1153 OBESITY IN CHILDREN. RELATIONSHIP BETWEEN ZINC (ZN) TRIGLYCERIDE (TG) AND TESTOSTERONE (T). S. Ziaadin Ghavami-Maibodi, Mariano Castro-Magana, Sanda Clejan, Shang Y. Chen, Vaddanahally T. Maddaiah and Platon J. Collipp. Nassau County Medical Center, SUNY, Stony Brook Health Sciences Center, Department of Pediatrics, E. Meadow, NY 11554.

Childhood obesity is caused by a variety of genetic and environmental factors which have been difficult to identify. We have recently found that some obese children have reduced levels of hair ZN. Our studies in rats and in adults with coronary artery disease have confirmed an inverse relationship between nutritional ZN status and serum TG levels. We have also reported a direct relationship between serum T and Zn nutritional status. We are here reporting data indicating that obese children can be divided into two groups. In Group 1 the serum TG level and hair ZN concentrations are significantly higher than in Group 2, and only in Group 1 is there a direct correlation between serum T and hair ZN. The cause of the lack of a direct relationship between ZN and T in Group 2 obese children is not known.

		Correlation Coefficient				
Group 1	Age	TG vs ZN	T vs ZN	Zinc	T	TG
(M 14,F 19)	10.4	-0.6300	+0.5193	216 ± 50*	127 ± 80	115 ± 56*
Group 2						
(M 12,F 14)	10.9	-0.5205	-0.1682	141 ± 27*	129 ± 80	85 ± 26*

Thus, obese children also had an inverse relationship between serum TG and hair ZN (*$p < 0.005$).

1154 PROTEIN-BOUND PLASMA CYST(E)INE: NUTRITIONAL AND METABOLIC SIGNIFICANCE. M.H. Malloy, D.K. Rassin and G.E. Gaull. NYS Inst. Basic Res. Mental Retard., Staten Island, NY, Dept. Ped., Mt. Sinai Sch. Med., CUNY.

A simple method for the measurement of plasma free, bound and total cyst(e)ine has been developed. Using this method, we found that plasma total cyst(e)ine is distributed equally between that bound to plasma protein (10.5 ± 4.0 μmoles/dl, mean ± S.D.) and that free (9.9 ± 2.5); compare with amino acid analyzer free ½-cystine (10.7 ± 2.2). Radioactive studies show the cyst(e)ine is bound mainly to albumin *in vivo* and is not an artifact of isolation.

Cyst(e)ine may be displaced from plasma protein by compounds that compete for the available sulfhydryl groups of plasma proteins, *e.g.* in 5 patients with homocystinuria, homocysteine was the predominant sulfhydryl compound bound to plasma protein. In addition, nutrition can affect both free and bound cyst(e)ine in the plasma, *e.g.* rats starved for 8 days had a significant decrease both in plasma free cyst(e)ine and bound cysteine.

We conclude: (1) Previous methods for the measurement of plasma cyst(e)ine underestimate the quantity of cyst(e)ine in the plasma. (2) The distribution of plasma cyst(e)ine between the bound and the free form may be influenced by the nutritional or metabolic state.

● 1155 NORMOTRIGLYCERIDEMIC ABETALIPOPROTEINEMIA: FATE OF APOLIPOPROTEIN B OF CHYLOMICRONS. Mary J. Malloy and John P. Kane. (Sponsored by Carolyn P. Piel), Cardiovascular Research Institute and Dept. of Pediatrics, University of California, San Francisco, California.

In this disorder (NA), normal plasma low density lipoproteins (LDL) are absent but intestinal absorption of fat is normal. Levels of spherical, triglyceride-rich lipoproteins, 300 to 1000 Å in diameter, rise markedly in plasma during fat absorption. Though 40% of their protein is insoluble in 4.2M tetramethylurea, it has only limited immunoreactivity with antibodies to apo B of LDL. Its apparent molecular weight (MWapp) in SDS is identical to that of the B-48 protein of normal human chylomicrons and it has a similar amino acid composition. The B-100 protein found in normal very low density lipoproteins and LDL, which has a much larger MWapp, is undetectable. This suggests that the B-48 and B-100 proteins are under separate genetic control and that NA results from selective deletion of the B-100 apolipoprotein. We purified chylomicrons from serum of a patient with NA, labeled them with ^{131}I, reinfused them and isolated B-48 protein from serum lipoproteins by SDS gel electrophoresis. The label disappeared with a $t_{\frac{1}{2}}$ of 52 minutes. Minimal labeling was detected briefly in lipoproteins of intermediate density (12% of total ^{131}I at 2 minutes, none thereafter) and none in the LDL interval. We conclude that the B-100 apolipoprotein is required for formation of LDL and that the B-48 apoprotein of chylomicrons is not converted to apo B of LDL.

● 1156 POLYOL PATHWAY HYPERKINESIS IN CHILDREN WITH DIABETES. John I. Malone, Gail F. Knox, Christopher Harvey. University of South Florida, College of Medicine, Dept. of Pediatrics, Tampa, FL.

Intracellular sorbitol accumulation has been implicated as an etiologic factor for many of the sequelae of diabetes mellitus. The human erythrocyte is a tissue that accumulates sorbitol in response to extracellular glucose concentration. Red cell sorbitol levels in children with diabetes were compared to those in red cells from non diabetics. Sorbitol was measured by a fluorometric assay utilizing sheep liver sorbitol dehydrogenase; glucose was measured by glucose oxidase. Red cell sorbitol in 140 children with diabetes (36.6±1.7nmols/gm Hb) was greater than in 50 non diabetics (10.3±.79) p<.0005. The influence of extracellular glucose was taken into account by expressing intracellular sorbitol in terms of plasma glucose concentration (S/G). The S/G ratio for red cells from diabetics was 20.6±.88 and non diabetics was 12.1±.95 p<.0005. When red cells from diabetics were exposed to non diabetic glucose concentrations for up to 14 hours the S/G ratio did not return to non diabetic levels. Non diabetic red cells exposed to elevated glucose concentrations (300 and 500 mg/dl) for 3 hours did not manifest elevated S/G ratios. Elevated S/G ratios were found in association with non diabetic hemoglobin A_{1c} values suggesting long term exposure to near physiologic glucose levels. Increased sorbitol production was found in most, but not all individuals with diabetes and seems to be an acquired characteristic. This is the first evidence of altered polyol pathway activity not directly related to substrate availability in spontaneous human diabetes.

1157 INFLUENCE OF CORTICAL BONE MINERALIZATION ON PERIOSTEOCYTIC LESIONS IN VITAMIN D-RESISTANT RICKETS. P.J. Marie* and F.H. Glorieux. Genetics Unit, Shriners Hosp., McGill University, Montreal, Canada.

Hypomineralized periosteocytic lesions (HPL) are peculiar bone features present with osteomalacia in vitamin D-resistant rickets (VDRR). In order to determine whether HPL result from defective bone mineralization, tetracycline dual labeled iliac crest bone specimens from VDRR children (4-16 years old) were analyzed before (n=6) and after treatment with phosphate supplements combined to vitamin D_2 (Pi + D, n=6) or to 1,25-dihydroxyvitamin D_3 (Pi + 1,25, n=13). The percentage of osteocytes with HPL was determined on microradiographs of undecalcified sections in the interstitial cortical bone (IB), the well calcified resting osteons (RO) and the less mineralized growing osteons (GO). Bone mineralization was assessed on the same samples by measuring the mineralization lag time (MLT), the osteon calcification rate (CR) and the bone formation rate (BFR). Although HPL were more prominent in less calcified bone areas (GO: 53.9%, RO: 43.4%, IB: 21.8%), no correlation was found before treatment between HPL frequency and bone mineralization parameters. In contrast to Pi + D, Pi + 1,25 reduced HPL frequency in GO (40.6%), RO (37.6%) and IB (12.8%) in correlation with improved bone mineralization (ΔHPL - ΔCR or ΔHPL - ΔBFR: $r = -90$, $p<0.0005$, ΔHPL - ΔMLT: $r = 0.66$, $p < 0.025$). However, HPL persisted in more than 30% of new osteocytes in GO despite normalization of bone mineralization. The data show that HPL result in part from defective bone mineralization. However, HPL were unrelated to the severity of osteomalacia and were persisting in the new well calcified bone, indicating that an abnormality of the osteocytic function, consequence of the proposed osteoblastic defect, is part of the VDRR phenotype.

1158 ZINC (Zn) AND COPPER (Cu) METABOLISM IN $CaNa_2EDTA$-TREATED CHILDREN WITH PLUMBISM. Morri E. Markowitz, John F. Rosen, Albert Einstein Coll. Med., Montefiore Hosp. & Med. Ctr., Dept. Ped. & CRC, Bronx, N.Y.

$CaNa_2EDTA$ is the primary agent in the treatment of plumbism in children. In vitro studies have shown that this agent also chelates several essential trace elements including Zn. We investigated the effects of a 5-day course of $CaNa_2EDTA$ (1000 mg/m^2/day) on serum Zn and Cu concentrations. 13 children with varying degrees of plumbism (blood Pb: 40-71μg/dl) had serum Zn and Cu concentrations measured 1 day before and 1 day after 5 days of therapy. In addition, 6 of these subjects had recovery (rec) phase levels drawn within 1 month of therapy.

Results:	Pre-Rx	Post-Rx	Rec	t(pre/post)	t(pre/rec)	controls
Cu	154 ± 34^a	164 ± 40	156 ± 49	1.43	-.811	148 ± 18^a
Zn	81 ± 18^{ab}	50 ± 15^b	83 ± 5^b	13.1[c]	-.302	102 ± 8^a

a=μg/dl, mean±SD; b=sign. different vs. controls, p<.001; c=p<.001; t=paired t test

Conclusions: 1) Mean serum Zn concentration in children with plumbism is significantly lower compared to normal children; 2) $CaNa_2EDTA$ treatment transiently and significantly further decreases serum Zn levels; 3) Return to pretreatment Zn levels occurs less than 1 month after $CaNa_2EDTA$ therapy; 4) Serum Cu concentration is unaffected by increased blood lead concentration or $CaNa_2EDTA$ therapy; 5) Zn and Cu supplementation to correct any $CaNa_2EDTA$-induced abnormalities appears unnecessary; 6) These data suggest that interactions between Zn and Pb may be of importance in the clinical expression of lead toxicity.