



IPWSO
International
Prader-Willi Syndrome
Organisation

IPWSO Health ECHO abstract

September 13, 2021: Diet and Exercise and the Prevention of Obesity

Please note this document is abridged from audio transcription of the Zoom session. Some errors resulting from the transcription process may be present.

IPWSO hosts: Georgina Loughnan (GL) Marguerite Hughes (MH)

Presenters: Constanze Lämmer (CL), Georgina Loughnan and Nikolinka Yordanova (NY)

Video links

Dr Constanze Lämmer, PART 1 of Diet and Exercise and the Prevention of Obesity: [Nutrition in PWS](#). PDF available [here](#).

Georgina Loughnan, PART 2 of Diet and Exercise and the Prevention of Obesity: [The Power of Exercise for People with PWS](#). PDF available [here](#).

[Q&A Following Contanze and Georgina's Presentation](#)

MH: I find it very interesting and welcome, the extent to which you emphasise the psychological element. It's not the case that everybody appreciates that.

Q: It's often a challenge to get the whole team on the same page, to the point where some team members will think there is not enough food in the house and they need to increase or supplement the amount. How do you try and get the whole team onto the same page with the same diet?

GL: A group home team, or family?

Q: A group home team.

GL: We do a lot of group home training and if a client is coming to see us in our clinic it's essential that the group home staff come as well, because they're really the ones that we're training. We're not training the client with PWS. We virtually challenge them, that if they're prepared to have someone with PWS live in their service, they need to provide the right environment, and this is what it's all about. It's not a matter of asking them, we tell them this is what they have to do.

Q: In the Netherlands we have the Dutch plan and many people with PWS use it. So they can choose their own food within the regulation of (for example) 1500 calories, but choose their own products to have 6 times through the day. We use that plan so they have choice. This helps to lose weight. And all of our Prader-Willi people move about 30-60 minutes a day so that is always our advice.

GL: That's excellent. As Constanze mentioned in her talk, we talk a lot about organising the food and managing the food and planning the food but certainly as people with PWS settle into where they are living or whatever the plan may be, once they are used to a new menu plan or a new exercise plan there is a lot of room for negotiating. My clients come with a list of things they would like to have and we discuss what they would need to remove in order to have that and still not gain weight. Negotiating is really important, especially because they have so little choice in their lives. We just set up the boundaries, and there is lots of choice within those boundaries.

Q: Does GH modulates the impact of exercise. In other words, do you get more muscle mass gain in someone who's on GH when they exercise than in people with PWS who aren't on GH when they exercise or is there no data?

GL: I think that answer is yes, you should do, provided they do as much exercise, however I would have to say the calf muscles I showed, and a couple of other clients of mine who've been exercising so regularly from a young age probably have better muscles than some of the guys on GH I know. We've only just started seeing people on GH over the last few years because it hasn't always been available for our age group.

So technically yes, they should have better muscles on GH because GH improves their muscle, not always up to the normal level, but pretty close. But can the muscle be equal just through exercise? It looks like it possible could be.

CL: I agree. I'd like to add that we see if someone is doing exercise on a regular basis and we have to stop growth hormone because the individual reached their final height, we can see in the body composition that muscle mass remains stable and they will not lose muscle mass after stopping GH.

On the other side, families that do not want treatment with GH but that are very focused on exercise and nutrition also have very good results in weight management and (the person with PWS) also has good body composition, in the lower normal range, if you compare muscle mass with that of their peers. A lot of things are possible with exercise and nutrition but it's hard work. For some families it's easier to give GH. We have to educate the families that GH is not instead of nutrition and exercise, it's only an additional component.

Case presentation (Abridged)

GL: Nikolinka Yordanova is a pediatric endocrinologist and core specialist of the PWS Multidisciplinary team at the First Pediatric Clinic, University Hospital "Sveta Marina" in

Varna, Bulgaria. Her clinical research interests are in the field of rare endocrine diseases, especially Prader-Willi syndrome.

Case presented:

Age and gender:

11-years old boy (born in Spain)

Diagnosis and date of diagnosis:

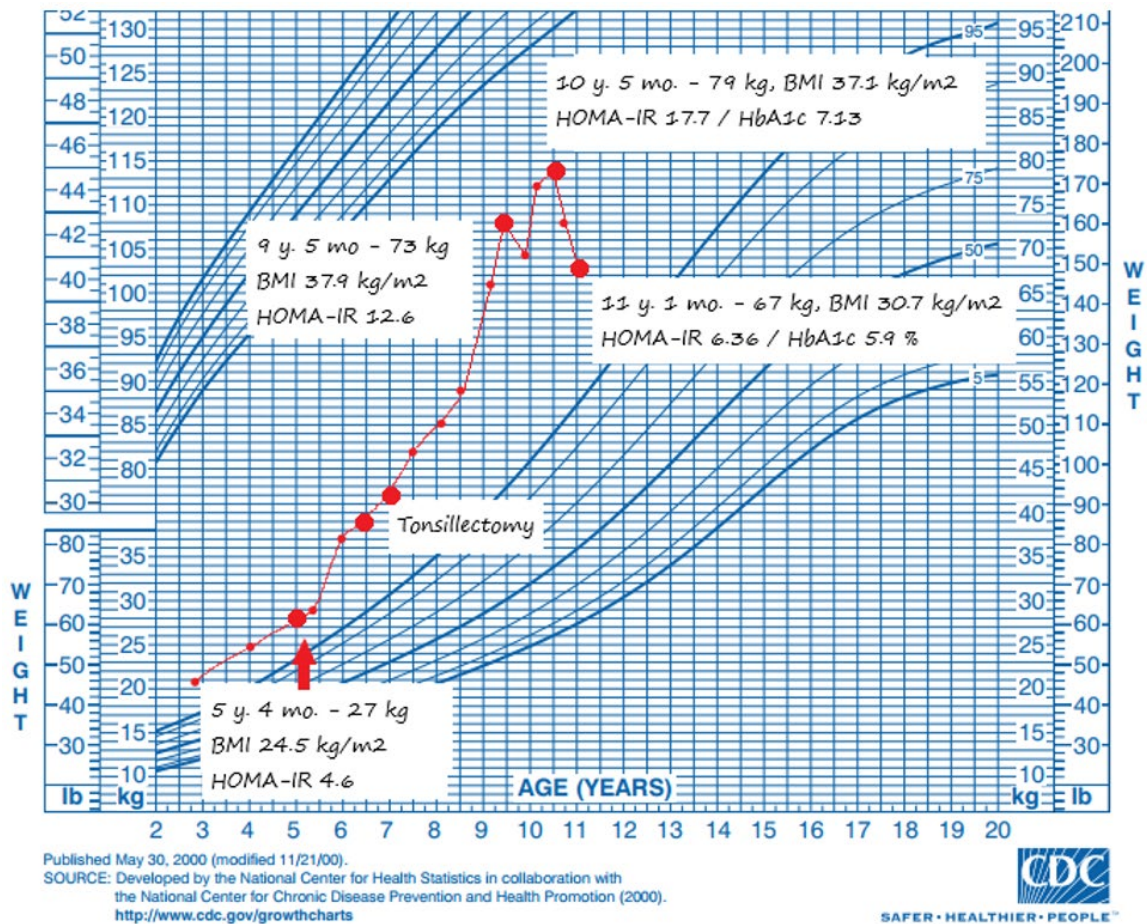
Prader-Willi syndrome diagnosed at two years of age (September, 2010 in Spain)

Treatment and significant information to date:

Growth hormone (from November, 2015 when he moved to Bulgaria), Metformin (from January, 2020), Chlorprothixene hydrochloride (from July, 2021)

Medical history:

- A boy born from first pathological pregnancy, C-section in 30-th week of gestation.
- Weight 1350 g; Length 40 cm;
- Complicated postpartum period: intubated, 80 days in incubator, tube feeding, severe hypotonia.
- Cryptorchidism of right testicle;
- Delayed milestones (walking at the of 1 y. 7 mo., speech at the age of 3 y.)
- Family history: mother – Morbus Basedow-Graves; father – severe obesity and allergic asthma
- Start of GH – at the age of 5



Psychosocial:

- Mild mental disability;
- Never worked with speech therapist and psychologist;
- Non-acceptance of the diagnose – unrealistic expectations that lead to disappointment and depression;
- The mother is raising her child ALONE, without help and empathy from her family;
- Lack of communication with the other parents from BG Organization/IPWSO – refusal to face the future and to receive help.

[Discussion following Nikolinka Yordanova's case presentation](#)

NY: I have some questions for discussion. They are more on the endocrinological side.

Do you use Metformin in PWS patients with severe obesity and similar metabolic parameters? When do you recommend it?

CL: Thank you for a very good presentation. We also use Metformin in such cases, especially if we have this high HOMA index. In the case of insulin resistance it's very helpful. We also do it in the same dosage, meaning we start with 500mg twice a day, and we have good experience with this medication. Often a side effect is that we have less obstipation problems under metformin and this is helpful for the family.

NY: When should we stop GH treatment if we have metabolic complications on board?

CL: It's an individual decision, when to stop growth hormone. GH is in competition with the high insulin levels you showed us. My experience is that if you have such high HOMA indicates the GH is not having a good influence on body composition. You have to balance the pros and cons. We always stop GH if we have diabetes which needs therapy with insulin. We continue GH when we use Metformin, and we can make an agreement with the family – that means the family opts for nutrition management and exercise. If we see, for example, rising weight, and no other success, then we also stop GH because GH is more risky for diabetes than it would be helpful. We make the decision together with the family.

NY: About HbA1c, for example if it is over 7.5 I think this is an indication to be stopped?

CL: Yes, this result of the HbA1c with Metformin and you cannot reduce this with higher dose of Metformin there is no benefit from GH and it is better to stop.

NY: With this boy we did not stop the GH. The family took the notes and made the changes in their daily routine, changed house, there is no free access to food, they changed the daily regimen. GH cessation was not done, for now, and I hope it will not become worse again.

We treat our patients sometimes with sub-optimal doses. This patient at the beginning was treated with suboptimal doses of GH (0.015-0.020 mg/kg/day) - do you think it was connected with poor control of the weight or is it just food restriction that counts?

CL: GH dosage is in competition with the GH level. High insulin means that is building up body fat and GH is supporting the building of muscle mass. We also often do not give the 0.035mg per kg because it would be a very high dosage, especially in your case, so we also do suboptimal doses. On the other hand we see also that IGF1 levels are high in children with high BMIs. Do you know about the IGF levels in this case?

NY: They are always at the upper limit of normal because of the weight. This is why we cannot increase the dose.

CL: That's correct. In that case I also would not increase the dose because we have to be more aware of other side effects. So we don't increase the GH, we have to lower the insulin level.

NY: Yes, we have worked very hard on this through all these ups and downs. It has been a big battle, but I think the result is good for now.

What do you think of the decision for the orchidopexy, postponing it because of the high risk of anaesthesia at that time so we waited for him to lose weight, but is there risk of malignancy? I did not find papers in medical literature connecting malignancy and PWS.

CL: We discuss these situations with our children's' surgeons and they told us there is not a big risk for developing a tumour if you have a situation where you can control the testes. If you have a chance to control it by ultrasound maybe every 6 months, if the boy shows up. If you can't control by ultrasound then they would decide to proceed with orchidopexy because then the risk for developing a tumour is too high. If you follow the anaesthetic guidelines and have low dosage it should not be high risk. This is the position of our children's surgeons.

NY: This was the position of our surgeons as well but he was really obese and we were upset by that. The bad thing here was that we could not follow the testes on ultrasound, we just have to follow MRI and can't do it every 3 months or 6 months so maybe the next step is surgery.

CL: If you look at <https://www.orphananesthesia.eu/> if colleagues follow these guidelines there should be no problems.

NY: How do you motivate the parents of your children to establish connections with the parents of other patients with PWS? This mother is upset every time she speaks on the phone with another parent. Especially if the patient is older than her son, she thinks they present to her what will happen in the future. After this she feels depressed and she refuses to establish connections. I cannot motivate her.

CL: We also have such families. What we sometimes do is that we mostly have 3 or 4 families attend the same day at the hospital. Then she will be given an appointment close to another family I think they will benefit from and we put them in a room side by side, so she meets other families by accident. Sometimes you can manage which families she will come in contact with first. So we organise the accidental meeting.

NY: We will try to manage this somehow. Thank you. So the take home message for me is that most important is food security and support from the family. Because if somebody is not following the rules, the situation becomes out of control.

GL: Thank you Nikolinka for a great presentation. As Constanze describes getting families to meet accidentally is also something we do with adults. If one adult is having trouble accepting the idea of moving to a group home I have another adult who comes in and has a bit of a chat with them which can work. Maybe getting her to meet someone, even in another country who has a child of the same age can help as well. I guess it's a matter of continuing to try.

Marguerite Hughes Summary

We heard very clearly how an appropriate and healthy diet can be designed and implemented and maintained for someone with PWS, and the benefits that this will bring. We heard a really strong and convincing argument for why exercise should be seen as a core component of any management strategy starting from birth and right throughout life. We had a brief but very interesting discussion on the extent to which

regular life-long exercise, beginning at a young age, can compensate for an absence of GH in cases where that is not available. Also it was clear that families need to understand that GH does not in any way eliminate the need for a good diet or exercise and that they compliment each other and both ideally should be available.

From the points that were raised, also in our Case Presentation, it was quite striking the extent to which we highlighted that so much ultimately depends on capacity of families or caregivers to implement the advice that professionals and indeed other families can give them. All of the speakers today made clear that positive outcomes are certainly possible, but they are by no means inevitable, even with good information and access to high quality services. Whether families can afford to buy healthy food and not rely on ready meals, or buy a treadmill, or have the time to support somebody to do exercise, or have the time to prepare fresh food – a lot does rely on individual families and for lots of reasons that means that success doesn't always happen. So I think there is an ongoing question for everyone as to how families who are particularly vulnerable or simply don't have the capacity to manage this syndrome can be better supported so that people with PWS can have better outcomes. I think that was illustrated very well by the case study where ultimately the mother involved had to move into a separate apartment. Her entire life was changed because of the necessity of providing support to the 11-year-old.

All-in-all I think we had a very strong and interesting discussion and I hope everyone took something from it.

Upcoming Health ECHO sessions

Tues 12 th October	<i>Caring and Living with PWS in Italy</i> Dr Maria Giulia Marini, Fondazione ISTUD, Milan, Italy
Tuesday 9 th November	<i>Socialisation and Communication in People with PWS</i> Norbert Hödebeck-Stuntebeck, Psychologist, Diakonische Stiftung Wittekindshof, Germany
Tuesday 7 th December	<i>Orthopaedics in PWS</i> Prof. Dr. Ralf Stücker, Altona Children's Hospital, Hamburg, Germany

Thank you very much to everyone who attended the session and participated. We look forward to seeing you on Session 8 in October.

Ends.