The switch in eating behavior in infants with Prader-Willi syndrome is associated with an increase in the AG/UAG ratio
Results of a longitudinal study

R.J. Kuppens[1,2], A.J. van der Lely[3], P.J.D. Delhanty[3], T.M. Huisman[3], A.C.S. Hokken-Koelega[1,2]

[1]Dutch Growth Research Foundation; Rotterdam, The Netherlands
[2]Dept of Pediatrics, Erasmus University MC-Sophia Children's Hospital, Rotterdam, The Netherlands
[3]Dept of Internal Medicine, Erasmus University MC Rotterdam, The Netherlands
R.Kuppens@kindengroei.nl

Conclusion

The switch from failure to thrive to excessive weight gain and hyperphagia in PWS coincides with an increase in AG/UAG ratio

Background

Prader-Willi syndrome (PWS) is characterized by a switch from failure to thrive to excessive weight gain and hyperphagia in early childhood. Hyperghrelinemia may be involved in the underlying mechanisms of the switch.

Ghrelin exists in two forms. Acylated ghrelin (AG) stimulates appetite and induces a positive energy balance. Unacylated ghrelin (UAG) acts as a functional inhibitor of AG. Thus the AG/UAG ratio may be crucial.

Aim

To assess the evolution of the appetite regulating hormones AG and UAG and the AG/UAG ratio in infants and children with PWS and to investigate their associations with the switch in eating behavior.

Participants & Method

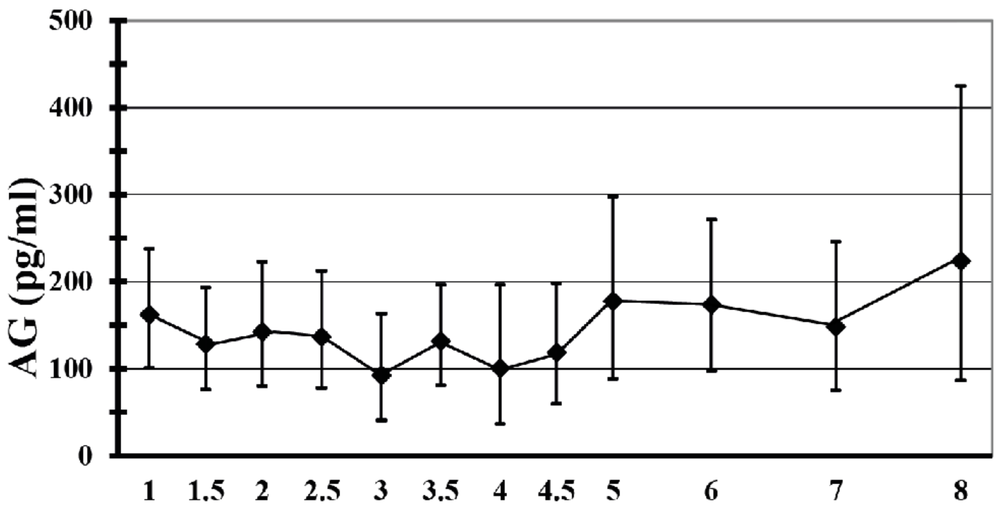
Longitudinal study in 44 infants and children with PWS aged 1 to 8 years during growth hormone (GH) treatment. Serum AG and UAG were assessed six-monthly for 2 to 5 times. AEBSF was added to the blood samples to inhibit deacylation of AG. Mixed models was used for analysis.

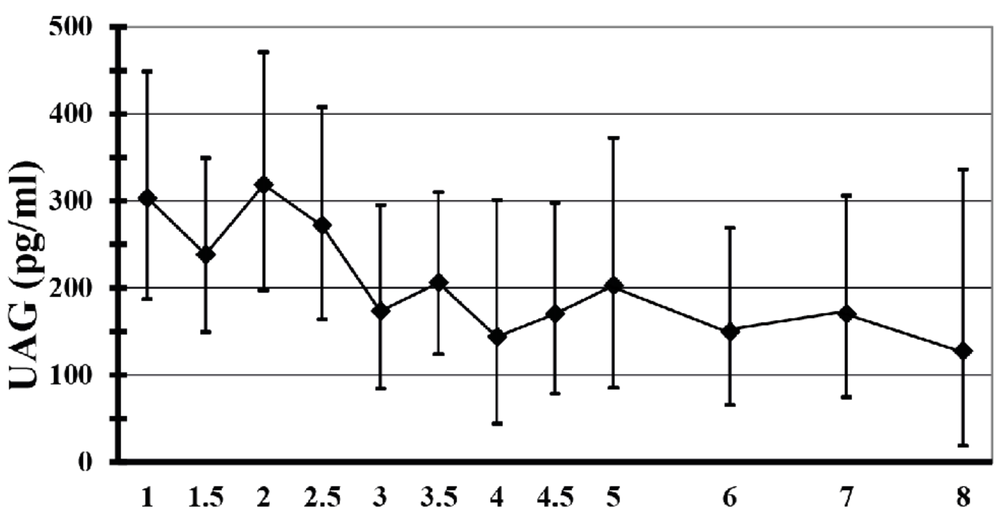
Baseline characteristics of 44 infants and children with PWS

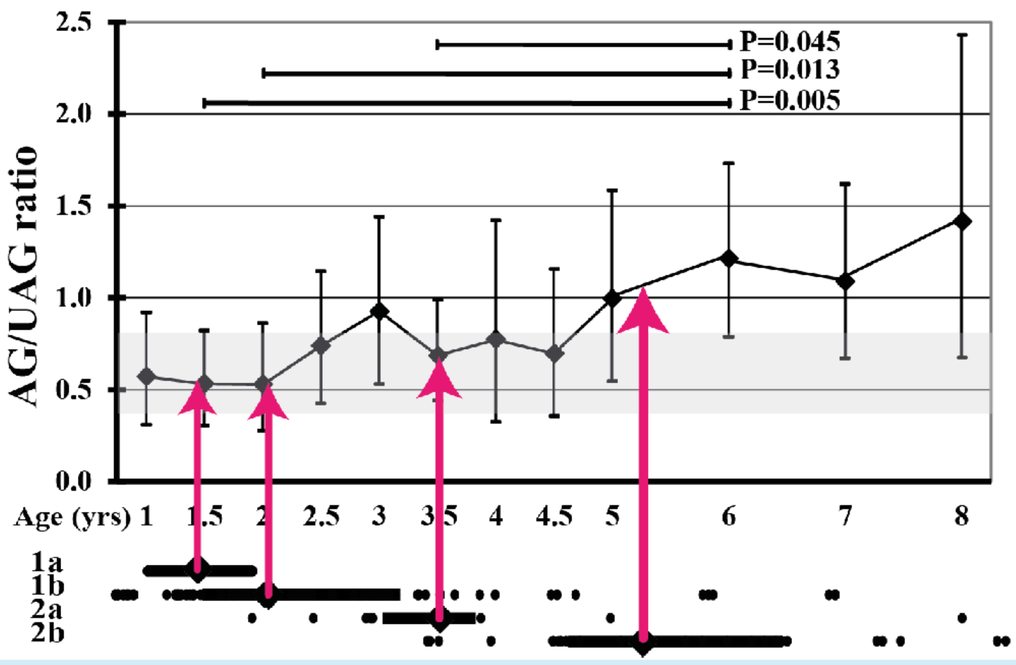
Age (years)	2.6 (1.1 to 4.6)
Gender (male / female)	22 / 22
Genetic subtype	
- Deletion	22 (50%)
- mUPD	18 (40.9%)
- ICD	1 (2.3%)
- Unknown	3 (6.8%)
Samples per child (2 / 3 / 4 / 5)	20 / 11 / 10 / 3
Height SDS	-0.2 (-0.9 to 0.3)
Weight for height SDS	-0.1 (-1.7 to 1.2)
BMI SDS	-0.3 (-1.7 to 1.3)
Fat percentage (%)	24.4 (21.3 to 33.5)
GH duration (years)	1.9 (0.5 to 3.7)
Nutritional phase[a]	
- Phase 1a	2 (4.5%)
- Phase 1b	30 (68.2%)
- Phase 2a	6 (13.6%)
- Phase 2b	6 (13.6%)

Age, Height SDS, Weight for Height SDS, BMI SDS, fat percentage, and GH duration are expressed in median (IQR); gender, genetic subtype, sample samples per child and nutritional phase are expressed in n(%). [a]Nutritional phase according to Miller et al.

Results







- AG levels decreased slightly during the first years of life and increased subsequently. UAG levels decreased until the age of 8 years.
- AG/UAG ratio was around 0.5 in the first 2 years of life and thereafter it increased gradually until an AG/UAG ratio of 1.4 at 8 years.
- At the age of 6 years, with the majority of children being in phase 2b, the AG/UAG ratio was significantly higher than at the age of 2 years, with the majority of children being in phase 1b.

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Renske Kuppens
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