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Introduction- Aims

Children with Prader-Willi syndrome (PWS) show alterations in each phase of infantile, childhood and pubertal growth when compared to the normal population. Growth Hormone (GH) therapy is recommended due to reported improvements in height velocity, muscle tone and body composition. This evidence has been reflected in a change in our clinical practice during the last 5 years: increased prescribing of GH and initiation of GH at a younger age.

The aims of the study were:

- To describe the patterns of growth in children with Prader-Willi syndrome attending our clinic in years 2000-2017 with regards to height and BMI
- To identify changes in growth parameters (height and BMI) between the periods 2000-2012 and 2013-2017
- To explore the effects of GH treatment on growth patterns

Methods

Sixty children (29 males/31 females) who attended the dedicated Prader-Willi clinic between 2000-2017 were included in the study. A total of 391 measurements of height and weight, with median number of 6 (1, 17) measurements per subject, were included. Only 1 measurement per year per subject was included. The median age at first measurement was 1.46 years (0.54, 16.21), the median age at last measurement was 10.07 years (1.04, 17.93) and the median duration of follow up was 5.36 years (0.5, 15.77).

- Comparison of growth parameters (height and BMI) to those for healthy children (expressed by Standard Deviation Scores, SDS)
- Comparison of growth parameters between children attended in 2000-2012 and children attended in 2013-2017
- Comparison of height SDS, height velocity SDS and BMI SDS in those children who had height and weight measurements available at -1, 0, 1 and 2 years in relation to the date of start of Growth hormone therapy

Results

Comparison with healthy children - Phases of Growth

Height SDS

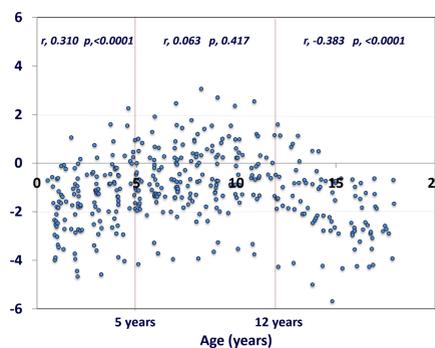


Figure 1. Height SDS against age

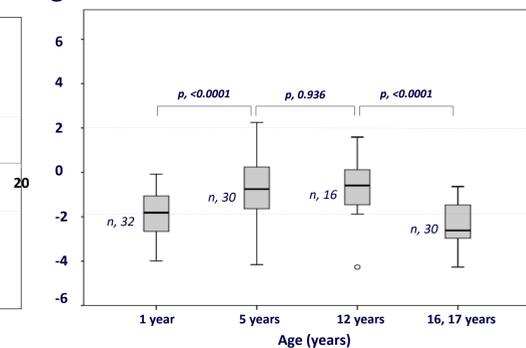


Figure 2. Height SDS at 1 year, 5 years, 12 years and 16, 17 years

BMI SDS

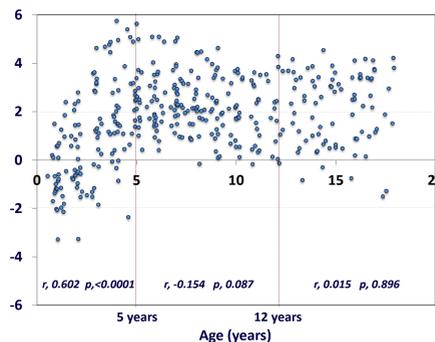


Figure 3. BMI SDS against age

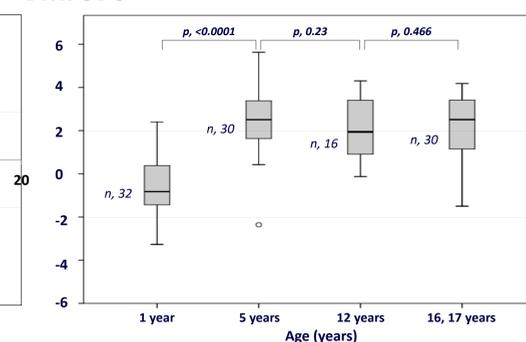


Figure 4. BMI SDS at 1 year, 5 years, 12 years and 16, 17 years

Phases of growth: **1-5 years**, with acceleration in both Height SDS and BMISDS; **6-12 years**, with stabilisation in both Height SDS and BMISDS; and **13-18 years**, with deceleration in Height SDS and no change in BMISDS.

Fourteen children had annual measurements available from 1-8 years

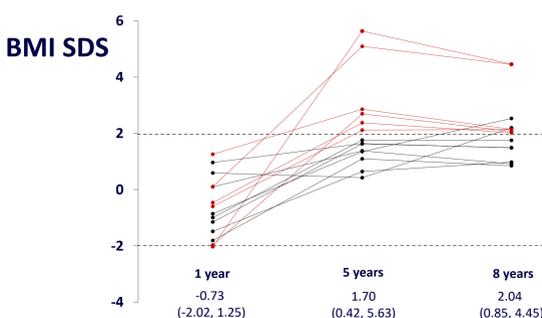


Figure 5. BMI SDS against age.

None of the 14 children had BMI>2 SDS at age 1 year. Six of those had BMI>2 SDS at age 5 years and all six of them (100%) continued having BMI>2 SDS at the age 8 years. Of the eight children who had BMI<2 SDS at the age of 5 years, only two (25%) had BMI>2 SDS at the age of 8 years.

Comparison of growth parameters between periods 2000-2012 and 2013-2017

Height SDS

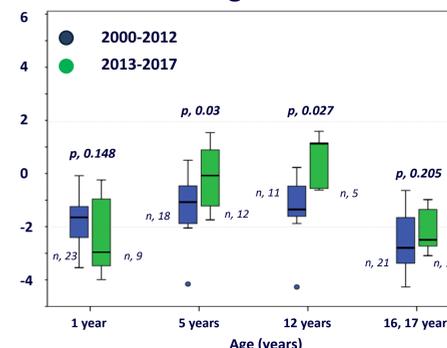


Figure 6. Children in the 2013-2017 were taller at the age of 5 years and at the age of 12 years.

BMI SDS

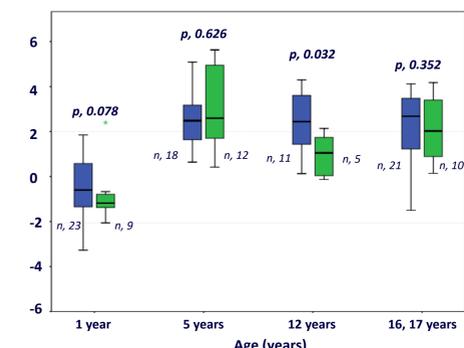


Figure 7. Children in the 2013-2017 had a lower BMI SDS at the age of 12 years.

Growth Hormone effect on growth parameters

Height SDS

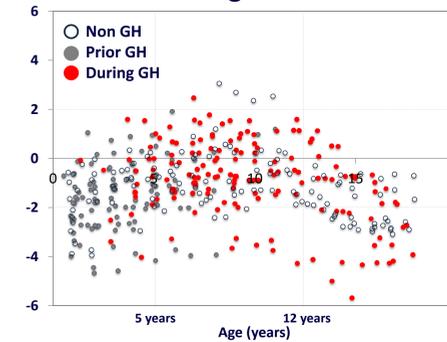


Figure 8. Height SDS against age of children prior and during GH therapy and of children who never received GH therapy.

BMI SDS

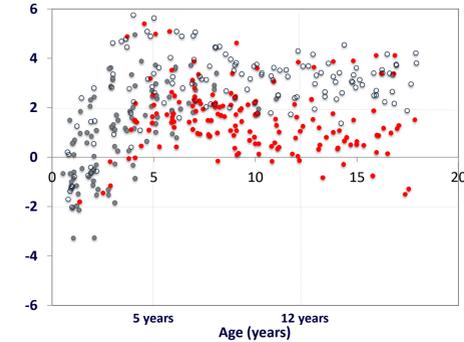
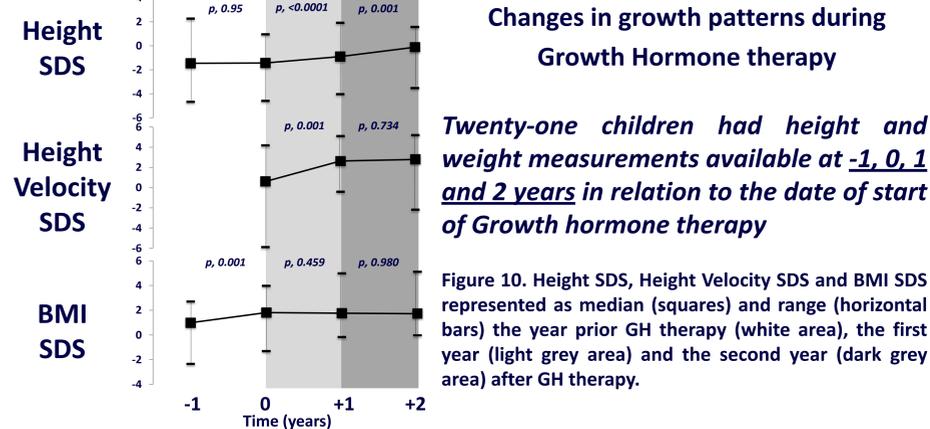


Figure 9. BMI SDS against age of children prior and during GH therapy and of children who never received GH therapy.



Changes in growth patterns during Growth Hormone therapy
Twenty-one children had height and weight measurements available at -1, 0, 1 and 2 years in relation to the date of start of Growth hormone therapy

Figure 10. Height SDS, Height Velocity SDS and BMI SDS represented as median (squares) and range (horizontal bars) the year prior GH therapy (white area), the first year (light grey area) and the second year (dark grey area) after GH therapy.

Conclusions

- Three phases of growth were observed: **1-5 years**, with acceleration in both Height SDS and BMI SDS; **6-12 years**, with stabilisation in both Height SDS and BMI SDS; and **13-18 years**, with deceleration in Height SDS and no change in BMI SDS.
- Children in the 2013-2017 period were taller at the age of 5 years and they were also taller and with a lower BMI SDS at the age of 12 years
- Growth hormone therapy improved Height SDS each year during the first two years of therapy. Height Velocity SDS improved during first year of therapy and was then maintained during the second year. BMI increased in the year prior to Growth hormone therapy and was stabilised after Growth hormone initiation.