865 LIPID CHANGES ASSOCIATED WITH PSYCHOSOCIAL DWARFISM (PSD). Nancy J. Hopwood, Dorothy J. Becker, Faye H. Hengstenberg and Allan L. Drash, Dept. of Ped., Univ. of Michigan and Pittsburgh, Ann Arbor and Pittsburgh

Turbidity of fasting serum and a distinctive pattern of changing blood lipids was observed during the first week of hospitalization in 14 children aged 1.9-14.3 yrs with PSD. All demonstrated remarkable catch-up growth with changed environment. On day 1 (morning of admission) mean ($\bar{x}$) fasting triglyceride (TG) was 69% and cholesterol (C) 156 mg%. Fasting sera in 11/12 were turbid without clearing after 48 hrs. During the first 4 days, serial observations in 10/11 children showed dramatic rise in TG (36-594%) with decrease to normal by day 6-8. Serum C gradually rose in 6/14 by day 6-9 (29-52%). During this time serum turbidity cleared. Paper lipoprotein electrophoresis of 10 hr fasting serum was normal except for a light chyle spectrum in 7/13 and an increased beta band in 1/13. An increase in prebeta fraction occurred on day 2-4 in 10/12. Recurrence of turbidity occurred after 15/21 parent visits, clearing again with parent-child separation. By contrast, 5 children with growth failure and protein-calorie malnutrition, 1.2-3.9 yrs, 24 children with hypopituitarism, 1.2-17.1 yrs ($\bar{x}$ TG 66 mg%; $\bar{x}$ C 182 mg%) and 16 children with other endocrine disorders undergoing similar hospital evaluations ($\bar{x}$ C 163 mg%) had clear fasting sera on day 1. No change in serum appearance, TG or C was seen in 8/8 of these followed serially during days 1-4. The presence of serum turbidity may provide a clue to diagnosis in psychologically induced growth retardation. The mechanism of this phenomenon is as yet unexplained.

866 FANCONI TYPE GLYCOGENOSIS: METABOLIC AND RENAL FUNCTION STUDIES. Mark Houser and Carol R. Angle, Univ. of Neb. Coll. of Med., Dept. of Ped., Omaha, Nebraska.

Fanconi Type Glycogenosis (FTG) is a unique syndrome characterized metabolically, in 9 cases, by the early onset of phosphaturia, glycosuria, amino aciduria and uricosuria and clinically by hypophosphatemic rickets, growth failure and hepatomegaly. Classic hepatic enzyme deficiencies are not found in these patients although there are metabolic similarities to Glycogen Storage Disease. Fasting hypoglycemia and lactic acidemia present less of a problem even though there is mild glucose (G),extreme galactose (Gal) intolerance. Renal function studies in a 26 month old white male with FTG demonstrated quantitatively similar increases in phosphate clearances (C_P) relative to inulin (C_{IN}) on G, Gal and xylose infusion while C_G and $C_{Uric\ acid}$ were unchanged.

Extreme tubular dysfunction allows use of the urine G:Nitrogen (N) ratio to define defective gluconeogenesis from protein, as demonstrated by reduction of the G:N ratio from 6.3 to 3.0 on high protein infusion. A good clinical response has been obtained by the introduction of high carbohydrate feedings by wearable infusion pump.

867 GLUCOSE PRODUCTION IN POSTABSORPTIVE WOMEN AT TERM GESTATION: LACK OF FETAL GLUCOSE RELEASE. Satish Kalhan, Larry D'Angelo, John Lovecchio, Samuel Savin and Peter Adam. Case Western Reserve Univ.Div. of Pediatr. Metab. at Cleve. Metro. Gen. Hosp., and Dept. of Earth Sciences, Cleveland, Ohio.

The effect of early diabetes on systemic glucose production rate(SGP)was evaluated in 4 normal (NM) and 3 gestationally diabetic mothers(GDM)undergoing cesarian section. Glucose-1-^{13}C was used as tracer according to prime-constant-rate infusion technique. Normoglycemia was maintained in GDM by dietary regulation. After a 12h fast SGPs in NM and GDM were similar while ^{13}C-enrichment ($\delta^{13}C‰$) of cord arterial glucose (CA)was lower than that of maternal venous glucose (MV) only in GDM (Mean ± SD).

	SGP	$\delta^{13}C‰$		
	mg/kg.min.	MV	CA	CV
Normals	2.4±0.6	75.6±14.4	76.0±13.3	74.4±17.4
GDMs	1.9±0.6	97.1±29.1	80.5±31.8	91.3±32.8

In addition,the natural ^{13}C abundance of cord blood glucose resembled the mother's in 5 NM and 5 insulin-dependent diabetic mothers. Conclusions: Brief maternal fasting normally does not initiate glucose production in the human fetus. GDMs apparently pass through a phase of reduced glucose assimilation while SGP during fasting remains normal.

868 ABNORMAL REGULATION OF CALORIC HOMEOSTASIS IN REYE'S SYNDROME SURVIVORS. Ellen S. Kang, Kathryn Schwenzer, Solomon S. Solomon, William Duckworth, John Smith and Gerald Billmeier. (Spon. by Walter Hughes). St. Jude Children's Res. Hosp., Lab. of Biochem., V.A. Hosp., Dept. of Med., Endocrine Sect. and the Univ. of Tenn. Centr. for the Health Sciences, Depts. of Peds. and Med., Memphis, Tenn.

The possibility that survivors of Reye's-like syndromes may be metabolically unique was explored by examination of their ability to mobilize metabolic fuels under endogenous and exogenous stimuli. 4 children, aged 5½, 8 10/12, 12 and 14½ years at the time of the acute disease were studied 1/3-3 years later. Oral glucose tolerance tests after 3-days CHO loading were abnormal: 3 had higher glucose and insulin and all had higher glucagon levels than normals. Fasting insulin and glucagon levels without CHO loading were repeatedly high and molar ratios were lower than normal in all. Glucose levels after epinephrine, glucagon and insulin were variable. The youngest child who received the max. recommended dose of epinephrine developed acute hepatomegaly (5cm below the RCM over a 3-4 hour period). Usual liver function studies were normal and enlargement was verified by technicium scintiangiography. Fasting glucose, insulin or glucagon levels were high in 3 of 4 parents and 1 of 2 asymptomatic siblings. These results indicate that these survivors are metabolically abnormal in their regulation of glucose and glucose substrates. It is likely that this was present prior to the acute disease, and we postulate that it imparts a risk for such a syndrome. Supported by USPHS NIH Grants HD09469, CA05176,AM18022 & VAMRIS 8036 & 1942.

869 MONOCYTE CELL RECEPTORS FOR INSULIN: QUANTITATIVE ESTIMATION BY RADIOAUTOGRAPHY. Solomon A. Kaplan, Barbara M. Lippe and Naomi D. Neufeld. Department of Pediatrics, UCLA School of Medicine, Los Angeles.

Insulin resistance has been found to correlate well with binding to target tissues,and circulating monocyte receptors present an accurate mirror of receptors in other tissues such as liver, muscle and adipocytes. Studies on children are severely limited by the amount of blood necessary for each assay, between 80 and 150 ml, and virtually no information is available regarding insulin receptors in children. We have developed an autoradiographic method which requires only a few ml blood for a complete assay. Our method consists of incubation of leukocytes with insulin labeled to high specific activity with I-125. An aliquot of the reaction mixture is transferred to microscope slides which are coated with a photosensitive emulsion. After staining, the silver grains associated with 15 to 25 monocytes are counted. A good parallelism was found between the autoradiographic and standard methods. Binding in the presence of 5 nanograms I-125 insulin, S.A. 100-150 uCi/ug, averaged 4.32% ± SE 0.55 in 7 experiments. Following addition of 1,10 and 100 ng insulin per ml the values were 3.3 ± 0.68, 1.96 ± 0.42 and 1.06 ± 0.36. Grain counts per monocyte at these four concentrations of insulin were 3.48 ± 0.48, 1.77 ± 0.37 and 1.46 ± 0.26. The method appears to be a valid method for estimation of binding of insulin to circulating monocytes. It should now be possible to determine insulin binding in normal and abnormal states in infancy and childhood.

870 PROTEIN SYNTHESIS AND CATABOLISM IN HEALTHY CHILDREN, BEFORE AND AFTER ELECTIVE SURGERY. Craig L. Kien, Dennis K. Rohrbaugh, John F. Burke, and Vernon R. Young, (Spon. by J. Richard Hamilton) Harvard Med. Sch., Shriners Burns Inst., Depts. of Ped. and Surgery, Boston, and MIT, Dept. of Nutrition and Food Science, Cambridge, Mass.

We measured whole body rates of protein synthesis(S) and catabolism(C) in seven healthy children, age 4 to 15 yrs., before reconstructive surgery and in six of them, 5 days postoperatively using a constant ^{15}N infusion technique. Pre-op, S and C fell between reported infant and adult normal values (g prot/kg/d) but did not vary with age of our subjects. S decreased 15% ($P<.05$) post-op although body weight, intake of protein and calories, N balance, and C did not change. S did correlate with protein ($r = +.75$) and calorie ($r = +.58$) intake. Pre-op, mean S (g prot/d) expressed per unit BMR (0.11) or per g protein allowance (5.0) was the same as that previously estimated for infants or adults. Our studies show: 1. Minor surgery causes decreased protein synthetic rate in face of unchanged protein and calorie intake, N balance, and catabolic rate. 2. Energy expenditure relates closely to protein synthetic rates at all ages. 3. The efficiency of N utilization in healthy children is similar to that in infants and adults.

	Pre-Op	Post-Op
S (g prot/kg/d)	3.9±0.8	3.3±1.1
C (g prot/kg/d)	3.4±0.6	3.3±1.1