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EFFECT OF RAPID AND PROLONGED WEIGHT REDUCTION ON VITAMIN STATUS IN OBESE ADOLESCENTS - 6 MONTH FOLLOW UP. Zwiauer K, Widhalm K, Brubacher G, Dept. Ped. Univ. Vienna, Austria; Dept. Nutr. Vit. Res., Hoffmann La Roche, Basel, Switzerland.

Much attention has been paid to the effects of weight reduction on protein and carbohydrate metabolism. However, only few reports exist in regards to vitamin status during weight reduction in obese adolescents. Therefore we investigated serum Vitamin A, E, and C (HPLC), retinol binding protein (RBP, RID), prealbumin (PA, RID), and LDL-C (LRC-methods) in 16 obese adolescent boys (45±11% overweight, 12.1±1.2 years, mean±SD) during a 3 week diet camp and after 6 months follow up. Weight loss after 3 weeks: 5.3±1.3 kg, weight gain after 6 months: 2.3±1.9 kg (1.3±11.9% overweight).

Results:	RBP	PA	LDL-C	Vit. A	Vit. E
baseline	5.3±0.2	28.1±1.2	132±18	505±21	9.8±0.4
3 weeks	3.0±0.1*	16.8±0.1*	82±18*	252±14*	5.9±0.4*
6 months	5.4±0.3*	30.8±2.1*	112±20*	432±21*	9.4±0.4*
	* p<0.001	+ p<0.01			

Vitamin A and E blood levels markedly decrease after 3 weeks but reached baseline levels 6 months later. Concomitantly RBP and PA showed similar changes, Vit. E an ongoing drop and rise. Vitamin C levels remained unaffected. The low vitamin A and E concentrations during the 3 weeks treatment (11 of 21 boys could be classified as "high risk for deficiency") can be explained by the marked fall of RBP and PA, due to the catabolic situation, in spite of adequate protein intake (0.9±0.2g/kg ideal body weight). We therefore suggest that rather the low concentrations in transport proteins than impaired vitamin intake caused these effects.

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GLYCOGEN STORAGE DISEASE TYPE I AND III (GSDI, III) AND PYRUVATE CARBOXYLASE DEFICIENCY (PCD): NOCTURNAL GASTRIC DRIP FEEDING VERSUS ADMINISTRATION OF UNCOOKED CORN STARCH! K. Ullrich, A. van Teeflen-Heithoff, Department of Pediatrics, University of Münster, FRG.

In three patients with GSD I, three with GSD III and one with PCD (age 4-20 y) nocturnal drip feeding (duration 2-8 y) was replaced by administration of uncooked corn starch in curd. To reach comparable serum glucose levels over night and in the morning (70-100 mg/dl) as during drip feeding, the starch had to be given 1-2 x per night. Patients aged below 14 y received 0.5-2.0 g/kg at 9 p.m. and an additional 3.0-4.0 g of starch/kg at midnight, whereas the two oldest patients got a single dose of 3.0-4.0 g/kg at midnight only. Caloric intake over night was 100-120 % of that during drip feeding. In addition, three of the younger patients obtained 1.0-2.0 g starch/kg one hour after breakfast and lunch. Following a period of 1.5-2.5 y no significant change occurred in serum lactate, alanine, GOT, GPT, uric, acid, triglyceride, cholesterol values and in the growth rate when compared to the last two years during drip feeding. The high starch doses were tolerated without side effects.

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LONG TERM USE OF D-HYDROXYBUTYRATE (DOHB) IN THE MANAGEMENT OF A PATIENT WITH 3-HYDROXY-3-METHYLGLUTARYL CoA (HMGCoA) LYASE DEFICIENCY. B. François and C. Bachmann, Dr Willems Instituut, Diepenbeek Belgium and Dept of Clinical Chemistry Bern Switzerland

Patients with inherited HMGCoA lyase deficiency show severe nonketotic hypoglycemia, acidosis and typical organic aciduria. In addition they are unable to produce adequate amounts of ketone bodies since the HMGCoA cycle is blocked in this disease. We report the effect of long term administration of DOHB (4mmol/bw/d) for treatment of a boy with this disorder from age 2 to 6 years. DOHB was applied in 4 daily doses. Monthly biochemical control analyses included glucose, ketone bodies, amino- and organic acids. In addition hepatic, renal and ocular function was assessed. DOHB assays showed adequate compliance with therapy. 3 episodes of hypoglycemia without acidosis were found associated with intercurrent infections. At the age of 6 years, the psychomotor development is normal. Height and weight remain at the 3rd percentile and head circumference is at the 50th percentile. No adverse effects of the DOHB administration were observed so far. We conclude that the long term use of DOHB together with a protein restricted diet allows feasible and effective treatment of HMGCoA lyase deficiency. The D- isomer of potassium hydroxybutyrate was a kind gift of Mr d'Oultremont, Brussels.

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PRESENCE OF DIURNAL VARIATION FOR IONIZED AND TOTAL Ca EXCRETION IN PREMATURE INFANTS.

A Moore, I Radde, A Huang, Research Institute, The Hospital for Sick Children, Toronto, Canada.

Diurnal variation in urinary calcium is observed in adult subjects, and is ascribed primarily to food intake, exercise habits, a day/night routine, and lastly an intrinsic rhythm. We wished to see whether premature infants showed diurnal variation in urinary Ca, and hypothesized that this would be largely an intrinsic rhythm since the infants were exposed to 24 h light and regular feeds around the clock.

We studied 5 infants (3 male, 2 female) birth weight 1.17±0.20 kg (mean±SD), gestational age 28±0.7 wks at 34±5 days of life, who were being fed every 2-3 hours. Urine was collected each time the infant voided for a period of 24 hours. Each specimen was analyzed for ionized Ca (iCa), total Ca (tCa), and pH, and excretion rates for Ca were calculated. The mean three-hourly Ca excretion rate for each infant was expressed as a percentage above or below the mean 3-hourly rate for the 24 h from midnight to midnight, and the average patterns of the five subjects graphed. Urinary iCa and tCa showed a diurnal variation (p<0.05). The values between 04.00 and 09.00 h were 17% above the 24 h mean and between 16.00 and 24.00 h were 19% below the 24 h mean. This is very similar to the intrinsic rhythm for Ca excretion described in adults, which coincides with the diurnal rhythm of cortisol secretion by the adrenal cortex, and is possibly linked to a rhythm in bone resorptive mechanisms. Thus this intrinsic rhythm of Ca excretion is present even in premature infants, and should be remembered when timed urine collections are made.

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A THERMODYNAMIC MODEL TO ASSESS THE METABOLIC RATE IN NEWBORN INFANTS.

Simbruner G., Entacher U., Universitäts-Kinderklinik Vienna, Austria.

Direct and indirect calorimetry are cumbersome and expensive conventional methods to assess the metabolic rate in neonates. We developed a thermodynamic model (TM) as an alternative method, whereby heat production is derived from heat loss to the environment and from heat storage, and converted into an oxygen consumption equivalent (4.83cal=1ml O2). In such a model the neonate is viewed to consist of a central cylinder (heat+torso) and a peripheral cylinder (limbs) whose volumes and surfaces are derived from body weight, length and standards. Heat loss from the surface of each cylinder is computed from its area, a combined heat transfer coefficient and the difference between mean skin and operative environmental temperature. The mean skin temperature of the central cylinder is represented by the temperature at the chest (between R nipple and xyphoid) and of the peripheral cylinder by the temperature at the calf (between knee and heel). Operative environmental temperature is approximated from incubator air and room temperature. Heat storage is computed from the rate of change of mean body temperature, the specific heat of tissue and bodyweight. The mean body temperature is derived from weighted mean skin and core temperature. The two skin, the core (esophageal) and incubator air temperature are measured by thermistors. The data are then smoothed by a special mathematical procedure and processed by a computer based analytic-theoretical model to yield oxygen consumption. The validity of the model was evaluated at the University of Vienna, Austria.

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EVALUATION OF THE THERMOMODEL 2C4T AT THE UNIVERSITY VIENNA, AUSTRIA.

Simbruner G. Entacher U., Universitäts-Kinderklinik Vienna, Austria.

The thermomodel 2C4T to assess oxygen consumption and a flow through method (FM) were synchronously applied to 16 neonates in order to determine the validity of the thermomodel (TM). The newborn infants were healthy, had a gestational age (mean±SD) of 33.1±3.3 wks, a postnatal age of 20±18 days and a weight of 1.97±0.5 kg at the time of study. All newborns were nursed in an Isolette single wall incubator with an operative environmental temperature of 30.5±1.1°C and a relative humidity above 70%. They were studied being naked and lying supine for a period of 60 min before and 40 min after feeding. For the flow through method we used a gas flow of 1 to 2 l/min/kg bodyweight sucked through a plexiglas hood, measured by pneumotachography and calibrated by a spirometer; and an oxygen analyzer OM11 (Beckman) the input of which received dry gas at stable pressure and temperature. The oxygen consumption (VO2) was calculated as product of flow and oxygen concentration difference. The thermomodel was applied the standard way. The mean VO2 during the study period was 7.00ml/kg.min determined by the FM and 7.97ml/kg.min by the TM. The VO2<sub>TM</sub> correlated statistically significant with the VO2 (r=0.58, N=138, y=3.21+0.68x, p<0.0001). The difference between VO2<sub>TM</sub> and VO2<sub>FM</sub> in each pair of measurement averaged 0.97ml/kg.min with a standard deviation of 1.44 ml/kg.min. All original data will be presented to demonstrate the good intra-individual correlation between VO2<sub>TM</sub> and VO2<sub>FM</sub>. The thermomodel reliable reflects relative VO2 changes but requires improvement to measure absolute VO2 with greater accuracy.