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THE EFFECT OF DIALYSATE VOLUME ON PERITONEAL CLEARANCES AND ON THE DEVELOPMENT OF HERNIAS IN CHILDREN TREATED WITH CAPD/CCPD. Tassilo von Lilien, Isidro B. Salusky, John C. Alliapoulos, Heinz E. Leichter, Marcy Wilson, Teresa L. Hall, Richard N. Fine. UCLA Sch. Med., Div. Pediatric Nephrology, Los Angeles, California.

The relation between inflow volume (IV), peritoneal clearance (PC) and development of hernias is not well established in pediatric pts. We therefore performed 73 PC studies in 40 CAPD/CCPD pts. aged 12.4 ± 5.4 (SD) yrs. After a 2 hr dwell with different IV a peritoneal fluid sample was obtained and clearances (C) of creatinine (Cr), urea-N (U-N), potassium (K), uric acid (UA) and phosphorus (P) were obtained. According to the IV, the pts. were placed in 3 different groups (A=20-34.9, B=35-44.9, C=45-66 cc/kg). In groups A,B,C the mean PCCr corrected for 70 kg BW were 9.4 ± 3.6, 11.1 ± 3.8, 14.4 ± 4.6 ml/min respectively (A vs B p=NS, B vs C $p<0.05$, A vs C $p<0.005$). PCU-N were 1.9 ± 2.8 (A), 14.8 ± 3.5 (B) and 19.5 ± 3.5 (C) ml/min/70 kg respectively (A vs B $p<0.005$, B vs C $p<0.001$, A vs c $p<0.001$). PCK and P were significantly higher with the increased volumes (K: 11.3 + 2.8, 13.8 ± 3.2, 18.3 ± 2.2 ml/min/70 kg $p<0.05$, <0.001, <0.001). P: 8.7 ± 2.6, 8.9 ± 3.9, 12.0 ± 3.1 ml/min/70 kg, p=NS, <0.05, <0.05). Only the PCUA failed to increase with increased volumes (8.7 ± 2.5, 9.0 ± 2.4, 11.8 ± 3.7 ml/min/70 kg p=NS). Seventeen of the 40 pts developed 27 inguinal, ventral or umbilical hernias after 16.5 ± 11.7 mos of dialysis with an IV ranging between 25.6 and 48.4 cc/kg (mean 37.3 ± 5.9). These data indicate that maximal dialysate volumes provide greater C. The IV does not per se effect hernia development. Pts should be carefully evaluated for hernias at the time of initiation of PD.

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ERYTHROCYTE CARNITINE STATUS IN PEDIATRIC PATIENTS ON CONTINUOUS AMBULATORY PERITONEAL DIALYSIS (CAPD). Bradley A. Warady, Peggy R. Borum, Charlotte Stall, Joan Millspaugh, Eileen M. Taggart, Gary M. Lum, (Sponsored by Stanley Hellerstein). The Children's Mercy Hospital, Dept. of Pediatrics, Kansas City, MO; The Univ. of FL, Dept. of Food and Nutrition, Gainesville, FL; The Univ. of Colo. School of Medicine, Dept. of Pediatrics, Denver, Colo.

An altered erythrocyte carnitine status was found in 9 (Group A) of 12 pediatric patients receiving continuous ambulatory peritoneal dialysis (CAPD). Upon entrance to the study, Group A patients had been on CAPD a mean of 17.2±6.2(SE) months, while Group B patients (normal carnitine status) had been on CAPD a mean of 9±2.6 months ($p<.05$). Mean red blood cell carnitine concentration (RC) in controls was 0.157±0.06 nmol/mg Hgb. Base line mean RC was .035±.027 nmol/mg Hgb and .151±.057 nmol/mg Hgb in Groups A and B, respectively. The base line mean weekly loss of carnitine in peritoneal effluent was 1350± 952 µmol in Group A and 478±119 µmol in Group B ($p<.10$). There was no difference in the plasma carnitine concentration between the two groups and all but two patients had a normal value. Eleven of the 12 patients received oral L-carnitine (100 mg/kg) for up to 3 months. The RC increased in all patients and a normal value was achieved in 7 of 8 treated Group A patients. The RC decreased to a level below that of the normal range once carnitine supplementation was discontinued. Patients on CAPD have been reported to have normal plasma carnitine concentrations. The RC may provide a more accurate assessment of carnitine status and has not previously been measured in such patients. We have observed an altered erythrocyte carnitine status, as assessed by RC, in 9 pediatric patients on CAPD. In addition, this abnormality appears to be responsive to oral replacement therapy.

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AN EXTENDED MAJOR HISTOCOMPATIBILITY COMPLEX (MHC) HAPLOTYPE ASSOCIATED WITH MEMBRANOPROLIFERATIVE GLOMERULONEPHRITIS TYPES I AND III. Thomas R. Welch, Linda Beischel and Clark D. West, Univ. of Cinti., Children's Hospital Medical Center, Div. of Nephrology, Cincinnati.

The (MHC) includes the alleles for HLA-A, B, DR; the complement components C2, Bf, and C4; and glyoxylase I (GLO I). C4 synthesis is controlled by two loci (A and B) and unexpressed ("null") alleles are fairly common (C4AQ0, C4BQ0).

Study of 26 patients affected by MPGN I or III and their families allowed unambiguous determination of 52 "disease associated" MHC haplotypes. The extended haplotype HLA-A1, B8, DR3, SC01 (BfS, C2C, C4AQ0, C4B1), GLO I 2 comprised 8 (15%) of these haplotypes, and was present in 31% of patients. This pattern, however, was found in only 1/102 normal (i.e. not occurring in MPGN patients) haplotypes ($p<.001$). Family members who did not share haplotypes with the patients did not differ from the normal population in haplotype frequencies. The presence of C4AQ0 on this haplotype accounts for our earlier finding of a high frequency of C4 nulls in MPGN.

The identical extended haplotype has been reported by others to occur with increased frequency in insulin dependent diabetes mellitus. Without data on the GLO I variant, it has also been reported in SLE. Although A1B8DR3SC01 is a common Caucasian haplotype (8% of our normal haplotypes), the disease associated haplotype always bears the GLO I 2 variant. These data suggest that a "disease susceptibility" gene(s) may be located between the DR/complotype loci and GLO I, and may be more closely linked to them than to HLA A and B.

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SHORT TERM FOLLOW-UP OF URINARY TRACT ANOMALIES DIAGNOSED IN UTERO: VALUE OF LASIX RENOGRAPHY Delbert R. Wigfall, Hooshang Kangarloo, Richard Ehrlich, Richard N. Fine. UCLA Sch. Med., Div. Pediatric Nephrology, Los Angeles, California.

With the advent of noninvasive radiologic imaging techniques, the early diagnosis of obstructive uropathy (OU) during pregnancy has become a major challenge. We have therefore undertaken a perspective follow-up of five infants with OU diagnosed by routine fetal ultrasonography (US). In each case, repeat ultrasounds were obtained at post-natal age 24 hours, 4 weeks, and 8-10 weeks. In addition, Lasix renography was obtained by age 8-10 weeks with concomitant US. By US, one infant was found to have bilateral hydronephrosis (2° to posterior urethral valves), two were found to have ureteropelvic junction obstruction, one had ureterovesicular obstruction and one had renal dysplasia. Lasix renography reconfirmed these diagnoses and also aided in evaluation of the degree of obstruction with resultant parenchymal destruction, and hence, the need for urologic imminent intervention. US alone was insufficient to delineate the specific site of obstruction or the clinical significance of the obstruction.

These observations suggest that the addition of Lasix renography to US appears to better differentiate functional versus anatomically significant obstruction. Therefore, Lasix renography should be included early in the evaluation of obstructive uropathy detected by fetal ultrasonography.

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IRON BALANCE IN A DEFEROXAMINE (D) TREATED HEMODIALYSIS PATIENT. Jerold C. Woodhead, Douglas N. Weismann, and Pedro A. de Alercon. University of Iowa, Department of Pediatrics, Iowa City, Iowa.

To study the route of iron removal by D in an anephric, hemodialyzed, iron-overloaded adolescent we performed iron-balance studies before and after D infusion on 2 separate occasions. Study #1 lasted 5 days and included 2 dialyses after D infusion. Study #2 lasted 4 days and included 1 dialysis after D infusion. Following baseline dialysis, carmen red was given as a stool marker and D was infused (2g IV, over 4 hrs). Carmen red was given again at the end of each study and all stool was collected between the markers. D was infused only once in each study. Dietary iron averaged 7.2 mg per day.

	STUDY #1 (Days)					STUDY #2 (Days)				
	Base	1	2	3	4	5	Base	1	2	3	4
Stool Fe (mg)	8.5	├---109.9---┤					15.6	├--------88.0--------┤			
								22.6	44.4	13.1	7.9
Dialysate Fe (mg)	0.8	5.8			1.5		0.8			7.0	

Average post D fecal iron in both studies was 22.0 mg/day, but study #2 showed that the majority was excreted during the 2 days following D infusion. Iron removal by dialysis was minimal compared with the fecal route. Our data demonstrate that D effectively removes iron when infused after dialysis and that stool is the principal route of iron removal.

† 1656

FAVORABLE EFFECTS OF L-CARNITINE SUPPLEMENTATION DURING HEMODIALYSIS (HD). Gaston Zilleruelo, Michael Freundlich, Milan Novak, Jose Foradada, Carolyn Abitbol, Jose Strauss. Department of Pediatrics, University of Miami, Miami, Florida.

Carnitine (C) plays an important role in lipid metabolism, being essential for the optimal oxidation of free fatty acids. Deficiency of C (MW 168), secondary to losses in dialysis effluents, has been described in patients undergoing HD. Attempts to correct hypertriglyceridemia by adding intravenous or oral L-Carnitine have given variable results. We studied the effects of adding L-Carnitine to dialysate fluid (2 g/dialysis x 4 weeks) on serum triglycerides (TG), free fatty acids (FFA), free carnitine (FC) and total carnitine (TC) in 10 patients ($\bar{x}$: age 18 yrs; range 9-21) undergoing chronic HD. Results ($\bar{x}$ ± SD) were:

Groups	Carnitine µmol/L		FFA	TG
	FC	TC	µmol/ml	mg/dl
Pre-trial	34±9*	59±15	0.55±0.26	236±146*
Post-trial	51±*,**	92±11*,**	0.44±0.27	186±123*
Normal Values	43±10	55±12	0.55±0.28	60±25

*p <0.01 compared to normals **p <0.01 compared to pre-trial.

These data suggest: a) children on HD have significantly decreased FC and increased TG; b) carnitine losses can be replaced by adding L-Carnitine into dialysis fluid; c) significant increase in FC and TC with a concommitant decrease in TG (average 28%) occurs after four weeks of L-Carnitine supplementation. This reflects a better oxidation of FFA with the resultant decreased synthesis of TG. Long term beneficial effects of L-Carnitine supplementation on dialysis hyperlipidemia deserves further evaluation.