**Introduction**

The classic 2-stage model of nutritional status in Prader-Willi syndrome (PWS) – initial failure to thrive due to hypotonia and poor feeding, followed by hyperphagic obesity (Figure 1) – has been replaced by a more complex 7-stage concept which includes a period aged 20-31 months during which weight increases without an obvious change in appetite (Miller et al, 2011)

**Aim of study**

To examine trends in body mass index (BMI) in order to determine a particular age zone at which inappropriate weight gain begins in preschool children with PWS.

**Methods**

BMI was plotted for each clinic visit in patients attending the PWS clinic at the Royal Hospital for Sick Children, Glasgow, over a 20-year period. BMI trends were categorised as showing a) no inappropriate increase in BMI (Fig 2); b) no definable age for BMI increase since child obese at referral (Fig 3); a definable age at which BMI increased (Figs 4 and 5).

**Results**

The case sheets of 76 patients were studied of which 36 had BMI increase since child obese at referral (Fig 3); a definable age at which BMI increased (Figs 4 and 5).

- No inappropriate weight gain had occurred in 10 patients by the time of study (see Figure 2). Seven were still young at 1.5 - 2.0 years in four patients, and 1.8 – 2.0 years in two patients, with three patients aged 6.6, 11.5 and 15.2 years at last data point.
- Age at BMI increase was indeterminate in 9 patients, all of whom were obese at age of first data point (see Figure 3). Median (range) age at presentation in this group was 3.2 (2.4-5.0) years.
- Age at onset of BMI increase could be ascertained in 21 patients (see Figures 4 and 5).
  - While their median (range) age was 2.0 (0.5-3.8) years, nine patients showed onset of BMI increase at ≤ 1.5 years; with onset 1.8 – 2.0 years in four patients, and ≥ 2.3 years (2.3, 2.5, 2.6, 2.8, 2.8, 3.8 and 3.8 years) in the remaining eight patients.
- No significant correlation could be found between age at onset of BMI increase and either gender or genotype.

**Discussion**

- Our data support the observation that weight gain in PWS may start **before** hyperphagia has become an obvious behavioural feature (e.g. 2-4 years).
- The median age at onset of BMI increase in our series was 24 months, which is within the 20-31 month range described by Miller et al.
- However, nine of 21 patients in whom age at BMI increase could be ascertained were aged 18 months or younger.

**Figure 1 At what age did the trend towards weight gain start in this boy with Prader-Willi syndrome?**

**Figure 2 No inappropriate increase in BMI trend**

**Figure 3 Age at BMI increase not ascertainable (obese at presentation)**

**Figure 4 Age at BMI increase ascertainable boy becoming obese**

**Figure 5 Age at BMI increase ascertainable in patient responding to diet**

**Conclusion**

- The phenomenon of weight gain preceding hyperphagia in PWS is attributable a decrease in energy expenditure due to hypotonia, which is particularly marked in the early years.
- A dual approach to management is needed from an early age:
  - giving a diet which is appropriate to age and body size
  - increasing energy expenditure to reach that of normal children (rather than reducing intake to offset the effect of the hypotonia)
- Practical measures to increase activity in infants and preschool children with PWS, including regular swimming, should be explored with the family.
