1177 CHRONIC NEUTROPENIA IN GLYCOGEN STORAGE DISEASE (GSD) TYPE Ib. Thomas F. Roe, spon. by George N. Donnell. Univ. of Southern California, School of Med., Childrens Hospital of Los Angeles, Dept. of Pediatrics, Los Angeles.

Two unrelated boys with typical clinical and biochemical features of GSD Type I had increased glycogen in liver but normal glucose-6-phosphatase activity (assayed after freezing). Both had chronic neutropenia from infancy, gingivitis and lesions of tongue, oral and anal mucosa. Their mean ± SEM blood neutrophil counts were 288 ± 94 and 494 ± 113 cells/mm^3. Aside from splenomegaly, there was no evidence of hypersplenism and investigation of other causes of neutropenia was negative. Their bone marrows showed mild granulocytic hyperplasia with no maturation arrest. In one patient tested, the blood neutrophil count rose after subcutaneous glucagon and intravenous hydrocortisone administration. Studies of immunologic competence gave normal results. Treatment with nocturnal glucose alimentation increased the childrens' growth rates but had no effect on the neutropenia. Conclusions: 1) Chronic neutropenia is associated with GSD Type Ib. 2) The neutropenia is probably not due to an arrest in granulocyte maturation. 3) The hepatic deficiency in Type Ib (Glu-6-P translocase) may also involve the neutrophil.

1180 SCREENING TERM LGA NEONATES FOR HYPOGLYCEMIA. Malini Satish, Gerald H. Katzman, J. Williams, Jose Urrutia, P.L.S. Amma, Irwin Weinfeld, Sidney Kripke, Venkatesan Krishnan. (Spon. by M. G. Robinson) Medical College of Ohio, The Toledo Hospital, Dept. of Ped., Toledo, Ohio.

Term large for gestational age neonates are at increased risk for hypoglycemia. It was postulated that identification of term LGA newborns by the Portland Intrauterine Growth Curve as opposed to the Colorado Intrauterine Growth Curve would allow for the classification of a lesser number of neonates as LGA but would identify the LGA neonates at risk for hypoglycemia. We reviewed retrospectively the charts of 100 inborn neonates born from August through October 1980 who plotted as LGA (wt.> 90th %tile) on the Colorado Chart. 61 of these 100 infants were LGA on the Portland Chart. All 100 neonates were screened for dextrostix values less than 45 mg.% in the first two hours of life. Of the 17 neonates found to have dextrostix <45 mg.%, 15 had a stat quantitative B.S. performed. All 15 neonates were symptomatic with tremors and jitteriness. 11 out of these 15 neonates had a B.S. <40 mg.%, requiring immediate treatment. All 11 hypoglycemic neonates were identified as LGA on the Colorado Graph whereas only 7 of these 11 neonates were identified by the Portland Graph as LGA. We conclude that in our population the Colorado Graph seems to be a better tool than the Portland Graph in identifying term LGA neonates at risk for hypoglycemia.

●1178 THE METABOLISM OF LEAD-210 (^{210}Pb) IN ISOLATED BONE CELL POPULATIONS. John F. Rosen, Albert Einstein Coll. Med., Montefiore Hosp. & Med. Ctr., Dept. Ped., New York.

Bone is the major storehouse of the body burden of Pb in man. Nonetheless, the biological significance of bone Pb is largely unknown. Studies in bone organ culture have shown that one subcompartment of bone Pb is readily exchangeable and the source of Pb chelated by $CaNa_2EDTA$. This subcompartment of bone Pb in vitro is regulated by the same hormones and ions that normally control bone cell metabolism. To characterize this subcompartment at the cellular level, bone cell populations from mouse calvaria were enriched for osteoclasts (OC) and osteoblasts (OB) by collagenase digestion. Once cells grew to confluency, OC and OB cells were subcultured in multiwells to which fresh medium and ^{210}Pb were added. After incubation with ^{210}Pb for 48 H, labelled cells were chased with ^{210}Pb-free medium. ^{210}Pb uptake was present by 1 H; a plateau was reached by 10H: OC: 4.51 x 10^3 cpm/mg cell protein; OB: 1.12 x 10^3 cpm/mg cell protein. Uptake increased linearly as medium ^{210}Pb was increased; by 48 H, OC accumulated 4-5 times more cpm compared to OB. In ^{210}Pb-free medium, OC and OB cells released 29.2 ± 1.1 and 5.6 ± .9%, respectively, of preincorporated label by 10 H. Vigorous washing of cells with $CaNa_2EDTA$, trypsin and TCA indicated that virtually all ^{210}Pb incorporation was intracellular. These data suggest that: 1) OC and OB cells incorporate ^{210}Pb rapidly; a significant fraction of incorporated label is exchangeable; 2) In this system, OC appear to be the major cell type involved in the metabolism of Pb in bone.

●1181 PLASMA KETONE BODIES DURING A 24-HOUR FAST IN NORMAL AND HYPOGLYCEMIC CHILDREN. RELATIONSHIP WITH AGE. Jean M. Saudubray, Cecile Marsac, Francois X. Coude, Jean M. Limal, and Christiane Charpentier (Spon. by Jerry A. Schneider), Department of Pediatrics, Hopital des Enfants-Malades, Paris.

The variations in blood ketone bodies (BKB), blood glucose (BG) and insulin were studied in 19 normal and 14 hypoglycemic children aged 4 months to 13 years during a 24 hour fast. Except in 4 patients (2 with hyperinsulinism, 2 with defect in ketogenesis), a significant and parallel increase in BKB was observed in controls and patients from 0h to 20h (in mmol/l, mean±SD) 0.20±0.16 at 0h, 1.54±1.26 at 15h, 3.05±1.45 at 20h, 3.87±1.48 at 24h. A significant progressive decrease in glucose levels was observed from 0h to 20h but not from 20h on (in mg/dl) 116±16.4 at 0h, 74.5±16 at 15h, 60±15.5 at 20h, 59.57±13 at 24h. A highly negative correlation between BKB and BG was found at each time of fasting ($r \geq -0.54$, $p < .001$ with a large dispersion of BKB especially for the ones corresponding to the BG between 45 and 65 mg/dl (SD=1.75mM). This dispersion was consistently reduced in a homogenous age group 4-6 years presenting with similar glucose levels (SD=0.92mM). There was a positive correlation between age and BG from 21h on ($r \geq 0.61$, $p \leq 0.01$), and an inverse relationship between age and BKB from 15h on ($r > -0.43$, $p < 0.001$). The same high inverse relationship between age and BKB was again observed when the variable of glucose level was factored out ($r = -0.73$, $p < 0.001$) demonstrating that the variation in BKB is indeed related to age. These findings deserve attention for the interpretation of fasting BKB especially when used as an aid in the diagnosis of the various forms of childhood hypoglycemia, and hypoketotic states.

●1179 MODULATION OF SOMATOMEDIN RECEPTOR SITES IN GROWTH HORMONE-TREATED HYPOPITUITARY DWARFS. Ron G. Rosenfeld, Stephen F. Kemp and Raymond L. Hintz, Stanford University School of Medicine, Dept. of Pediatrics, Stanford, California.

We have previously demonstrated that somatomedin (SM) can induce the loss of specific SM-C receptors on cultured IM-9 lymphocytes in a time- and concentration-dependent manner. To investigate the acute regulation of SM binding under in vivo conditions, we have evaluated SM receptors on circulating mononuclear cells obtained from 12 hypopituitary dwarfs prior to and after 4 days administration of hGH, 0.1 U/kg/day. Plasma SM levels, measured by radioreceptorassay, rose from 0.37 ± 0.08 U/ml (mean ± SEM) to 1.00 ± 0.10 ($p < 0.001$). Concomitantly, specific binding of 125-I-SM-C to 50 x10^6 mononuclear cells/ml fell from 13.61 ± 0.97% to 10.40 ± 0.85% ($p < 0.02$). Scatchard analysis demonstrated that the decrease in specific binding was predominantly secondary to a reduced number of SM-C receptor sites per cell, with no alteration in receptor affinity. Overall, a significant inverse correlation was observed between plasma SM levels and mononuclear cell binding of 125-I-SM-C ($r = -0.62$, $p < 0.001$).

The data demonstrate that treatment of hypopituitarism with conventional therapeutic doses of hGH results in significant acute increases in plasma SM levels in the majority of subjects, with a reciprocal decline in the specific binding of 125-I-SM-C. We conclude that SM, like insulin, is capable of regulating homologous receptor concentrations under both in vitro and in vivo conditions.

1182 EXPERIMENTAL MODELS OF MICROVESICULAR FATTY CHANGE IN RAT LIVER. Marian Statter, Ephraim Sagi, Alex Russell, Nelly Livni & Richard Deckelbaum (Spon. by Ingeborg Krieger), Wayne State University, Dept. of Pediatrics, Detroit & Hadassah University Hospital, Depts. of Pediatrics, Pathology and Gastroenterology, Jerusalem.

An association of microvesicular hepatic fatty change with hyperammonemia and orotic aciduria (OAuria) has been noted in several pediatric conditions, including Reye's syndrome. Experimental OA supplementation produces fatty changes in rat liver. In the present study severe hyperammonemia (2-3 mg%), without OAuria was produced by repeated administration of urease (25 U/kg) for periods up to 24 hrs. The hyperammonemic rats were found to have hyperglycemia (374±21 mg%: controls: 172±6 mg%), elevated plasma FFA (815±62 μEq/l: controls 491±25 μEq/l) and hepatic triglycerides (15.6±1.4 mg/gm: controls: 2.7±0.2 mg/gm) (SEM). Liver sections stained with oil red O showed microvesicular fatty changes. To evaluate the role of OA in the development of fatty changes, conversion of OA to uridine monophosphate (UMP) was blocked with azauridine. Under these conditions fatty changes resulting from urease-induced hyperammonemia were markedly less than those observed without azauridine. It is concluded that increased availability of carbamyl phosphate in hyperammonemia leads to overproduction of OA and that the conversion of increased amounts of OA to UMP rather than OA elevation was an essential prerequisite to the induction of hepatic microvesicular fatty changes.