●1255 LIPID INFUSION ENHANCES GLUCONEOGENESIS FROM AMINO ACIDS IN PREMATURE INFANTS. Renate D. Savich, Sandra Finley, Mary E. Bussey, Edward S. Ogata, Northwestern Univ. Med. School, Depts of Peds, OB-Gyn, Chicago.

Glucose production in premature neonates depends upon available gluconeogenic substrates and intact enzymatic and hormonal systems. To determine whether lipid alone or in concert with amino acids alters glucoregulation, we determined the glycemic response from intravenous lipid in 8 premature infants (BW 1226 ±100 g, GA 28.6±1.4 wks). At 4±2 days of age, we administered either Intralipid (L) alone (.25 g/kg over 20 min followed by .5 gm/kg/hr x 100 min) or an identical Intralipid dose plus amino acids (Aminosyn, .5 gm/kg over 4 hrs) (LA). All infants received concurrent glucose at 5-6 mg/kg/min throughout the study. Free fatty acid concentrations increased within 10 min of infusion and peak values were equivalent in L and LA infants (4345 vs 3100 μEq/L). LA infants demonstrated a brisk glycemic response compared to L infants (23±3 vs 7±2% increase at 10 min). In addition, LA infusion resulted in a far greater increase at 150 min compared to L (65±11 vs 30±7%). Preliminary data in infants receiving A alone suggest a limited glycemic response relative to L and LA infants. Insulin responded appropriately in L and LA infants increasing from 10 to 75 μU/ml in response to increasing glucose, while glucagon demonstrated no consistent pattern in either group. L infusion results in a mild glycemic response in premature infants, while LA administration more promptly increases glucose. These data suggest that lipids enhance gluconeogenesis from amino acids in premature infants.

1256 ALTERED ERYTHROCYTE MALONYLDIALDEHYDE (RBC-MDA) FORMATION IN REYE'S SYNDROME. K.B. Schwarz, J.P. Keating, P. Kanjananggulpan, and C.D. Fitch (Spon. by T. Aceto, Jr.) St. Louis University School of Medicine, Cardinal Glennon Children's Hospital, Departments of Pediatrics and Internal Medicine, and Washington University School of Medicine, St. Louis Children's Hospital, Department of Pediatrics St. Louis, MO.

Peroxide-induced erythrocyte malonyldialdehyde formation (RBC-MDA) has been used as a measure of polyunsaturated fat autooxidation, a process believed to alter membrane transport and enzyme function. Evidence for this process was sought in erythrocytes of RS patients using the method of Bidder (Life Sciences 30:1021, 1980). For 7 children with stage II-IV RS, RBC-MDA was 432 ± 36 nmoles/g hemoglobin on admission, 323± 25 at the nadir, and 478 ±67 1-4 months after RS. Nadir values differed significantly from admission and from values for 7 siblings (431 ± 28), 5 pediatric intensive care unit (ICU) patients (517 ± 32) and 5 adults (530± 40). Corresponding values for plasma vitamin E in RS patients were 556±37 mcg/dl at admission, 473±36 at the nadir, and 905±30 after RS. Nadir values were significantly different from recovery, siblings 1001±43, and ICU patients 740±98. Conclusion: The observed decrease in RBC-MDA in RS could be explained by decreased substrate (if RBC polyunsaturated fatty acids had undergone peroxidative degeneration) or, less likely, given the low plasma vitamin E, increased antioxidant activity.

†1257 OPTIMAL RATE OF NASOGASTRIC (NG) CARBOHYDRATE IN PATIENTS WITH TYPE I GLYCOGEN STORAGE DISEASE (GSD I) W. F. Schwenk and M.W. Haymond, Mayo Clinic and Foundation, Dept. of Pediatrics, Rochester, MN 55905.

Although noctural NG infusion (INF) of carbohydrate (CHO) is standard therapy in GSD I, the optimal rate of CHO has not been established. To determine if a CHO INF of ~6 mg/kg·min (glucose flux in overnight fasted normal children) suppresses hepatic glucose (GLU) production and optimizes plasma lactate, ketone body (KB) and free fatty acid (FFA) concentrations, 6 GSD I children (4.2 ± 0.8 yrs) received NG INFs containing Vivonex (3 mg of CHO/kg·min) with [$^2H_2$]Glucose plus varying GLU to achieve total INFs of 10.5, 8.6, 5.8 and 3.0 mg of CHO/kg·min. Each INF period was 3 hr with blood sampling during the last 30 min. GLU decreased ($p < 0.05$) from 106 ± 5 to 90 ± 5, 75 ± 5 and 52 ± 3 mg/dl during the 4 INFs whereas lactate (4.0 ± 0.5, 4.9 ± 0.6, 6.4 ± 0.7 and 8.8 ± 1.2 mM, respectively) and FFA (0.7 ± 0.1, 0.9 ± 0.1, 1.1 ± 0.2 and 1.6 ± 0.1 mM, respectively) increased. Lactate, pyruvate, KB and FFA were increased ($p < 0.05$) at CHO INFs of ~6 mg/kg·min compared to the higher INFs. Total GLU flux decreased ($p < 0.05$) from 10.5 ± 0.3 to 9.1 ± 0.2, 7.2 ± 0.1, and 6.0 ± 0.4 mg/kg·min for the 4 INFs while estimates of endogenous GLU production rose ($p < .05$) from ~0 to 0.5 ± 0.2, 1.4 ± 0.1 and 2.9 ± 0.3 mg/kg·min, respectively. In summary, NG INF of CHO at ~6 mg/kg·min in GSD I fails to suppress endogenous glucose production and optimize organic acidemia. In conclusion, if organic acidemia has an adverse effect on growth in GSD I, NG INFs in excess of ~6 mg/kg·min may be optimal.

1258 EPIDEMIOLOGY OF RICKETS ADMITTED TO COMBINED UNIVERSITY AND MUNICIPAL HOSPITALS IN 30 MONTHS. Binita Shah, Irma Fiordilisi and Laurence Finberg, SUNY, Downstate Medical Center/Kings County Hospital, Brooklyn, New York.

In 30 months the diagnosis of rickets was made 36 times as follows: Nutritional (N)-8, Familial Hypophosphatemia (FHP)-7, Vitamin D Dependent 1 (DDI)-1, Vitamin D Dependent 11 (DDII)-2, Renal Osteodystrophy (RR)-16, and Hepatic Rickets (HR)-2. The 16 N, FHP and DDI patients were all from self referral families with relevant complaints. The 8 N patients all had dark skin; 4 of these were exclusively breast fed with low calcidiol levels in serum. One had dietary CA deficiency with increased calcitriol levels in serum. Three siblings had multiple dietary deficiencies because of diet faddism. 5/8 studied did not have amioaciduria or increased PTH. The 8 genetic metabolic bone disease patients had varied skin color. The remaining 20 patients had rickets as a complication of severe renal disease or liver disease or were specifically referred because of DDII. We can distinguish N from FHP by an objective therapeutic test (600,000 u Vitamin D in 24 hours) within five days when other criteria are inconclusive. Serum PO_4 rises in 4-5 days and x-ray changes are visible at one week in patients with N. We conclude: 1) rickets in the inner city U.S. population is relatively common; 2) the primary varieties (N and FHP) are readily distinguishable by simple clinical means; 3) secondary aminoaciduria and hyperparathyroidism are frequently absent in nutritional rickets in this series.

1259 GLYCOHEMOGLOBIN BY AFFINITY CHROMATOGRAPHY: A THIRTY MINUTE PROCEDURE IN CLINIC. Shirish C. Shah and John I. Malone. University of South Florida, Department of Pediatrics, Tampa, Florida.

The utility of glycohemoglobin (GHb) in the management of diabetes mellitus (DM) is undisputed. Clinical decisions frequently are deferred 3-5 days until the GHb results are available; hence, follow-up instructions must be given by telephone or letter. The delay and lack of personal attention may markedly reduce compliance; conversely, postponing new recommendations until the next evaluation may reduce the utility of GHb to a mere academic exercise. We evaluated affinity chromatography (AC) for the possibility of obtaining GHb results while the patients were in the clinic. From 23 children with insulin dependent DM, 25 μl of blood was collected by a fingerstick and GHb was measured by AC. Results for up to 5 specimens were available in 30 minutes. A venous blood specimen was collected simultaneously from the same 23 children and HbA_{1c} was measured by high pressure liquid chromatography after removal of labile fraction (LF). A remarkable correlation was found between the two methods ($r = 0.89$). The study was repeated 2 months later and similar results were obtained ($r = 0.91$, $n = 25$). Even though LF was not removed in AC, the correlation between the two methods indicates that LF may only be an insignificant portion of total GHb. The AC was less expensive than the conventional methods and was more acceptable to the children as venipuncture was not required. We conclude that the utility of GHb may be improved in clinical practice utilizing AC, as prompt results can be obtained at less expense without the necessity of a venipuncture.

1260 ALANINE-INDUCED HYPERAMMONEMIA IN HYPERLYSINEMIA AND IN SACCHAROPINURIA. Olli Simell, Päivi Luukkainen and Ilkka Sipilä (Spon. by David Rosenblatt). University of Helsinki, Helsinki, Finland.

It has been suggested that lysine inhibits urea cycle function both in vivo and in vitro mainly by decreasing arginase activity. We gave an L-alanine load to a patient with hyperlysinemia (fasting plasma lysine 860 μM on a 1.5 g protein/kg diet) at age 5 and 10 years, and to a patient with saccharopinuria (and hyperlysinemia) at age 3, 12.5 and 16 years. The alanine was given as a 5% aqueous i.v. infusion (6.6 mmol/kg) over 90 min and changes in blood ammonia, serum urea urinary orotic acid were measured.

In healthy subjects, serum urea increases rapidly, but blood ammonia and urinary orotic acid remain unchanged after such a load. The patient with hyperlysinemia developed hyperammonemia (peak values 760 and 210 μM at 5 and 10 years of age), and orotic aciduria increased 10-fold in the first 6-h collection. Serum urea increased slowly. The saccharopinuric patient's peak ammonias were 60, 280 and 410 μM at 3, 12.5 and 16 years of age, respectively; serum urea increased always slowly. Orotic acid excretion in the latest test increased 320-fold. Addition of 1.1 mmol/kg of l-ornithine to the alanine loads abolished the hyperammonemic response totally in both, and urinary orotic acid increased only 2-3 fold. The findings suggest that patients with constant hyperlysinemia or with saccharopinuria have decreased nitrogen tolerance and may benefit from ornithine supplementation.