



# Feeding problems in Barth Syndrome: the UK perspective

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## Background

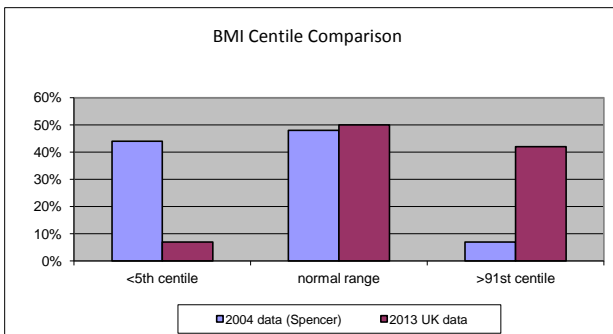
Feeding problems are common in Barth syndrome (BTHS): many boys are slow to transition to solid food, have poor chewing skills and small portion sizes compared to their peers. Supplemental feeding is often given due to concerns about slow weight gain. Barth Syndrome Foundation Registry 2010 data shows that 46% of all males in the registry had been fed by enteral tube [1]. A recent national French survey showed long term enteral feeding in 70% [2]. Since its inception, the NHS Barth Syndrome Service has cared for 25 males with the disease, of whom 21 are currently alive. Regular face-to-face and telephone dietary advice has been given by a qualified specialist paediatric dietitian. Height, weight and body composition have been recorded by bioelectrical impedance analysis (Tanita BC-418) in all boys aged over 7 years on an annual basis. Data is presented to show that increasing the rate of weight gain faster than the rate of growth leads to gains in body fat and not increases in height; by keeping calories at the appropriate level and fat intake to at or below current guidelines (35% of energy), body fat percentage can be kept within the healthy range. Careful monitoring of supplemental enteral tube feeding is required to prevent excessive weight gain. Supplemental feeding is not indicated if weight gain velocity is in concordance with height velocity.

## Types of Diet in the UK cohort

- 10% (2) are currently receiving long term enteral feeding.
- 28% (6) have been taken off enteral feeds and are now eating and drinking orally.
- 3 of these 6 remain on prescribed nutritional supplement drinks to maintain their weight.

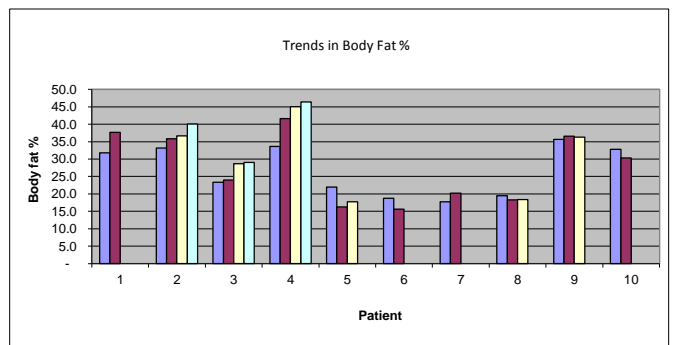
## Changes in Body Mass Index (BMI)

- BMI is used to assess if weight is appropriate for height.
- The healthy range BMI changes according to age, and under 18 years BMI centile charts are used.
- BMI has been increasing over the last 9 years therefore increases in weight gain have not led to increases in height growth.
- BMI has limitations and does not discriminate between muscle or fat.



## Trends in Body Fat Percentage

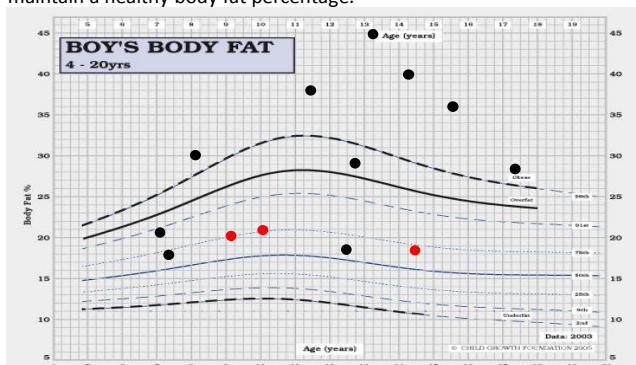
- Body fat percentage increases in patients whose rate of weight gain is greater than their rate of growth.
- Patients who maintain a weight gain velocity in concordance with their growth maintain a stable body fat percentage.



Bars represent body fat % measured using Tanita BIA scales at annual reviews for patients over 7 years. Patients 1-4 have rates of weight gain in excess of growth and show increasing body fat percentage each year. Patients 5 and 6 have been exclusively tube fed and patient 7 has a high intake of nutritional supplements. These have a controlled amount of calories and fat and have kept a stable body fat percentage in the healthy range. Patients 8-10 have maintained or reduced their weight gain velocity in line with their growth by reducing the calories and fat in their diet and have a stable body fat percentage.

## Body Fat Percentage

- Body fat percentage in patients over 7 years ranged from 14.9-45.3% (median 24%). The healthy range for boys aged 7-18 years is 10% to 22%.
- Therefore 54% of these males have body fat percentage higher than the healthy range.
- Patients who receive a controlled amount of calories and fat (35% of calories) from tube feeding or nutritional supplements (shown in red below), or who have smaller portions and a lower fat diet are able to maintain a healthy body fat percentage.



McCarthy HD, Cole TJ, Fry T, Jebb SA, Prentice AM. New body fat reference curves for children. *Obesity Reviews (NAASO Suppl) A156*

## Late Catch-up Growth

- The rapid growth phase in BTHS occurs between 18 and 22 years.
- This typically involves crossing several height centiles and the average BTHS male reaches the 50th centile for height [2].
- It is unclear whether body fat percentage will return to the healthy range spontaneously and consistently during the pubertal growth spurt. DXA scans are being performed annually to assess this.

## Recommendations

- Being significantly overweight is likely to have negative effects during adolescence including increasing fatigue and muscle pain.
- Keeping weight velocity tracking no more than 2 centiles above height centile is a working target.
- Careful monitoring of supplemental enteral tube feeding is required to prevent overfeeding. Supplemental feeding is not indicated if weight velocity is in concordance with height velocity.
- Healthy eating advice focused on appropriate calorie intake and keeping fat intake to at or below current guidelines (35%) is recommended.

## References

- [1] The Barth syndrome registry: distinguishing disease characteristics and growth data from a longitudinal study. Roberts AE et al., Spencer CT: *Am J Med Genet A* 2012, 158A:2726-2732
- [2] Natural History of Barth syndrome: a national cohort study of 22 patients. Rigaud C et al. *Orphanet Journal of Rare Diseases* 2013, 8:70.