showed serum alpha fetoprotein (AFP) levels in TOX-treated animals were greater than controls. This may be due to the reversion of the damaged liver to AFP synthesis and is consistent with elevated levels of serum AFP noted in NH. These findings suggest that TOX is a reliable fetal hepatotoxic agent and may be used as a model system to study the pathogenesis of NH.

412 FREE FATTY ACID METABOLISM DURING INTRALIPID INFUSION. G. Andrew*, G. Chan*, D. Schiff**. Department of Pediatrics, Obstetrics & Gynecology, University of Alberta, Edmonton.

Fatty acid composition of Intralipid (IL) differs from that of milk in that IL consists of 50% linoleic acid (L) and 10% palmitic acid (P) as compared with 8% (L) and 42% (P) in milk. To determine the effects of IL infusion on plasma free fatty acid (FFA) patterns, 6 neonates received 1 gm/kg of IL over 4 hours. The infants ranged from 32 - 38 weeks gestation and were of appropriate birth weight. Samples were obtained during infusion and at 2 hours post infusion for FFA pattern analysis by gas liquid chromatography. Results:

Time (hr)	FFA mg/dl mean \pm SEM	P/FFA mean \pm SEM	L/FFA mean \pm SEM	P/L mean \pm SEM
0	8.51 \pm 2.61	0.42 \pm 0.08	0.08 \pm 0.01	5.71 \pm 1.75
4	27.64 \pm 6.69	0.18 \pm 0.04	0.33 \pm 0.02	0.57 \pm 0.15
6	17.44 \pm 5.05	0.25 \pm 0.03	0.27 \pm 0.03	1.08 \pm 0.31

During infusion, the change in FFA concentration was significant ($p < 0.02$). There was a significant decrease in P/FFA ratio ($p < 0.01$) and increase in L/FFA ratio ($p < 0.001$) and reversal of P/L ratio ($p < 0.001$). In the post infusion period from 4 to 6 hours, there was a reversal of these altered ratios towards baseline values, but this change was not statistically significant. There are both quantitative and qualitative changes in plasma FFA during Intralipid infusion. These alterations reflect the fatty acid composition of Intralipid. Slow return of FFA to the normal pattern, after stopping Intralipid infusion, depends on the rate of cellular uptake or metabolism of the FFA.

413 FREE FATTY ACID (FFA) PATTERNS IN NEONATES RECEIVING INTRALIPID (IL). G. Andrew,* G. Chan,* D. Schiff.* Department of Pediatrics, Obstetrics & Gynecology, University of Alberta, Edmonton, Alberta, Canada.

FFA deficiency develops on fat-free intravenous alimentation (IVA). Minimum requirement for essential FFA is 0.5% of total calories. Milk contains 1.3% of total FFA as linoleic (L) and 23% as palmitic (P). IL contains 50% as L and 10% as P. The effects of IL as the only source of fat on sequential FFA patterns were determined in 6 infants (in the first 3 weeks of life) before and during two weeks of IVA and after milk had been re-established. A control group (C) received only milk.

GROUP	Day of IL	IVA gm/kg/day	Total FFA mg/dl	P/L \pm Sem	P/FFA \pm Sem	L/FFA \pm Sem
IL	0	0	7.57 \pm 2.00	6.52 \pm 1.04	0.37 \pm 0.11	0.06 \pm 0.01
	3	1	15.23 \pm 3.00	1.13 \pm 0.36	0.21 \pm 0.08	0.24 \pm 0.01
	7 - 10	3	8.81 \pm 0.95	1.02 \pm 0.01	0.25 \pm 0.01	0.27 \pm 0.04
	Milk for 1 week		15.17 \pm 1.54	1.08 \pm 0.02	0.17 \pm 0.08	0.25 \pm 0.07
	Milk for 2 weeks		10.76 \pm 1.35	0.73 \pm 0.30	0.22 \pm 0.04	0.33 \pm 0.02
C	(no.=39)	Milk	9.68 \pm 0.96	7.09 \pm 1.01	0.41 \pm 0.02	0.09 \pm 0.01

FFA deficiency was not observed. In infants receiving IL, FFA concentration was not significantly increased. Compared to milk-fed infants and to their own pattern before IL, the FFA pattern was significantly altered with increase in L/FFA ($p < 0.01$) decrease in P/FFA ($p < 0.05$) and reversed P/L ratio ($p < 0.001$). Changes were independent of the dose of IL, and persisted 2 weeks after IL was discontinued. Persistence of high L indicates mobilization of lipid that had its composition altered during IL infusion.

414 DISACCHARIDASE AND LYSOSOMAL ENZYME ACTIVITIES IN AMNIOTIC FLUID AND HUMAN FETAL SMALL INTESTINE. Irena Antonowicz and Aubrey Milunsky, HMS, CHMC, Boston

Brush border membrane-bound disaccharidases (lactase, sucrase, maltase) and lysosomal enzymes (α -glucosidase, β -D-fucosidase and N-acetyl- β -glucosaminidase) were studied in amniotic fluid (AF) obtained at 14th to 28th week of gestation from 176 mothers who delivered healthy babies. AF enzyme activities were correlated with gestational age and the activities of the same enzymes in fetal intestinal mucosa, on a week by week basis. Disaccharidase activities in AF decreased with advancing gestational age. The same pattern was also found for lysosomal α -glucosidase and β -D-fucosidase while no marked changes in the activity of N-acetyl- β -glucosaminidase were noted. The maximal activities for sucrase, maltase, α -glucosidase, β -D-fucosidase and N-acetyl- β -glucosaminidase were about 10.0, 25.0 units/min/gP, 11.0, 150.0 and 155.0 units/hr/gP respectively at 16th week of gestation and decreased to nearly zero values at 26th week except for N-acetyl- β -glucosaminidase. Comparison of disaccharidase activities in intestinal mucosa and AF showed that sucrase and maltase were 6 to 9 times higher in the intestinal mucosa between the 14th and 20th week of gestation. By contrast, the lysosomal enzyme, α -glucosidase, demonstrated comparable activity (1:1 ratio) in intestinal mucosa and AF during the same period of gestation. The differences in distribution between AF and intestinal mucosa for brush border bound enzymes and α -glucosidase may reflect the ubiquitous origin of lysosomal α -glucosidase, only intestinal mucosa produces disaccharidases which are segregated in utero and only available to the AF by exfoliation.