



**IPWSO**  
International  
Prader-Willi Syndrome  
Organisation

# IPWSO Health ECHO abstract

## November 9, 2021: Socialisation and Communication in PWS

*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) Tony Holland (TH)

Presenters: Norbert Hödebeck-Stuntebeck (NHS), Raquel Corripio (RC)

### Video link

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Norbert Hödebeck-Stuntebeck, [Socialisation and Communication in PWS](#)

PDF available [here](#).

### [Q&A Following Norbert's Presentation](#)

**Q:** You say that you feel we should educate and train the people with PWS and their careers to understand these different ears. Do you find that you're able to train most people with PWS in this concept or is it limited to some that may be higher on the IQ level?

**NHS:** I do not think it depends on IQ. I think we can find ways to describe this, maybe visualize by painting things. When you paint these or make it visual, these four ears these four heads with big ears, to the people with PWS they are really able to work with these pictures and to make meta communication with themselves when we support them. Yes, it works, and it works very good with support of staff.

**TH:** What I wanted to ask is whether people with PWS can generalize from one situation to another. I could imagine how in a particular setting you might help someone with PWS to better use their four ears, but do they then learn to do the same in a different setting?

**NHS:** Yes, they do, if we are able to support the transfer. These days, we make a training program and involve these things, and it is a group of people with PWS who live in different houses. I also did it, I think four or five years ago, without staff, only with people with PWS, and it worked very good in the room, and in this setting, but they did not transfer it in a good way to their working field or living field. So this time we include the staff, so that they see what we are doing.

We gave them homework, after each session, and they support the homework, and they support the transfer, and this is a much better way, then doing it without the staff, or it could be with parents.

GL: Do you think one ear is predominantly used when people with PWS are anxious?

NHS: Not really, what do you think?

GL: I think possibly the interpersonal ear. It's all about them when they're anxious. I think sometimes, even before they're anxious, if they see people talking they're immediately assume that the people are talking about them. Especially if it is carers, or myself and a parent. And I think it's teetering on anxiety but possibly more it's this interpersonal ear, it has to be that we have to be talking about them, and that causes anxiety.

**NHS:** And if we can make clear that they do not talk about you or that that or we can make clear what the others are saying, the anxieties, goes down. I think that you're right, with the interpersonal ear, that they open it very big. I think if they if you can go with them on a meta level in this situation that they have this concept of these four ears, maybe then we can talk with them about what happens now. Now your interpersonal ear is going bigger, you know, now it is coming up. It could be helpful to understand what happens in this moment for them to handle the situation better.

**TH:** Do think that what we're seeing in people with PWS is the same as any of us would respond, just in a sense, more marked and more obvious? Do you think the phenomena are the same? When Georgina asked that question about anxiety, I thought, how would I be? What would my ears do in anxiety and I think they might do the same, but it may not be quite as dominant.

**NHS:** When I do this with staff and train in this communication module I always make a task with them - I give them 15 sentences and they should bring the answer to the different ears. What they find out is that maybe one of them has a big interpersonal ear, one of them has a big appeal ear... It's the same, but the difference is that they can do this meta level and they can think about themselves and see what happens in the situation. If they are a good staff. If not, then they handle in the same way as people with PWS handle situations. But if they are trained in this, and this is what I think we can do with people with PWS too, when they are trained in this they are more sensible about the ears, and can use these to reduce stress and to reduce conflicts.

My question to the others is, do you have experience with these four ears and talk about these with people with PWS or with the staff?

**Comment:** I was just thinking, we do we operate 12 dedicated services, I'd have loved to have some managers on because they would witness a lot of interaction. Typically, in say, an eight-person service there is quite a range of abilities. So it would be interesting to see how they interact with each other with these four ears. But it'd be very difficult

to re-educate some of the people that have lived there for some time and obviously the staff as well.

**TH:** Presumably you would want to see intervention starting in childhood.

**NHS:** Yes, and if parents have the concept of this, then they can use it. And then they can train their child to develop the ears, and know about it, and ask them on which ear they hear now, but we have to inform parents about this concept.

**Q:** You must manage people whose anxiety goes through the roof very quickly, so from nought to 10 very quickly. So you've re-educated some of them to manage communication between themselves better. Have you seen the evidence that actually people manage their anxiety better as well?

**NHS:** Yes, but not in this situation. When they come into crisis situation then you can only handle the crisis situation. But after the situation, when they calm down when the arousal is normal and we can talk with them, then we reflect on the situation and then we can talk about this concept. Maybe the first time we talk about it we paint it and so they are able to understand.

If we do this 50, 60, 80 or 100 times in the reflection I think then they can use it in one of the next situations but not the first time - it needs time.

**Q:** And most people are responsive? Because some of our people don't like to revisit a situation which has been particularly stressful for them and possibly others they're living with. Are they receptive to revisiting the event maybe 24 hours later or an hour later?

**NHS:** Yes, they are. But in the same way as we do when we should talk about negative things, we try to avoid it. But if they know that this happens, maybe a day or two later, it's more and more normal, and as Tony said when we start with children to reflect on their behaviour, then they are able to do this when they're 20 or 30. So we have to start this very early, as early as possible. Normally not five minutes later or half an hour, but often on the same day, or a day or two days later. And then they can remember.

**GL:** Do you think we could ever get to the point where parents and caregivers, and even professionals could say to the person with PWS, "I am now talking to your appeal ear. I'm now talking to your factual ear." once they understand the concept?

**NHS:** Yes. We have to we have to do it in different ways. Some like to paint it, for someone else it may be helpful to make a role play. There are different ways to transfer these concepts. Yes, but we see that people are able to do this, but not in the first weeks.

**GL:** I imagine it would be really good training for professionals as well for medicos, etc. Because so often they will just assume that the patient is being uncooperative or in a bad mood, when really they're just perceiving it differently.

**TH:** We think of people with PWS as having an atypical pattern of development or delayed development. To some extent, what you described is what one would see in children, an inability to understand the meta aspects of communication, but of course we develop in a way that hopefully allows us to do that better. Do you think this is understandable in terms of delayed development or atypical development? How do you actually explain the phenomena that you described and why it affects people with PWS?

I just wonder whether you think that what you've described in terms of the communication problems is a reflection of delayed development, generally. Because I think in children you would see these problems differentiating the different years but in PWS it continues into their adult life. Is that a useful or is that an unhelpful way of thinking about it?

**NHS:** We have to accept and have to find out what is the developmental age in different areas. If we know where they are, then we can start things. If someone is in a situation that he cannot walk, and we cannot train him to run. Yes, we have to support and to find out, in which areas they are in the developmental age of 3 or in the developmental age of 10, or in the developmental age of 18, because maybe he's 20. And we see that he's cognitive, in a way of 2 years, that we cannot use training programs which needs developmental age of 10 or 12. And so there are limits, and we cannot do it to all of them in the same way. This is what we see in the training program there. There's one in this program who is not able to do it at the same speed as the others do, so we can't overwhelm her.

**RC:** Can you recommend any paper on educating our four ears, some supplementation for careers and to teach our families, how to educate that?

**NHS:** We have no paper just a manual for when we do this whole program, then there's a one day for staff or for parents to understand these concept and to train communication. Friedemann Schulz von Thun "[Miteinander reden](#)" is a book where you is the book where you can read about the theory.

## Case presentation (abridged)

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**GL:** I now have the great pleasure of introducing Raquel Corripio, who is a pediatric endocrinologist from the Parc Tauli Hospital Universitari, in Sabadell, Spain.

**RC:** Today we are speaking about socialisation, and this was a clinical case that I thought it could be an interesting case to share. I will speak about Pitolisant as a treatment. I have the good fortune to belong to a multidisciplinary team here in Sabadell, this is the childhood team, and this is the adult team, and we treat in our clinic around 50 children and 50 adults not only from Catalonia or Barcelona, also from all of Spain. I am the clinical coordinator of the children patients.

Our patient is a female, and now seven years old. She was diagnosed with Prader-Willi syndrome at two weeks old, in Canada, and she's under growth hormone treatments in since she was 3 months.

Parents explained sleep apnea, hypersomnia related with carbohydrate intake, language delay. She was receiving physiotherapies also. She has mild skin picking, mild scoliosis, no hyperphagia at the moment.

The parents were worried about the hypersomnia and the language delay. At their first visit here in Sabadell they explained they moved from Canada 4 years ago and had lived in Spain 2 years. The first visit they asked me what I knew about the use of Pitolisant. My answer at that moment was nothing, so I had to study it and we learned that Pitolisant is a histamine H3-receptor antagonist which enhances the activity of brain histaminergic neurons that are involved in sleep, muscle tone, and was approved in 2016 for the treatment of narcolepsy. I contacted a French group that was using that for narcolepsy, and contacted with my group here in the hospital with adult pneumologists and no one had a lot of experience with that pharma.

The parents knew of a [paper](#) by the Canadian group that reported cognitive improvements in a child with PWS following this treatment. This only described the experience with 3 patients, and they conclude that the benefits of Pitolisant appear to extend beyond and improved sleepiness, improved cognition, less behavioural problems, the patient feels happier and love more. And this is the reason why we asked the hospital pharmacy for the approval to do an experimental treatment, a pilot treatment, and also to the Spain health ministry, and it was approved.

We began in September 2020 within an initial dose of 4 milligrams per day and the dose was increased every four weeks or less to the maximum dose that was 18 milligrams per day. After six months, no adverse effect was reported and the parents reported more activity, less clothes intolerance, because our patient has dislike of wearing clothing, less sleepiness, language improvement and better socialisation.

We did a complete neuro psychological evaluation before, and six months of the treatment with Pitolisant, and also a weekly cognitive online questionnaire.

I have another patient whose mother is a teacher in primary school and for her socialization is one of the most important aspects to consider in Prader-Willi syndrome patients because they often play alone during recess. They don't disturb anyone, so it's very difficult to the detect when there is a problem if you are not very alert.

After seven months, our president explained a visual aura in left eye. We did a complimentary examination by a paediatric neurologist. MRI and topological evaluation were normal. After that we did reduce the dose but the problems persisted so we stopped the treatment, and the neurological symptoms disappeared.

The parents were to leave again to Canada, so we don't we don't have more information. We will have for sure because we have a very good relationship, and we will be in contact by email.

For discussion, it would like to ask these questions:

Would you try a lower dose? The parents certainly want to because they saw improvement in some issues that were very important for them. And what is your experience with Pitolisant? I will be happy to discuss with you the clinical case. Thank you very much.

#### [Discussion following Raquel Corripio's case presentation](#)

**GL:** Many thanks Raquel, that's very interesting. I'm in Australia and we call it modafinil.

**RC:** I know it's not a common case as this experience is very small. Only one paper, our experience was in good, only at the beginning the patient improved and after that the adverse effects were bigger than the benefits.

**TH:** Can I ask whether the side effects that were described, the visual aura and the panic, are those side effects that are described with the use of Pitolisant in the general population? Are they recognised side effects of the medication?

**RC:** It was a new effect Headache is a typical effect from Pitolisant. We thought perhaps it was an aura from migraine, but it wasn't really because she never had a headache. And no, panic crises, and visual aura are not described with this treatment so it was very panicky for us also. It was after six months of treatment, and ophthalmologists, MRI, paediatrician, we couldn't find any other reason. But the fact is that after stopping the treatment these symptoms disappeared so we think it was the Pitolisant. To my knowledge it is not described in the literature.

**TH:** I guess the only way to establish whether this is a side effect would be to try it again.

**RC:** I think so. Speaking to the parents, they think that with perhaps with a smaller dose, 8-13mg per day, they can maintain the good benefits of the treatment without the adverse effects. I did a medical report to the Canadian doctors, and the intention was to try to restart the treatment with a low dose.

**GL:** I'm not an endocrinologist but I can tell you about one of our clients who is around 28, and he's been on modafinil (Pitolisant) about six years. He gets headaches, and I don't know if it's ever been equated to the modafinil, but he gets headaches so he takes paracetamol for it, and they then they pass. But he was probably one of the sleepier clients we had ever seen, and our previous endocrinologist said he used to slide off the chair, because he was so sleepy, he would just flop. And since, the modafinil he's much more alert. So, he treats the headaches but he's remained on it.

**RC:** Yes, because it's useful for him.

**GL:** Yes, very useful. He's also a lot older though.

**RC:** At the beginning the health system in Spain, and my hospital told me to try other things but when I explained the parents were so clear from the beginning that they wanted to know if Pitolisant is useful because they read about it, and I provided their documentation, I was surprised to receive permission that I could do that. And after your opinion of your adult patient, for sure in sleepiness patients I should try again, perhaps.

**TH:** Presumably, Pitolisant is treating Central Sleep Apnoea. So what would you do if this patient of yours, put on a lot of weight, and became sleepy, possibly because of Obstructive Sleep Apnoea. When would you use Pitolisant and when would you not use it for sleepiness?

**RC:** This patient was a very thin 7-year old. Not normal weight, thin, because the parents did a perfect dietary management, and she did exercise every day, so obstruction here was not a possibility. For sure if I have an obese patient, it's I think it's better and more sure to begin with a C-pap, or another treatment.

**TH:** Can I just come back to this issue of medication prescribing in people PWS, because I think this case is a very nice example of the dilemmas. We see this in in psychiatric practice all the time - do you use a medication or not? - and of course it is a balance between the likely benefits and the likely risks. I think what you have very nicely shown is that there appear to be quite substantial benefits. And if there are substantial benefits then what are the what are the risks and it seems that the risks are small. They may be having visual aura, it may be developing headaches. But if the risks are no more than that, then the benefits may outweigh the risks. I think particularly in people with Prader Willi syndrome who may be more sensitive to certain medications that's the sort of analysis one has to do all the time.

**Chat:** Sorry I cannot contribute about Pitolisant, in Dk we first do a sleep study, sleep apnea during night can be the reason for daytime sleepiness.

**Q:** Was your treatment of the patient through the Covid period? Covid has interrupted trials with some of the major drug companies in the States, because the outcomes were affected by Covid restrictions and change of lifestyle. But I doubt that that would be the case with your particular situation because you're dealing just with the drug that is affecting the mental state. Was your treatment of this patient through the Covid period or before?

**RC:** I don't think the Covid, per se, changed the response to the treatment in this patient because they were living in a little village and near to the sea, and I don't think that they were very affected by the situation. Certainly we have patients that were living in a very difficult position during Covid, but I don't think is the situation with this family. The patient was living with a little sister and she was happy, and she was

learning on an online classroom so I don't think that the situation with Covid made the situation for my patient worse.

**GL:** Am I correct in remembering from your presentation you said this little girl had narcolepsy?

**RC:** No, here in the hospital pneumologists use modafinil/Ptilosant for their treatment, but they didn't have any experience with Prader-Willi syndrome patients. So that was the reason why I contacted different investigators and for example, the French group that was doing a trial with narcolepsy patients, they also didn't have experience with Prader-Willi syndrome, and that's what the reason we asked. Because the family was looking for a place where a clinical trial was going on. They are a very highly educated family, and they have information about different families in Canada, that were very happy with the use of that medication, but we need more information about this treatment.

**Chat:** Many children with who have many activities during the day can be easily tired, and even at 7 some of them need a nap or at least a break during the day.

**RC:** Yes every son or daughter is different. From their other child they could compare what the activity, and the time asleep. It was very challenging and very difficult to combine with a normal lifestyle but I don't think that a person who is seven years old that sleeps 11 hours at night, usually they don't need a nap at 11 in the morning.

**Chat:** No not normal situation , but their sleep regulation is not normal, and if medication can help, we shall try, but they are all different in their symptoms, and their need for rest and break, and with an active family it can be very hard, because they need so much time to digest impressions ( and some children become stressed, ) thank you so much for telling about the girl and her symptoms, muchos grazias.

**GL:** We certainly see an increase daytime sleepiness even if they do have a long nap and a good sleep at night The particular person I spoke about, he had severe daytime sleepiness and narcolepsy.

**RC:** Do you relate this with obesity and obstructive apnoea, or do you think it is not related? Because I have children patients with sleep apnoea with sleepiness but I have a lot of patients with PWS in childhood without sleepiness in the day. I think this is more in adulthood than infancy, and it's regardless of their BMI.

## Tony Holland Summary

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Thank you for a really interesting presentation on socialization and communication. You started by very nicely illustrating what you call the macro communication environment - the communication of the person with Prader-Willi syndrome with his parents, with his family, extended family, but also with teachers and school, neighbours, health professionals, and so on. And just how complex that



communication environment is, and how people may arrive at different understandings within that environment.

You then went on to talk about how people with PWS may vary in their abilities but there will be a range, and if they're in the right environment they may be able to then achieve their optimal abilities within that environment.

You then moved on to consider the micro environment for me this was the particularly interesting aspect of your talk - this communication between sender and receiver and how sender and receiver changes as the other person replies he becomes or she becomes the sender and the other person becomes the receiver.

You reminded us that a lot of communication is nonverbal, and that even with verbal communication that the spoken form can vary in the way that it's delivered, the rate, the pauses between words or sentences and so on. Then the nonverbal communication, the body language, the space. It was really important for us to be reminded about the real central relevance of nonverbal communication.

We then went on to talk about the complexity of meaning, the factual, the interpersonal, the self-revelation, and the appeal forms of communication. You ask is the receiver able to understand what the sender means, and that then led to the four ears which was a helpful way of thinking about this.

You then address the question of how does this model help us, help people with PWS to improve their communication, and by improving their understanding and their communication, helping their anxiety and helping their behavior.

This really came out in the discussion as well. How you can do that ideally in childhood, but continue that into adult life. You talked about the value for example of people having homework to do, or use a role play and different ways of trying to develop these, these skills.

Then Raquel's case presentation was exactly what and ECHO is about - to hear these examples from somewhere in the world, in this case it happens to be Spain, but someone could have reported from another country about this, so that we can learn from each other because Prader Willi syndrome is rare and the experience of using something new life has taught us and is rare as well so this was a really helpful case presentation that demonstrated I think the benefits to this particular child, but also you described the dynamic when we don't have very much information about the use of this particular drug. So you are now a bit stuck because the person had apparent side effects. But we have no experience to know whether these are serious side effects or not so there's a difficult decision for the family - do they restart the medication or not?

Like Georgina I've also heard examples of individual people with Prader-Willi syndrome who have been given this medication which we also call modafinil here, and reported benefits, but it seems to me there is a clear need to

run a trial and it would be very nice if your presentation could encourage people to set up a more formal trial.

Those were the main points that I took from the didactic presentation and from the case presentation, and I thought that those were extremely helpful and informative so thank you.

## Final Health ECHO 2021

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Tuesday 7<sup>th</sup>  
December

*Orthopaedics in PWS*  
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 10 in December.*

Ends.