



6th International Prader-Willi Syndrome Caregivers' Conference

21-23 May 2024, Berlin, Germany

Ageing in PWS

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Workshop Summary Ageing in PWS

Workshop Leaders:

Susanne Blichfeldt, MD, The Danish Prader-Willi Association, IPWSO Clinical and Scientific Advisory Board (CSAB), Denmark and Larry Genstil, Psychologist, Executive Director, Genstil Institute of Human Behaviour, Israel

Workshop description:

This workshop took place from day 2-3 of the conference. During the workshop, participants focused both on physical and mental ageing, and the social and family circumstances. Issues such as fatigued and/or ageing parents, or when parents pass away were also discussed.

The goals of the workshop were:

- to equip participants with knowledge in the themes of ageing in PWS that require caregiver support;
- to give participants tools to support people with PWS who have reached ages in which there is decline;
- to provide participants an understanding of the varying needs of this population as they continue to age.

The workshop also allowed ample time for open discussions between participants, with brainstorming of ideas and activities that will positively impact ageing people with PWS.

The 30 participants came from many countries in Europe and also some from USA. Most work daily with adults with PWS, and many have years of experience with adults with PWS above the age of 40-50 years. All expressed a wish to learn more about ageing in PWS and to be prepared for how to manage the environment and support people with PWS as they become older.

Workshop presentations

Susanne Blichfeldt, [Ageing in Prader-Willi syndrome \(click to link\)](#) which focused on which medical conditions are common in PWS, and which if left untreated can lead to early ageing and death. See abstract, **Appendix A**.

Larry Genstil, [Issues of Ageing in PWS \(click to link\)](#) which focused on support needs for older people with PWS in social situations.

After both presentations there were Q&A sessions with many important points made, and participants also shared their presented experiences on the topics.

Breakout sessions:

Participants worked in smaller groups to discuss the below topics related to ageing in PWS, and at the end of the workshop presented on key points.

Many of the participants have worked with people with PWS for several years, and there were examples of working with the same individual over years, or decades, where the caregiver observed changes in both physical symptoms and mental state with age.

The below-mentioned observations are all based on individual cases which participants reported, so we can not conclude that the recorded situations are typical for all people with PWS, but they are important to know about and be aware of should the same be seen in others.

Food, eating/weight, and physical condition

- Weight loss was observed
- Difficulty gaining weight after weight loss
- Weight gain was also observed
- More difficult to lose weight gained after family visits
- Eating more slowly
- Increasing difficulties with chewing and swallowing: cases were mentioned where blended food and/or tube feeding was needed
- Losing interest in eating
- Less interest in food and eating was not a general observation (as proposed in the literature) for ageing adults, but some eat more slowly with age
- Despite getting older, many look forward to family visits and the food served there
- Gradual loss of energy was often seen

Cognition and mental health

- Losing interest in activities and hobbies which were previously enjoyed. Noted this was lost at an earlier age than expected (compared to the general population)
- Dementia was often raised as a topic, but seems not to be often reported in the PWS population
- Depression and other psychiatric conditions were observed, also arising in older age

So, about the reported cases in the workshop, we can say that in general there are changes to be aware of with increasing age, especially after age 40-50. But those with PWS are not ageing in a specific “PWS-way”, there are many differences.

Susanne notes that from a medical point of view, it could be of value to know the physical condition, who is treated for hormonal deficiencies, has a diagnoses of type 2 diabetes, and so on, to determine if any of these factors influence the ageing process.

Key questions

One of the breakout groups also discussed ***How to determine if decline in function is decline or depression or a medical problem?***

And their recommendations were:

- Regular medical assessment to monitor physical development
- Regular cognitive assessment (for example, Montreal Cognitive Assessment, MoCA) to monitor the cognitive development
- Thorough knowledge of the individual person with PWS so the caregiver will be able to identify any change in behaviour
- Working with the person with PWS from a young age to help them to notice and put words on feelings and physical sensations/changes, like pain, and so forth

And a breakgroup was also asked ***What therapeutic tools can we use to help ageing with PWS process feelings?***

Recommendations were:

- Social stories
- “Theraplay” - Sensory integration/play: putting sensory stimulation into words

Conclusion

The participants were keen to learn about ageing in PWS, but the workshop leaders have to acknowledge that there is so little literature about ageing in PWS. There are case reports, and a study looking at a particular group of adults, but their medical conditions were very different. So today, the real experts about ageing in PWS are the caregivers who have worked with individuals with PWS across years and decades.

Our hope is that all adults with PWS will receive necessary medical treatments (hormonal deficiency, avoiding obesity and thereby type 2 diabetes, measures to avoid osteoporosis) secure safe and supportive surroundings, and treatment for psychiatric diseases when needed.

More research is needed about ageing in PWS, to know if hormonal deficiency causes early ageing, what the ageing process in PWS looks like when there are no untreated physical problems, no diabetes and normal weight, and meaningful occupation and engagement. There are still many questions to ask, so that we can give the best possible care and support for people with PWS as they age.

Bonus interview

Watch workshop leaders Susanne and Larry answer some commonly asked questions about ageing with PWS.

Video link: <https://youtu.be/uZ8KdabyzD8>

Ageing in Prader-Willi Syndrome:

Medical conditions and diseases to be treated

Susanne Blichfeldt, MD

The Danish Prader-Willi Association, IPWSO Clinical and Scientific Advisory Board (CSAB), Denmark

Introduction: Prader Willi syndrome (PWS) is a complex disease. Thanks to growing knowledge, treatment possibilities and care, persons with PWS now survive into adult age and can live years healthy and happy, and face the possibility of becoming “old” and then need extra support.

Methods: Many treatable symptoms in PWS are well known. We need more systematic investigations about how adults are actually treated in various countries, how often they have medical contact and which medical treatment is given. We know from the general population that diseases often found in PWS, if untreated, can cause early ageing, reduced lifespan and perhaps dementia. Conditions to be aware of in PWS are: Hypogonadism, growth hormone deficiency, hypothyroidism, diabetes, obesity that causes heart and circulatory problems, respiratory diseases including sleep apnea, vitamin deficiency and physical inactivity.

A recent Dutch investigation * showed that among 115 adults with PWS 61% had one or more untreated health problems, and 25% had multiple untreated health problems.

Psychiatric diseases can arise and be treated. Dementia is not a common symptom in PWS, but if suspected testing and care is possible.

Due to ageing, new needs and symptoms can arise and individual support is always needed.

Conclusion: Knowing symptoms and diseases that can be seen in PWS, and knowing from the general population the effect on ageing and survival if these conditions are not treated, it is recommended that all with PWS regularly receive lifelong medical evaluation and treatment together with continuous information and counselling for caregivers and families. A proposal for systematic medical contact will be presented.

*Reference:

Pellikan K et al: Missed Diagnoses and Health Problems in Adults With Prader-Willi Syndrome: Recommendations for Screening and Treatment

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