1176 DISSOCIATION OF CHLORIDE (Cl^-) AND SULFATE (SO_4) PERMEABILITIES IN THE CYSTIC FIBROSIS (CF) SWEAT DUCT. D.E.C. Cole and D.A. Landry (Spon. by J.F.S. Crocker) Dalhousie University, Dept. of Pediatrics, Halifax, N.S., Canada

Defective anion transport has recently been put forward as the proximate cause of elevated Cl^- in CF sweat. To further explore this possibility, we studied the behaviour of a Cl^- analogue, inorganic sulfate (SO_4), in serum and sweat obtained from controls (n=22) and adult CF patients (n=6). A minimum of 50 mg of sweat was collected from both forearms onto acid-washed, low-sulfate absorbent disks after the standard pilocarpine electrophoresis. Cl^- and SO_4 were measured by conductimetric means on the same sample using controlled-flow anion chromatography. Serum Cl^- and SO_4 were similar in controls and CF patients. In controls, sweat SO_4 and Cl^- were significantly correlated (r=.52, p<.01). Also, controls had sweat:serum concentration ratios for SO_4 (0.28±0.02, $\bar{x}$±SE) that were not different from Cl^- (0.26±0.01), suggesting that both anions are reabsorbed to a similar extent during the formation of the final secretory product from serum ultrafiltrate. As expected, Cl^- was raised in CF sweat (104±3 mmol/L vs 19±2 mmol/L in controls, p<.01); however, sweat SO_4 was significantly lower in CF patients (48±5 µmol/L) than in controls (81±3 µmol/L, p<.01). Although SO_4 and Cl^- are known to share the anion exchange system in the red cell membrane and are apparently handled similarly in the normal sweat duct, their transport by the CF sweat duct is not the same. Any model of defective anion exchange in the CF sweat gland must take this dissociation of SO_4 and Cl^- permeabilities into account.

●1177 HEPATIC MONOACYLGLYCEROL ACYLTRANSFERASE ACTIVITY IN SUCKLING RATS. Rosalind A. Coleman and Elaine B. Haynes, Department of Pediatrics, Duke University Medical Center, Durham, North Carolina 27710

Hepatic monoacylglycerol acyltransferase (MGAT) activity is 700-fold higher during the suckling period than in the adult rat. Specific activity in total particulate preparations rose from 9.4 nmol/min/mg before birth to a peak of 78 nmol/min/mg on the 6th postnatal day. MGAT activity fell sharply after day 8 and was 1.6 and 0.1 nmol/min/mg on day 28 and in adult rats, respectively. The activity had a pH optimum at 8.0 and was activated by albumin and by phospholipids. With (^{3}H)palmitoyl-CoA and sn-2-monooleoylglycerol, more than 96% of the products were di- and triacylglycerols. 90% of the diacylglycerol product was the 1, 2 isomer. The activity was stable at 43° for 50 min and had t 1/2 values of 8 min and 4.5 min at 53.5 and 55°C, respectively. Suckling rat MGAT activities in liver and intestinal mucosa were 100- to 300-fold higher than in other tissues. MGAT activity was 12.5 fold greater with C16:0-CoA than with C8:0-CoA. Acetyl-CoA was not a substrate. The sn-2-monoacylglycerols were strongly preferred over the sn-1 isomers. No direct relationship was noted between monoacylglycerol chain length and apparent K_m value. The presence of high MGAT activity suggests that the monoacylglycerol pathway provides a major route of glycerolipid synthesis in suckling rat liver.

1178 HEPATIC RESPONSIVENESS TO A GLUCOSE INFUSION IN PREGNANCY. Richard M. Cowett, Brown University, Women & Infants Hosp. Dept. of Ped. Providence, RI

To enhance glucose transfer to the fetus, the pregnant woman may evidence prolonged hyperglycemia after feeding. To evaluate if hepatic responsiveness to glucose and insulin, in contrast to decreased peripheral glucose uptake, contributes to this metabolic response, the glucose production rate (GPR) was measured in 15 pregnant non-diabetics (P-ND) (G: 37.3±0.3 wks), in 16 pregnant controlled (HbA_{Ic} 7.2±0.5) insulin dependent diabetics (P-ID) (G: 37.4±0.4 wks), and in 12 non-pregnant non-diabetic controls (C). Nineteen women were infused with 3mg·kg^{-1}·min^{-1} glucose 60 min prior to the kinetic analysis in which the GPR was measured with D[U-^{13}C]tracer glucose.

(M±SEM)	Saline Turnover				Glucose Turnover			
	N	Gluc. mg/dl	Ins. µU/ml	GPR mg·kg^{-1}·min^{-1}	N	Gluc. mg/dl	Ins. µU/ml	GPR mg·kg^{-1}·min^{-1}
P-ND	9	78±1	18±1	1.7(0.6-3.3)	6	115±6**	60±11**	0.2(0-0.6)**
P-ND	9	74±7	46±7	1.5(0.7-2.1)	7	129±16*	65±12	0.5(0-1.2)*
C	6	87±1	14±1	2.0(1.5-2.4)	6	114±7*	28±3**	0.1(0-0.3)**

unpaired t test (saline vs. glucose) *p<.01 **p<.001 (range)

All glucose groups had elevated pl. glucose compared to the saline groups. Both the P-ND and C had elevated pl insulin in response to glucose but the P-ND rise was greater(p<.05). The GPR was suppressed in all glucose groups; the GPR of the P-ID was similar to that of the P-ND but more variable than that of the C (p<.05). In the third trimester the P-ND and P-ID have similar hepatic responsiveness to glucose and insulin compared to C which does not contribute to the observed hyperglycemia.

1179 UNCOOKED CORNSTARCH (UCS): A DIETARY SOURCE OF CONTINUOUS GLUCOSE FOR PATIENTS WITH TYPE I GLYCOGENOSIS (GSD-1): II.OVERNIGHT STUDIES. Joseph I.Wolfsdorf, Lori B. Laffel, Ruth A. Flowers, John F. Crigler, Jr., Dept. of Ped., Harvard Med. Sch., Dept. of Med., Children's Hospital, Boston

We have examined the ability of UCS to provide a continuous source of glucose during the night in 10 children (ages 4-17yrs) with GSD-1. The amount of orally administered UCS was calculated to be equal to the rate of hepatic glucose production and given in either 4 or 5hr loads. All patients had been on a regimen of continuous overnight dextrose (COD) with D25 or D50 via gastrostomy and 1-3hr daytime D feedings prior to study.

Blood glucose (BG), lactate (L), free fatty acids (FFA), insulin (IRI), growth hormone, cortisol and glucagon levels obtained from 2200-0700hrs during nocturnal UCS and COD were compared. The results are shown in the following table (mean± SEM; n=number of determinations per 10 patients):

	BLOOD GLUCOSE mg/dl(n)	SERUM INSULIN µU/ml(n)	BLOOD LACTATE mM/L(n)	SERUM FFA µEq/L(n)
COD	89±3(140)	18±2(140)	3.2±0.5(50)	784±111(44)
UCS	79±3(164)	12±2(132)	3.6±0.6(50)	1142±216(55)
p value	0.001	0.014	0.55	0.035

Mean BG and IRI levels were lower with UCS than COD but both were in the normal postabsorptive range. Blood L's were not different. Lower serum FFA levels with COD were associated with persistently higher BG and IRI levels.

UCS successfully has provided continuous glucose without complication in 15 patients with GSD-1 for 3-16 months.

1180 EFFECT OF HYPOXIA ON MITOCHONDRIAL ELECTRON TRANSPORT COMPLEXES AND ADENINE NUCLEOTIDES IN THE NEWBORN HEART. Helen H. Young, Suguru Matsuoka, Shigeru Uemura and Jay M. Jarmakani, UCLA Medical Center, Dept. Peds., Los Angeles, CA.

Effect of 60 min hypoxia (N_2) and subsequent 40 min reoxygenation (re-O_2) on mitochondrial state 3 respiration (QO_2), electron transport complexes (ETC), adenine nucleotides (AdN= ATP+ADP+AMP) and adenine nucleotide translocase (AdNT) activity were studied in isolated, arterially perfused newborn (NB) and adult (A) rabbit hearts. In the oxygenated muscle (control) QO_2, AdNT and ATP were significantly ($P<0.05$) greater in NB than in A. After N_2, QO_2 and AdNT decreased significantly ($P<0.01$) in both age groups, and the values of QO_2, AdN, and AdNT in the NB were significantly greater ($P<0.05$) than in A. N_2 depressed ETC I in the A but not in the NB. After re-O_2, QO_2 and AdN returned to control values in NB (96 ± 10% of control) but not in A (56 ± 3%), and the recovery of AdNT in NB (70 ± 2%) was significantly greater than in A (39 ± 5%). There was good correlation between QO_2 and AdN as well as AdNT in both age groups. Conclusion: 1) the effect of N_2 and re-O_2 on QO_2, AdN, AdNT and ETC in NB is less than in A, 2) AdN is important in maintaining mitochondrial function.

1181 CONTINUOUS INTRAGASTRIC FEEDING(CIF) IN FAMILIAL HOMOZYGOUS HYPERCHOLESTEROLEMIA(FHH). D.Daneman,E. Regelink & V.Rose,(Sponsored by R.Ehrlich),Hosp. for Sick Children,Univ. of Toronto,Toronto,Canada.

We report the effect of CIF on lipid profiles in 2 children with FHH refractory to medical therapy: A: 5yr boy with receptor negative FHH; B: 16yr girl with receptor-deficiency(15%). The protocol consisted of CIF administered as Vivonex(V) to supply full caloric & fluid needs via a soft silastic nasogastric tube. Total cholesterol(TC),LDL-C,HDL-C & triglycerides(TG) were measured 3times prior to the start of CIF, & 12,36,72hrs, 1 & 2wk thereafter. In A, CIF was discontinued for 4hr/day after 1wk, & completely at 2wk. In B, CIF was maintained for 2 wk, then changed to 12hr nocturnal feeding(NF) to provide 1/3 daily needs & continued for 5 wk, supplemented with Colestipol for the final 3wk. Results: CIF caused a decrease in TC (772-692mg/dL(10%) at 1wk in A; 567-427(24%) at 3 wk in B); LDL-C [709-623(12%) in A; 516-400(23%) in B]; HDL-C [31-15(51%) in A; 144(71%) in B]. TG concentrations varied greatly. In A, discontinuation of CIF for 4hr/day led to return of all levels to baseline. In B, NF maintained levels of TC, LDL-C & HDL-C at 85,84&77% of baseline. Patient A refused further CIF. Conclusions: CIF leads to a fall in all cholesterol components in FHH. The response varied greatly (and may be dependent on receptor status). These results are similar, though quantitatively less than those previously reported. The mechanism of cholesterol lowering with CIF is unknown. However, a trial of this therapeutic approach may be warranted before more invasive measures (portacaval shunt/plasmaphoresis) are attempted.