Royal Cornwall Hospital NHS Trust

"A Modified Macronutrient Diet for Children with Prader-Willi Syndrome Does Work"

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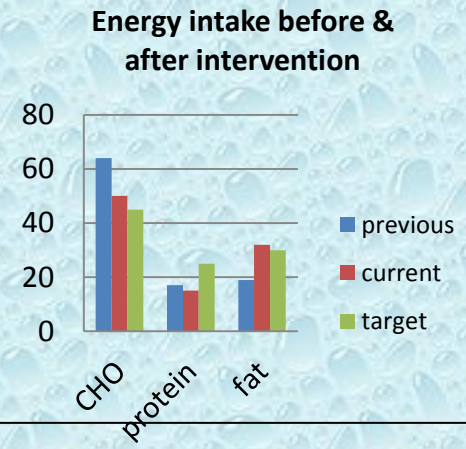
Background:
Children with Prader-Willi syndrome (PWS) have a predictable pattern of weight gain, with obesity beginning in early childhood and worsening as they get older. They have low tone and as a result their energy requirements are lower (typically 60% estimated average requirement for energy, EAR) than age matched controls[1].

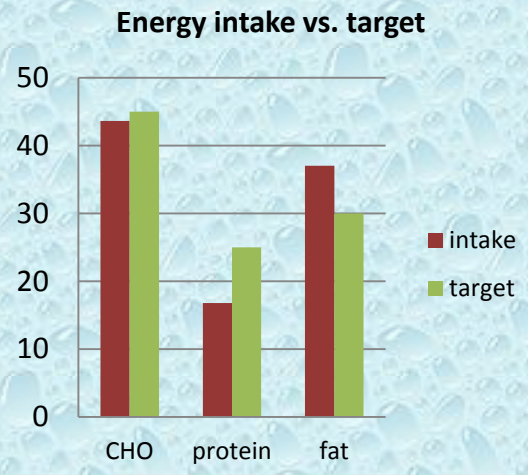
The modified macronutrient diet aimed to increase the proportion of energy from protein (25%) and fat (30%) and lower the proportion from carbohydrate (45%) compared to guidance for the general population, as well as having a high fibre content (>20g/day)[1].

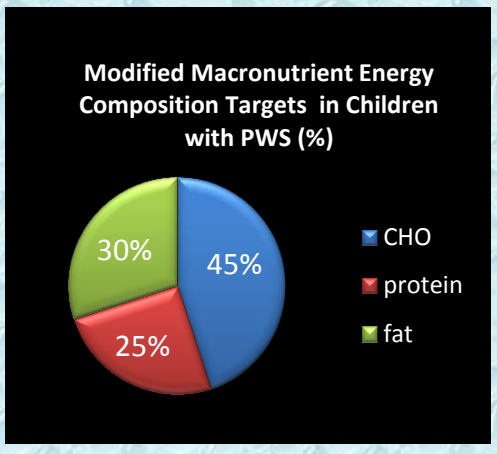
Individuals worked with their dietitian to create personalised stepwise plans to move their current intake towards these levels. Follow-up measurements were taken between 9 and 12 months after starting the dietary changes.

Case 1:
An 11 year old boy
- BMI decreased from 26.7 (>99.6th centile) to 23 (98th centile).
- fat mass decreased from 24.5kg (41.4%) to 19.6kg (35.2%)
- muscle mass increased from 32.9kg to 34.2kg.
- fibre intake 22.7g



Case 2:
18 year old female and
- fat mass reduced from 30.7kg (43.6%) to 27.6kg (39.2%)
- muscle mass increased from 37.6kg to 40.7kg





Case 3:
A 5 year old boy, is following the same diet but is too young to accurately measure bio-impedance with current equipment.

Positive effects on BMI have been seen though from 22.1kg/m^2 (above +3.5sd) to 19.1kg/m^2 (above 98th centile) over a 1 year period

Conclusion:
The results from these three cases indicate that an energy restricted diet with a well-balanced macronutrient composition and fibre intake improves both BMI and body composition in children with PWS compared to a simple energy-restricted diet.

References:
1. A reduced-energy intake, well-balanced diet improves weight control in children with Prader-Willi syndrome. JL Miller et al. Journal of Human Nutrition and Dietetics 2012 pages 2-9
2. Nutrient intake of young children with Prader-Willi syndrome. M Lindmark et al. Food and Nutrition Research 2010. 54:2112