648 ENERGY EXPENDITURE AND BODY COMPOSITION IN PRADER-WILLI SYNDROME.

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The assessment of energy expenditure (EE) in humans in a normal environment has recently become possible through measurement of the differential excretion of doubly labeled heavy water ($^2H_2{}^{18}O$) following oral ingestion. After $^2H_2{}^{18}O$ distribution, CO_2 production and EE may be calculated from differential ^2H and ^{18}O enrichment in urine samples at 1 and 7 days. Further, isotope dilution at 4 hours may be used to calculate total body water and fat-free mass (FFM).

We studied 10 individuals with Prader-Willi syndrome (PWS) (8-24 years) and 5 normal obese children (NO) (7-16 years). None had other chemical abnormalities, and both groups were similarly obese (PWS 157±9.7%, NO 152±11.7% ideal body weight). PWS were 47.9±2.3% fat comp. to NO 45.6±1.9% (N.S.). However, % fat linearly correlated with body mass index (kg/m^2) only in PWS ($r=0.93$). Two normal-weight PWS had 40% and 37% body fat. EE in PWS was 2103±203 kcal vs. 3478±407 kcal, NO ($p<.005$). EE/kg FFM, 71.6±3.4 PWS vs. 76.3±6.4, NO, or EE/kg body weight, 37.6±2.6 PWS vs. 42.0±4.5, NO, was not significantly different, but the measured EE/calculated basal metabolic rate was less in the PWS, 1.34±.08 vs. 1.65±.12, NO, ($p<.05$). PWS have increased body fat even when of normal weight. EE in PWS normalized for FFM or weight is not significantly lower than in NO. These studies clarify the nature of obesity in PWS using a non-invasive technique with potential application to other disorders of EE and intake.

† 651 USE OF ^{13}C-GLUCOSE BREATH TESTS IN THE CHARACTERIZATION OF ABSORPTION OF CARBOHYDRATE IN FORMULA BY INFANTS WITH DIARRHEA.

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The appearance time (AT) of the first detectable levels of $^{13}CO_2$ (> .01% dose/min) in breath following the oral administration of UL-^{13}C-glucose (^{13}C-G, 5 mg/kg) provides an estimate of the time required for orally administered ^{13}C-G to reach the primary site of absorption and utilization. The AT for ^{13}C-G will increase as the absorption of glucose occurs at more distal sites, until a maximum is reached when the glucose enters the colon where it is rapidly oxidized to CO_2. The AT of ^{13}C-G administered alone and with formula (60-240 ml) has been used to characterize glucose absorption and utilization in 6 infants (16-93 d) with diarrhea and malnutrition (arm/FOC < 0.28). A maximum value of 27 min for the AT of ^{13}C-G from formula was determined in an infant with congenital glucose-galactose malabsorption, in whom colonic utilization of CHO was demonstrated by breath H_2 production and partial recovery of ^{13}C in stool. AT for ^{13}C-G in formula, obtained for the 6 infants measured, were 5,7,15,20,30,30 min, respectively, providing indirect evidence for the colonic utilization of dietary glucose in at least 2 of the infants. In the absence of formula, AT for ^{13}C-G decreased to 10,15,10,10,2,10 min, respectively. This suggests that the CHO load in the formula may have contributed to the small bowel malabsorption of glucose in those infants with near maximal formula AT values.

649 NUTRITIONAL REHABILITATION OF MALNOURISHED CYSTIC FIBROSIS PATIENTS BY NOCTURNAL GASTROSTOMY FEEDING.

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Progressive malnutrition associated with deteriorating lung function in CF patients is a major clinical problem. Our initial approach is intensive dietary counselling and oral supplementation. In 14 subjects (5 male, mean age 15.6yr; 9 female, 11.4yr) the oral approach was unsuccessful and a feeding gastrostomy (G-tube) was inserted for nighttime supplementation (1200kcal) with an elemental formula. Daytime meals with exogenous pancreatic enzymes were as usual. Changes in growth, body composition and habitual physical activity were measured. Growth rates in all subjects were initially reduced ($\bar{X}$ height velocity=69% predicted; $\bar{X}$ weight velocity=25% predicted). In response to G-tube supplementation, height and weight growth, body fat and total body potassium (TBK) were significantly increased; total body nitrogen (TBN) did not change. The table shows changes seen after a mean period of 0.8yr (range 0.3-2.5yr) of supplementation.

	Δ% Body Fat	GROUP	Δ% Ht V	Δ% Wt V	ΔTBK(g)	ΔTBN(kg)
M	+4.9(p<0.05)		+163.0	+596.0	+8.5	+0.10
F	+3.1(p<0.005)		p<0.025	p<0.005	p<0.025	NS

Two subjects with the worst lung disease died which points to the need for careful patient selection criteria for G-tube placement. In the remaining 12 subjects with restoration of a positive energy balance there has been marked improvement of habitual physical activity. We conclude that in CF patients unable to meet energy needs orally, supplemental nocturnal G-tube feeds offer a reasonable and important alternative.

652 THE DAMAGED INTESTINAL MUCOSA: STRATEGIES FOR SELECTING ORAL HYDRATION SOLUTIONS (OHS).

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Optimal concentrations of glucose (Glc) and Na have previously been determined in OHS using an in vivo perfusion model (Pediatr. Res. 17:194A, 1983). A similar approach was used to investigate the effect of varying [Na] and [Glc] in W.H.O.-type OHS on the absorption of H_2O and Na by a damaged mucosa. Mucosal injury, as evidenced by large numbers of damaged cells, was produced by a 1 hr perfusion of rat jejunal segments with 5 mM deoxycholate (DCh) added in one of the OHS. Controls (C) were perfused with DCh-free OHS. The segments were then equilibrated for 1 hr and sampled for 2 hr. Results (in ul or uEq/min. cm):

OHS mM		OHS Osmol.	Intestinal Absorption H_2O		Intestinal Absorption Na	
Na	Glc	mOsm/kg	DCh	C	DCh	C
30	55	146	$1.20\pm.13^{a,b}$	$2.91\pm.12^{b}$	$-208.6\pm23.7^{a,b}$	-100.5 ± 9.7^{b}
60	111	210	$0.63\pm.08^{a,b}$	$1.63\pm.06$	-111.9 ± 17.0^{a}	-43.2 ± 7.3^{b}
90	111	314	$0.10\pm.07^{a}$	$1.36\pm.14$	-110.8 ± 12.8^{a}	46.7 ± 7.4

[a] $p<0.01$ vs C; [b] $p<0.01$ vs 90 mEq/l Na

Our data indicate that in the DCh-damaged mucosa, H_2O absorption is better maintained with a hypotonic, low-Glc, low-Na OHS at an approximate 2:1 [Glc] : [Na] ratio. Isotonic, high Na OHS (90 mEq/l is especially inappropriate for H_2O uptake by the damaged mucosa. These results may be relevant for the design of OHS therapeutic approaches in patients with compromised mucosal integrity.

● 650 MUCOSAL AND LUMINAL IgA IN EXPERIMENTAL BACTERIAL OVERGROWTH. EFFECT OF MALNUTRITION.

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We have previously shown that malnutrition hastens the onset of mucosal pandisaccharidase deficiency (PDD) in the experimental blind loop model (Pediat. Res. 16, 177A, 1982). Since malnutrition may reduce the sIgA response and possibly encourage the growth of selective pathogens, we studied mucosal and luminal sIgA in experimental blind loops from nourished (N) and malnourished (MN) rats at varying time periods corresponding to the evolution of PDD. Self-filling (SFBL) and self-emptying (SEBL) blind loops were formed 7 cm distal to the ligament of Treitz. MN received 50% of the ad libitum intake consumed by littermates. Antibody to (N) rat sIgA was purified by affinity chromatography on Sepharose 4B linked sIgA. SIgA was determined by ELISA. N developed PDD at 4 weeks and MN at 1 week. Mucosal sIgA in N was unchanged for 8 weeks. In MN mucosal sIgA was significantly reduced at 1 week ($p<.025$), but not subsequently, although mean values were consistently lower than in N. In N blind loops, luminal sIgA rose 9.5 x to 17 x the sIgA levels in comparable segments of non-operated intestine at 1, 2 and 4 weeks. In MN, comparative levels of luminal sIgA were significantly lower at 1 week ($p<0.005$) and 4 weeks ($p<0.05$). There was no change in mucosal and luminal sIgA in SEBL. MN therefore slows and reduces sIgA accumulation in SFBL. A blunted immune response may hasten the evolution of PDD.

653 ABNORMAL RECTOANAL FUNCTION IN CHILDREN RECOVERED FROM CHRONIC CONSTIPATION AND ENCOPRESIS.

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Anal sphincter function is decreased in children with chronic constipation and encopresis, but no information is available for children after long-term treatment for CE. We compared anal tone and anal sphincter relaxation on rectal distension (RSR) in 20 healthy children (C) and 20 children with CE (Pts). Measurements were obtained using an intraluminal pressure transducer and a balloon for rectal distension prior to treatment with laxative, and were repeated in 11 Pts recovered and 9 Pts not recovered from CE after 2.5-4 years of therapy and follow-up. The anal resting tone (ART) was defined as the tone in mm Hg at the low points of the rhythmical waves, and the highest ART in the anal canal was used. Anal pressure (APr) was recorded by rapidly pulling the transducer through the anal canal. APr is the pressure difference in mm Hg between the peak of the pressure tracing and atmospheric pressure, and is computed as the average of six measurements at rest. The highest amplitude of RSR in mm Hg for distension of the rectal balloon with 60 ml of air and the lowest volume in ml to elicit RSR of ≥ 5 mm Hg (RSRT) were determined. ART, APr, and RSR60 were significantly lower in Pts than in C, and remained lower even in the recovered patients ($p < 0.05$). RSRT was similar in Pts and C. It appears that abnormal anorectal function is the underlying cause of CE.

	Controls	Children With CE		
		Pretreatment	Recovered	Not Recovered
ART	53 ∓ 12	37 ∓ 10	41 ∓ 11	42 ∓ 14
APr	133 ∓ 14	103 ∓ 28	117 ∓ 26	109 ∓ 32
RSR60	52 ∓ 13	29 ∓ 12	33 ∓ 7	32 ∓ 9
RSRT	14 ∓ 5	16 ∓ 11	20 ∓ 10	17 ∓ 10