1165 EPISODIC HYPOGLYCEMIA WITH ORGANIC ACIDURIA. J.D. Miller, E. Colle, O.A. Mamer, J. Montgomery. McGill Univ. Mass Spectrometry Unit-Montreal Children's Hospital, Dept. of Pediatrics, Montreal, Quebec.

Two female children with febrile episodic hypoglycemia(glucose 10 and 34 mgm/dl) and elevation of SGOT (103 and 163 U/L) had increased amounts of urinary C6-10 dicarboxylic fatty acids as well as the ω-1-OH and ketoderivatives of these acids. Acetoacetate and β-OH butyric acid are only moderately elevated.

The compounds are present between attacks, increase after a medium (MCT) or long (LCT) chain triglyceride load and are not found in the urine of normal children. Young infants with ketosis will show dicarboxylic aciduria but only in the presence of greatly increased acetoacetate, β-OH butyric and lactic acid.

Urinary Organic Acids (mgm/gm creatinine)

	First Patient				Second Patient		
	First Episode	Second Episode	MCT	Health	(5 days post) Episode	MCT	Health
Acetoacetate	20	930	13	19	-	-	-
3OH Butyrate	26	485	20	21	-	70	-
5OH Hexanoic	70	120	13	26	8	3	3
7OH Octanoic	30	270	540	39	7	340	2
Adipic	80	630	49	27	140	100	8
Suberic	145	430	361	135	86	215	2
Sebacic	-	415	378	21	130	110	-

The data suggest a deficiency of medium chain acyl coA dehydrogenase activity limiting β-oxidation of fatty acids which becomes significant during periods of increased energy demands.

1166 LIPID AND LIPOPROTEIN CORRELATIONS IN BLACK AND WHITE ADULT AND CHILD SIB PAIRS. J.Morrison, R.Horvitz, K. Kelly, P.Khoury, P.Laskarzewski, C.J.Glueck. Univ. Cincinnati, College of Medicine. LRC, GCRC, Cincinnati, Ohio.

To compare intrafamilial associations among siblings(S) who still share and no longer share a common household environment, S-S correlations for total cholesterol(TC), high and low density lipoprotein cholesterol(C-HDL,C-LDL), and triglyceride(TG) were calculated.For the 54 black(B) and 230 white(W) adult(>20yrs old) (A) S-S pairs, and for the 97 B and 274 W child (6-19 yrs old)(C) S-S pairs, the correlations (*p<.02,**<.01,†<001) were:

	C-LDL B	C-LDL W	C-HDL B	C-HDL W	TC B	TC W	TG B	TG W
A S-S	.534†	.471†	-.06	.159*	.508†	.508†	.353*	.378†
C S-S	.363*	.451†	.608†	.374†	.259**	.395†	.332†	.110

There were no consistent B-W differences in lipid-lipoprotein correlations in A or C S-S pairs. S-S correlations for C-LDL, TC, and TG were either comparable or stronger in A S-S than in C S-S pairs, despite the loss of shared households in the A S-S pairs. This suggests a lasting and persistent influence of genetic factors on C-LDL and TG, or, speculatively, persistence into adulthood of shared environmental effects. In contrast, S-S correlations for C-HDL were stronger in C than in A S-S pairs, suggesting a substantial influence of environmental factors on C-HDL, revealed when common households are no longer shared. Explication of environmental factors which affect C-HDL might be realized by longitudinal studies of lipoproteins from childhood to adulthood.

●1167 ADRENOLEUKODYSTROPHY: ABOVE NORMAL LEVELS OF VERY LONG CHAIN FATTY ACIDS IN PLASMA: IMPLICATIONS FOR DIAGNOSIS AND DIETARY THERAPEUTIC TRIAL. Hugo W. Moser, Ann E. Moser, Mary Ann Van Duyn, Daniel Stowens, John Barranger, Joseph D. Schulman. J.F. Kennedy Institute, Baltimore; National Institutes of Health, Bethesda, MD.

Adrenoleukodystrophy (ALD) is a progressive X-linked disorder associated with accumulation of saturated very long chain fatty acids in brain and adrenal. We have used a capillary gas liquid chromatography method to quantitate each of the saturated very long chain fatty acids (C20-C26) in the plasma total lipids of 18 adrenoleukodystrophy (ALD) hemizygotes, 17 obligate ALD heterozygotes and 50 normal and disease controls. C26 levels in the controls were .015% ± .0032 (SEM) of total fatty acids with chain length in excess of C14, compared to .081% ± .0066 in the ALD hemizygotes and .057% ± .0063 in heterozygotes. Differences were significant with p value <.001. C23, C24 and C25 levels were also significantly increased in the ALD patients. The procedure permits the diagnosis of the ALD hemizygote, even presymptomatically, and aids the identification of the carrier. Since there is evidence that the accumulated fatty acids are of dietary origin (Bioch. Bioph. Res. Comm. 96:69, 1980) we have developed a diet which contains 3 mg of C26 fatty acid per day instead of the usual 15-40 mg. This diet brought about a rapid reduction of plasma C23 and C25 levels in one patient.

●1168 CORD PLASMA C-PEPTIDE/GLUCOSE [C-P/G] IN INFANTS OF DIABETIC MOTHERS [IDM]: RETROSPECTIVE AND PROSPECTIVE INDEX OF B-CELL FUNCTION. Edward S. Ogata, Boyd E. Metzger, Norbert Freinkel (Spon. by C. Hunt). Northwestern University Medical School, Prentice Women's Hospital, Departments of Pediatrics, Ob Gyn, Medicine; Chicago.

We have reported evidence of ↑ fetal B-cell function [C-P/G] in umbilical cord plasma from IDM, including gestational diabetics with minimal degrees of glucose intolerance. In this report, we examined the relationships between C-P/G to antenatal (metabolic environment and intrauterine growth) and postnatal (glucose homeostasis) events in 87 term IDM (White Classes A-D) with uncomplicated neonatal courses. Retrospectively, maternal hemoglobin A_{1c} (HbA_{1c}) at term correlated with C-P/G (r=.3450, p<.01) and adiposity as reflected in skin fold thickness measurements (p<.05) and weight/height ratio (p<.01). Thus, even in tightly regulated diabetics, gradations of metabolic control reflected in HbA_{1c} elicit ↑ B-cell function which is expressed in birthweight and adiposity. Prospectively, C-P/G correlated with the nadir of plasma glucose in the first 4 hours of life (r= .4976, p<.01) as well as with rate of glucose disposal (K_t r= .5830, p<.01) and acute B-cell stimulatory responses (ΔC-P r= .7128, p<.01) in a subgroup of IDM challenged with intravenous glucose at 2 hours of life. Thus, cord C-P/G relationships at the pivotal transition between in utero and extrauterine life are sensitive measures of ongoing B-cell function providing a retrospective index of antenatal fetal metabolic environment and augmented growth and a prospective index of altered neonatal glucose homeostasis.

1169 EXERCISE IN THE HEAT IN CYSTIC FIBROSIS PATIENTS. David M. Orenstein, Kathe J.Germann, David L.Costill, Peter W. Lemon. (Spon. by Robert C. Stern). Case Western Reserve University, Rainbow Babies & Childrens Hospital, Cleveland.

Cystic fibrosis (CF) is characterized by high sweat sodium (Na) and chloride (Cl), yet little is known about CF patients' response to exercise in the heat. We examined the responses of 5 CF patients with mild lung disease and 3 non-CF controls (N) to single sessions of cycle exercise in the heat. Subjects had free access to water, and pedalled at 50% peak VO_2 for 60-90 minutes in a heat chamber at 37-38°C (DB), 24-29°C(WB). Urine and serum osmolality and electrolytes were measured before and after exercise, as were serum renin and aldosterone. Sweat volume, rate, and electrolytes were measured. Rectal temperatures (Tr) and ECG were monitored continuously. N and CF had similar Tr (101.1 ± 1.3, 101.0 ±.08, p>.5), peak HR (175 ±16, 165 ±12, p>.25), sweat rates (5.8 ±2.0, 8.1 ±2.1 $ml \cdot m^{-2} \cdot min^{-1}$, p>.1). CF had the expected higher sweat Na (76 ±24 $mEq \cdot L^{-1}$) and Cl (70±21$mEq \cdot L^{-1}$) than control (44 ± 5 and 37 ± 7 $mEq \cdot L^{-1}$, p <.05). CF showed a normal rise in renin (6.8 before, 21.4 $ng \cdot ml^{-1} \cdot hr^{-1}$ after, p<.05) and aldosterone (13.6 before, 45.4 $ng \cdot dl^{-1}$ after, p<.05) with exercise. CF urine Na fell with exercise (172.2 to 119.8 $mEq \cdot L^{-1}$, p<.05). Despite this evidence of renal salt conservation, CF serum Cl fell (102.4 to 98.8 $mEq \cdot L^{-1}$, p <.05) while control serum Cl did not change (104 $mEq \cdot L^{-1}$) and CF serum osmolality fell (272.2 to 263.8 $mosm \cdot L^{-1}$, p<.05). Even though CF patients can mount hormonal responses to exercise/heat stress and can conserve salt through renal mechanisms they lose more salt than normal people. Prolonged or repeated exercise in the heat may be dangerous for CF patients.

1170 STUDIES WITH A NEW COMPETITIVE INHIBITOR OF α-GLUCOSIDASES. Stephanie S. Padilla, Peter H. Burrill and James B. Sidbury, Neonatal and Pediatric Medicine Branch, NICHD, Bethesda, MD.

Lysosomal α glucosidase, deficient in type II, GSD, has not been fully characterized in the diseased state. Inhibition by turanose and stimulation by high concentrations of KCl have been used to distinguish the lysosomal pH4 enzyme from the microsomal bound enzyme with pH optimum 6.5. In the human placenta the Km of the pH4 enzyme is 1.1 mM whereas the pH6.5 enzyme is 12. μM, utilizing 4 methylumbelliferyl-α-D-glucopyranoside. The low Km pH 6.5 activity was not found in urine. A new competitive inhibitor was found to be useful in distinguishing the two enzymes: Bay G-5421 is a complex oliogosaccharide, containing an unsaturated cyclitol unit bound to 4,5-didioxy-amino-D-glucopyranae within a chain of α-1, 4-glucopyranose units. The KI for the pH4 enzyme was .05 mM, whereas that for the pH 6.5 enzyme was 1.5 mM. Urine enzyme showed only the pH4 type inhibition. The urine enzyme could be concentrated by percipitation with 50% alcohol.

This technique will be applied to tissues from patients with late onset type II GSD who have reduced amount of enzyme with an altered Vmax. This additional tool should be helpful in defining heterogeneity in the late onset form.

It is hoped that urinary enzyme assays may be useful in detecting heterozygotes.