

## Prader-Willi syndrome

### Adrenarche, gonadal function, cognition, psychosocial aspects and effects of growth hormone treatment in children

1. Children with PWS aged 3 to 10 years, have significantly higher DHEAS levels, are younger at onset of pubarche and have a higher prevalence of premature pubarche than healthy peers. (this thesis)
2. Primary testicular failure, which becomes apparent after onset of puberty, appears to be the main reason of hypogonadism in the majority of male patients with PWS and not hypogonadotropic hypogonadism. (this thesis)
3. The primordial follicle pool and number of small antral follicles are conserved and the impairments of maturation of follicles and progression of pubertal development are not absolute, thus ovulation and conception cannot be ruled out in girls and female adolescents with PWS. (this thesis)
4. GH-treatment prevents deterioration of cognitive skills in children with PWS on the short term and improves abstract reasoning and visuospatial skills on the long term. (this thesis)
5. Health related quality of life in children with PWS increases during GH-treatment, in contrast to untreated children with PWS. (this thesis)
6. Over the recent years, early diagnosis, use of multidisciplinary teams, introduction of GH-treatment, control of the food environment, and better understanding of the behavioral and psychiatric aspects have made significant improvements in the life of those with PWS. (A.P. Goldstone et al. *Journal of Clinical Endocrinology and Metabolism* 2008 Nov; 93 (11): 4183– 4197)
7. Excessive fruit juice consumption is associated with increased risk for obesity. Sucrose consumption without the corresponding fiber, as is commonly present in fruit juice, is associated with the metabolic syndrome, liver injury, and obesity. (J. Wojcicki. *American Journal of Public Health* 2012 Sep;102(9):1630-3)
8. Off-label use of drugs in paediatrics is associated with a significantly increased risk of adverse drug reactions, however, off-label use in children is so common that it is often not even mentioned when such a drug is administered. (C. Lenk et al. *European Journal of Pediatrics* 2009 Dec;168(12):1473-8)
9. Good doctors use both individual clinical expertise and the best available external evidence, and neither alone is enough. (David L Sackett. *British Medical Journal* 1996 Jan;312(7023):71-2)
10. We become ourselves through others. (Lev S. Vygotsky, Russian psychologist)
11. As it net sile wol, moat men lavearje. (Gysbert Japicx, Friese renaissanceschrijver)

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