1147 PLASMA SOMATOMEDIN IN CYSTIC FIBROSIS ADOLESCENTS IS NORMAL. Christopher Landon, Ron G Rosenfeld, Ray Hintz, Norman Lewiston, and Ray Nagashima. Childrens's Hospital at Stanford, Palo Alto, California.

Growth retardation and malnutrition are common manifestations of cystic fibrosis (CF). Since chronic malnutrition is generally characterized by reduced somatomedin (SM) activity despite normal levels of growth hormone (GH), defects in the hypothalamic-pituitary-SM axis have been proposed as etiologic in the growth retardation of CF. Accordingly, we have measured GH, insulin, and SM levels in 15 adolescents with CF and compared them with 9 age matched controls of normal height and weight. All CF subjects were below the 30th percentile in height with 9/15 below the 5th percentile in height and 12/15 below the 5th percentile in weight. Weight to height percentile ratio was 50% or below in 11/15. Fasting 8 AM GH values were similar in both groups: 5.33±1.16 ng/ml (mean±SEM) in CF and 5.76±1.33 ng/ml in controls. Fasting insulin levels were moderately reduced in CF patients: 8.07±0.80μU/ml versus 12.94±1.89μU/ml ($p<0.01$) although there was no evidence of clinical diabetes. The total plasma SM content, measured by placental radioreceptor assay following G-50 chromatography in formic acid was normal or elevated in all CF patients. SM levels ranged from 0.93-3.54 U/ml, with a mean value of 1.92±0.23 U/ml; control values ranged from 0.89-2.49, with a mean of 1.67±0.20 U/ml. This demonstration of normal plasma SM in subjects with CF contrasts with previous reports of decreased bioassayable SM. We suggest that the H-P-SM axis in CF is intact despite a clinical malnutrition, and that growth retardation may reflect a defect in cellular responsiveness to SM peptides.

1148 MITOCHONDRIAL ABNORMALITIES IN A MALE WITH ORNITHINE TRANSCARBAMILASE DEFICIENCY(OTC). R.Longhi, C.Buttè, R. Valsasina, L.Rossi, M.Borzani, (spon. F.Seroni), Università di Milano, Clinica Pediatrica I°, Milan, Italy.

The case of a 4 yr.old boy with partial deficiency of hepatic OTC is presented. This X-linked dominant disease is lethal, for males, in the newborn period. Our patient presented with lethargy, ataxia and irritability, not preceded by signs of viral infection. Laboratory tests were normal with the exception of metabolic acidosis, hyperammonemia (257γ/100cc) and oroticaciduria (1680γ/mg creat.). The clinical picture progressively worsened to coma, and death occured on the 13th day of hospitalization. A liver biopsy on the 12th day showed a high residual OTC activity (10,9% of controls). All the other urea cycle enzymes were normal. No fatty liver degeneration was present, but an E.M. study showed the mitochondria to be rounded with loose matrix and only a few short cristae. An E.M. kidney biopsy examination showed fatty degeneration of renal tubules, but no mitochondrial involvement. Reye's syndrome was ruled out for various reasons such as the atypical clinical course, normal liver function tests, delayed death and absence of fatty liver degeneration. Abnormalities of liver mitochondria have not been previously reported in male patients. The claims that mitochondrial abnormalities support the diagnosis of Reye's syndrome versus OTC deficiency have to be reconsidered.

●1149 ALTERED Na, K ATPase IN SPONGY DEGENERATION (CANAVAN'S DISEASE). Ira T. Lott, David S. Reiss, and Victor S. Sapirstein. Harvard Med Sch, E.K. Shriver Ctr-Mass Gen Hosp, Depts Ped/Neurol, Boston.

Spongy degeneration (SD) of Van Bogaert and Bertrand in infancy is characterized by white matter edema and splitting of myelin at the major dense line. The cause of myelin breakdown in this disorder is believed to be secondary to the edema. Na, K ATPase activity was studied in 3 SD brains since inhibition of this enzyme is associated with edema. Na, K ATPase activity in homogenates from frontal cortex was reduced 35-60% in SD brains compared to leukodystrophic and non-neurological controls. The decreased enzymatic activity in SD was not due to altered affinity for either the activating cations Na and K or for the substrate ATP. Measurements of /^{3}H/ ouabain binding indicated that SD brain had approximately 2X the number of enzyme sites per mg membrane protein as compared with controls. The ATPase per enzyme site (molecular specific activity) in SD cortex was only 29-50% that of controls. In the single sample of SD kidney available, ATPase activity was normal. These data suggest that an attenuation in the brain specific form of NA, K ATPase may be related to the pathogenesis of intralamellar myelin edema in SD. (Supported in part by USPHS grants HD 05515 and HD 04147)

1150 TOTAL PROTEIN (TP), 3-METHYLHISTIDINE (3-MeH), AND FREE WATER CONTENT OF SKELETAL MUSCLE IN FETUSES AND INFANTS AT VARYING GESTATIONAL AGES. Victor Lunyong, Zvi Friedman, Baylor College of Medicine, Department of Pediatrics, Houston, Texas.

Urinary excretion of 3-MeH provides an index of the rate of myofibrillar protein breakdown. In order to assess fractional turnover of muscle protein in infants, the content of 3-MeH was analyzed in skeletal muscle obtained from two fetuses and four infants of 15 to 38 weeks gestation (GW). Wet and dry weight (wwt/dwt) were measured from duplicate specimens of the psoas and femoral muscle of each fetus and infant. One set of the muscle specimen was hydrolyzed and assayed for 3-MeH and total nitrogen content by high pressure liquid chromatography and micro-Kjeldahl technique respectively. The results are as follows:

Groups	No. of sample	Gest. age (Wks)	dwt/wwt (%)	TP/wwt (%)	TP/dwt (%)	3-MeH/g.TP (μmole/g)
I	2	15-18	10.4	9.0	78	1.11
II	4	26-32	12.4	10.1	82	2.64
III	3	37-38	14.8	11.7	79	3.68

Conclusion: 1. Free water content of muscle decreases with advancing gestational age. 2. The higher content of 3-MeH in muscle in advanced gestation may reflect either an increase proportion of myofibrillar protein of skeletal muscle or an increase methylation of histidine in myofibrillar protein molecule. This information is useful for the measurement of the fractional turnover of muscle protein in infants.

1151 ZINC (ZN) IRON (Fe) NUTRITIONAL STATUS IN OBESE CHILDREN. S. Ziaadin Ghavami-Maibodi, Shang Y. Chen, and Platon J. Collipp. Nassau County Medical Center, SUNY, Stony Brook Health Sciences Center, Department of Pediatrics, E. Meadow, NY 11554.

Forty-five obese children, (>2SD above ideal weight), were divided into two groups based on their hair Zn concentration. Twenty-four children with hair Zn below 180 ug/gm, had significantly higher Hgb, Hct, serum Fe:Fe-binding capacity, and significantly lower urine Fe, than twenty-one other children with hair Zn above 180 ug/gm.

		Zn		Fe				
	Age	Hair	Urine	Hair	Urine	Fe:FeBC	Hgb	Hct
Group 1 (M10,F14)	11.4	150±17	575±238	38±18	49±49	26±6	14.1±0.6	42±3
Group 2 (M6,F15)	11.3	231±33 $p<0.001$	600±294 $p<0.8$	27±14 $p<0.1$	150±85 $p<0.05$	19±6 $p<0.005$	13.5±0.7 $p<0.05$	38±2 $p<0.005$

Hair, serum, and urine copper concentration was also determined in both groups and were not significantly different. This inverse relationship between Zn and Fe, has also been seen in children with pure red cell anemia and Cooley's anemia.

1152 FAMILIAL ZINC (Zn) DEFICIENCY. S. Ziaadin Ghavami-Maibodi and Platon J. Collipp. Nassau County Medical Center, SUNY, Stony Brook Health Sciences Center, Department of Pediatrics, East Meadow, NY 11554.

Some children with constitutional growth delay (CGD) and children with obesity have been found to have low levels of Zn in their hair. Some of them have the physical features which have been reported in Zn deficiency (leuconychia, abdominal striae, hyperkeratosis, sparse hair, etc.). We have noted similar findings in some of their relatives and have now studied the familial pattern of low Zn levels. Children with either condition who have low hair Zn levels have families with significantly ($p < 0.01$) low hair Zn levels. It is not clear whether this is hereditary or dietary or both but in 304 families we did not find any correlation in Zn levels between husbands and wives, suggesting that heredity may be important in determining hair, serum and urine Zn.

Children	CGD(14)	Controls(125)	Obesity(48)	Obesity(43)
Age	11.5	12.0	10.7	10.3
Hair ug/gm	134±22	186±23	150±17	231±33
Blood ug/ml	100±20	105±23	137±47	160±43
Urine ug/gm C	701±245	-	575±238	600±299
Parents				
Hair	144±38	177±42	170±33	210±40
Blood	118±35	110±20	-	-
Siblings				
Hair	143±48	-	-	-
Grandparents				
Hair	145±38	-	-	-