

Table 1
Demographics, body composition, resting metabolic rate, fasting and postprandial fat and carbohydrate oxidation, diet induced thermogenesis and dietary intake in people with PKU and Controls.

	Children		Adults		All	
	PKU	Control	PKU	Control	PKU	Control
Gender (M/F)	6 (4/2)	6 (4/2)	10 (4/6)	9 (3/6)	16 (8/8)	15 (7/8)
Age (years)	11.5 ± 2.0	11.0 ± 2.2	33.9 ± 5.0	28.8 ± 5.9	25.5 ± 11.9	21.7 ± 10.2
BMI §	0.71 ± 1.7	0.53 ± 1.3	27.8 ± 8.3	25.9 ± 8.4	–	–
PKU severity **	0, 100	–	40, 60	–	25, 75	–
Metabolic control ***	237.5 (5%)	–	917 (100%)	–	546 (46%)	–
Compliance to overall PKU management ‡	87 (16)	–	65 (49)	–	76 (29)	–
Compliance to prescribed protein substitutes ‡	90 (7)	–	73 (42)	–	84 (18)	–
Compliance to prescribed protein exchanges ‡	79 (27)	–	60 (29)	–	68 (30)	–
$\dot{V}O_2$ max (ml/kg/min)	43.7 ± 9.7	44.7 ± 11.9	32.4 ± 6.3	34.4 ± 12.4	36.3 ± 9.5	38.1 ± 12.9
FM %	28.9 ± 7.9	27.8 ± 8.3	39.4 ± 8.2	34.3 ± 11.1	35.5 ± 9.43	31.7 ± 10.3
FFMI (kg/m ²)	14.4 ± 2.9	13.7 ± 2.0	18.9 ± 3.0	19.1 ± 2.8	17.2 ± 3.68	17.0 ± 3.68
FMI (kg/m ²)	6.3 ± 3.5	5.5 ± 2.3	12.9 ± 4.6	11.0 ± 5.8	10.4 ± 5.28	8.86 ± 5.43
RMR (kcal/day)	1266 ± 597	1308 ± 394	1660 ± 489	1611 ± 440	1511 ± 553	1489 ± 436
RMR/FFM (kcal/kg/d)	35.6 ± 8.5	42.0 ± 9.6	29.4 ± 5.8	28.3 ± 4.9	31.8 ± 7.3	33.7 ± 9.7
RMR/ BW (kcal/kg/d) ‡	23.9 (9.0)	26.8 (16.4)	18.8 (3.9)	17.6 (3.6)	19.3 (6.7)	20.3 (9.5)
Fasting FOx (mg/min)	55.0 ± 15.2	44.8 ± 15.2	80.0 ± 13.3	111.7 ± 33.0	70.6 ± 10.2	61.9 ± 9.6
Fasting CHOOx (mg/min)	104.8 ± 30.7	147.7 ± 43.1	126.3 ± 18.6	86.6 ± 18.8	118.3 ± 16.0	136.9 ± 26.3
Fasting RER (mg/min)	0.85 ± 0.03	0.86 ± 0.08	0.81 ± 0.05	0.82 ± 0.06	0.82 ± 0.04	0.84 ± 0.07

DIT (% increase above RMR) ‡	18.5 (62.0)	14.0 (23.5)	10.3 (38.0)	16.9 (29.5)	10.6 (53.9)	15.6 (21.3)
DIT (increase as % of EI) ‡	10.5 (7.7)	4.4 (6.8)	2.5 (4.8)	8.2 (8.8)	5.2 (8.5)	7.0 (7.1)
Postprandial FOx (mg/min) ‡	48.1 (32.0)	49.0 (17.6)	58.4 (43.0)	58.0 (30.5)	55.7 (34.0)	51.4 (23.4)
Postprandial CHOOx (mg/min) †	177.5 ± 42.1	169.8 ± 23.9	211.2 ± 14.7	235.5 ± 22.5	198.6 ± 17.9	209.2 ± 18.1
Postprandial RER (mg/min)	0.88 ± 0.02	0.89 ± 0.02	0.88 ± 0.04	0.89 ± 0.04	0.88 ± 0.04	0.89 ± 0.04
MPAD (min/week) †	138 ± 52	264 ± 84 *	27 ± 10	77 ± 26 *	73 ± 26	152 ± 43 *
MPAEE (kcal/week) †	618 ± 226	972 ± 187 *	221 ± 115	587 ± 215 *	404 ± 127	741 ± 153 *
EI (kcal/day) †	1965 ± 216	2208 ± 154	2721 ± 509	2138 ± 292	2343 ± 287	2173 ± 156
EI from carbohydrate (%) †	63 ± 4	52 ± 4 *	57 ± 3	50 ± 3	60 ± 2	51 ± 2 *
EI from sugar (%) †	13 ± 5	9 ± 2	15 ± 4	17 ± 3	14 ± 3	13 ± 2
EI from fat (%) †	21 ± 2	33 ± 4 *	27 ± 3	37 ± 3 *	24 ± 2	35 ± 3 *
EI from protein (%) †	16 ± 2	15 ± 1	16 ± 2	13 ± 1	16 ± 2	14 ± 1

Values are mean ± SD unless indicated, t-test for comparison

§ BMI: kg/m² for adults and Z-scores for children

** classical (%), mild-moderate (%) PKU

*** Median of phenylalanine plasma levels in µmol/L (% of phenylalanine measurements above age-specific treatment targets recommended by the medical research council⁽¹⁾)

‡ Median (interquartile range)

† Mean ± SE

* p value is significantly different (p≤0.05)

PKU, phenylketonuria; M, male; F, female; BMI, body mass index; $\dot{V}O_2$ max, maximum of oxygen consumption; FM, fat mass; FFMI, fat free mass index; FMI, fat mass index; RMR, resting metabolic rate; FFM, fat free mass; BW, body weight; FOx, fat oxidation; CHOOx, carbohydrate oxidation; RER, respiratory exchange ratio; DIT, diet induce thermogenesis; EI, energy intake.

⁽¹⁾ MRC (1993) Recommendations on the dietary management of PKU. Report of Medical Research Council Working Party on PKU. Arch Dis Child 68, 426-427.