

## GROWTH AND BODY COMPOSITION IN CHILDREN WITH PHENYLKETONURIA (PKU)

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When dietary regimens were first introduced for the management of PKU, a major concern was that a dietary restriction of phenylalanine (PA) might interfere with normal growth. A cross-sectional analysis of growth data from our clinic demonstrated a lower standard deviation (SD) height score ( $P < 0.01$ ) in the younger children (4-6yr) than in the older children (6-11yr),  $-0.66 \pm 0.94$  vs  $0.17 \pm 0.85$ , respectively. Despite this the younger patients were consuming higher amounts of protein  $2.3 \text{ g/kg} \pm 0.2$  vs  $2.04 \text{ g/kg} \pm 0.33$ ,  $P < 0.001$ , respectively. These results suggested that the restricted PA intake to maintain lower plasma PA levels in the younger children might have interfered with such patients achieving their true growth potential.

To assess this possibility a prospective study with sibling controls was instigated in prepubertal children to determine their protein nutritional status and the influence of plasma PA on this parameter.

Protein nutritional status was assessed by total body nitrogen measurements using in-vivo neutron activation analysis. Lean body mass was calculated from estimations of body fat from 4 skinfold measurements. Dietary protein, energy and PA intake was measured by 4 day weighed food records. Plasma PA levels, height and weight were also recorded.

Preliminary dietary data (mean  $\pm$  SD) shows lower fat but similar energy intake between the 2 groups. The percentage of energy from protein and carbohydrate was higher and fat lower for the PKU children compared to their siblings. As was expected, the PA intake was higher in the controls.

	Energy (kJ/kg)	Fat (g/kg)	% Energy:			
			Protein	Fat	Carbohydrate	PA (mg/kg)
PKU (21)	$266.1 \pm 55.0$	$1.45 \pm 0.45^{***}$	$16 \pm 7^*$	$21 \pm 8^{***}$	$63 \pm 6^{**}$	$30.3 \pm 15.1^{***}$
Siblings(13)	$262.8 \pm 32.6$	$2.42 \pm 0.30$	$11 \pm 2$	$38 \pm 5$	$51 \pm 6$	$96.6 \pm 20.9$

Different from controls by Mann Whitney \* $P < 0.05$ , \*\* $P < 0.0005$ , \*\*\* $P < 0.0001$

There was no difference in the mean SD height and weight scores, percentage body fat, lean body mass and total body nitrogen between the 2 groups.

However, the younger PKU children (<6yr) had a lower mean SD height score than the older PKU children ( $-1.02 \pm 0.47$  vs  $-0.03 \pm 0.92$ ,  $P < 0.06$ ) although the numbers are small. Also, the younger children with PKU had total body nitrogen measurements below the average previously established for normal children.

These preliminary results support our initial observation that younger children with PKU have a compromised growth pattern.

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