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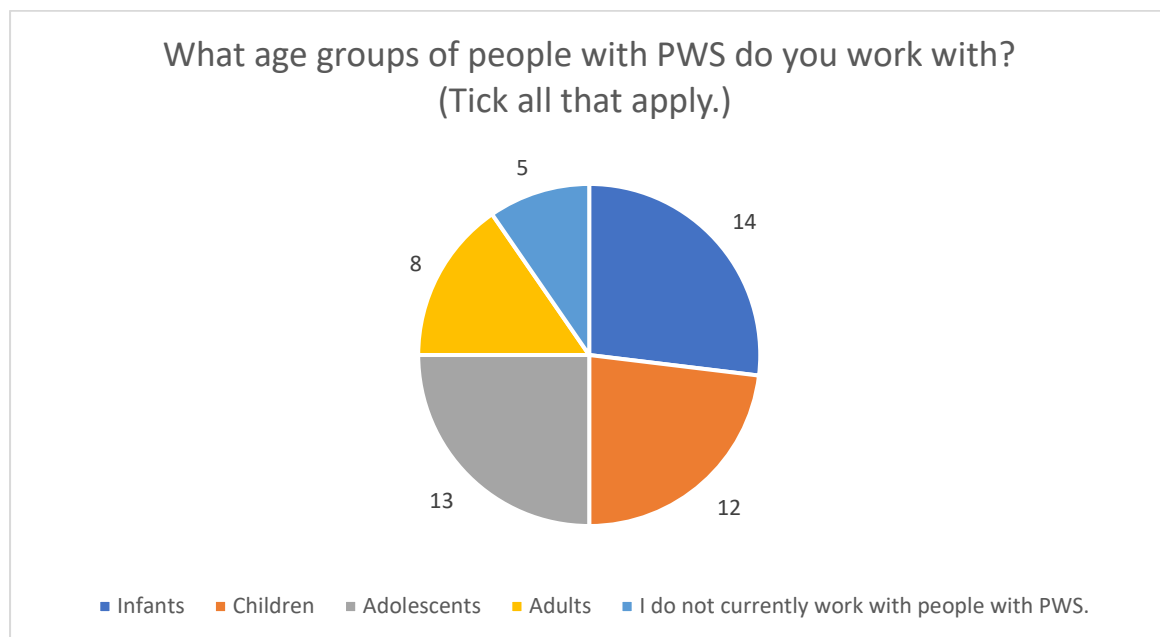
March 16, 2021: The Care of the Infant with PWS

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) and Tony Holland (TH)

Presenters: Dr Constanze Lämmer (CL) and Dr Susanne Blichfeldt (SB)

Poll Results



24 responses, multiple choice.

Video link

[Dr Constanze Lämmer: The Care of the Infant with PWS](#)

Q&A Following Dr Constanze Lämmer's Presentation

Q: I'm just wondering what the breastfeeding rates are for neonates and how do you support that if you're aiming to feed every 4 hours. Our breastfeeding rates in Ireland are quite low, particularly with these children. I'm just wondering how I can support them more, and what the breastfeeding rates are like for you.

CL: We have specialists for breastfeeding, so they advise mums, which would like to give breast milk of these special feeding systems and often we combine breastfeeding with tube feeding because nearly no infant is able to manage all the amount which would be necessary, so it works, only in some cases. We also have mums who take the breast milk and feed this by bottle to have all these positive effects of breast milk for the baby, but this is very hard work and only a few will do this over a longer time.

Q: Yes, and I'm just wondering if you're also using tube feeding which is common and you said you're looking for 2/3 of the energy requirements of an equivalent healthy infant; a lot of my experience is that we need to give higher energy formulas and give them more energy so that they can transition from the tube to oral feeding. Is that your same experience?

CL: Yes, some infants grow with these 2/3 of energy amounts, but I agree with you. That is why we use these weight charts. The families are often afraid of (having an) obese child and that's why they often do not give the whole amount.

And it's hard for them to feed, so intensive a child, which doesn't cry, which doesn't show signals of hunger and so with this growth/weight chart we like to show them what is really necessary. If the infant's spontaneous motoric development is very rare, you have enough energy with 2/3 of the intake. Otherwise, if you do very intensive physiotherapy, very intensive Vojta gymnastics, then you're right, the infant needs more and then we add some fortifier.

Q: And one last question, are you using the WHO growth charts or PWS growth charts?

CL: Yes, we use regular growth charts because when we combine our patients with the PWS growth charts they always have better height and a lower weight. So we see nothing in detail. So we explain to the parents we use regular growth charts because our aim is (on) these regular developmental charts and we're happy if the child reaches the 3rd to the 10th percentile of weight in the first year.

And that is why we use the regular one and not the Prader-Willi one.

Q: We have the experience that in the very beginning, within the first year of life we with growth hormone treatment we have to increase the nutrition of our patients because if not their weight is very, very low. So do you think that if you begin their growth hormone treatment in the first months of life we should adapt to increase our treatment nutritional treatment for this patient?

CL: Yes, of course, if you have this situation, I would increase the energy intake because if you have more growth hormone you have more movement, maybe the energy expenditure is higher than in an infant which only lies in bed, for example So yes, I would increase the energy intake so that I can reach the 3rd to the 10th percentile and always keep in mind that growth and weight should be in the same range.

When do you start with growth hormone? At which age?

A: At the age of 6 months.

CL: Yes, that's the same in our country.

SB: Well, I just think it's so important to stress that the weight should not be as you expect for normal children because the body fat in small children with Prader-Willi is bigger than in other children and people often think that the weight should be in the normal range and (as Constanze can tell you) if the weight is a normal range then there's very, very much fat on the body.

CL: Yes, I agree completely with you. That's why we try to be between the 3rd and 10th percentile and not the 15th, because otherwise we have high amount of body fat but not more muscles.

We mostly have the problem in the first year of life that we have underweight children because the parents are afraid to give food to the child, they are afraid of obesity, and we see that this lack of energy is not good for the psychomotor development and we see very small head circumferences and all these things stop. And then I also have no positive effect of growth hormone if I have no energy in the car.

SB: This is right and we see these cases in Denmark, too. But I think in many countries if the dietitian is not aware of the body composition of the child with Prader-Willi, then you see, I have seen many babies being quite overweight because they're tube fed with very much energy so the dietitian should really know about Prader-Willi and the body composition in small children.

CL: I think tube feeding is a chance for the family to have time with the baby, to interact with the baby and not only sit with a bottle. So I think in this case tube feeding can be helpful. The parents can change who is feeding the infant during the night, the other one gets enough sleep for the next day, so I think it's helpful and sometimes if the infant is getting (?) you will not have to go to the hospital the next day because you have a chance to manage the food at home.

SB: Yes, I completely agree. Just like you do not put too much calories in the tube.

CL: Yes, right. Of course at first, feeding and active drinking, and then only add what is necessary.

Q: Could you tell us where to find more information about the Castillo Morales feeding approach please?

CL: I think in Germany these Castillo Morales techniques are provided by the physiotherapists. There is also a booklet about Castillo Morales therapy in infant in hypertone infants, and also some speech therapist also can offer Castillo Morales.

About stimulating all these muscles, it's not that you have to go there every week. Often you have consulting and then you get an idea how to stimulate muscles more, give pressure to the muscles, how to have a good feeding position and then you have the next meeting with the Physiotherapist, three or four weeks later, it depends on how things go.

Q: You mentioned that you noticed a restriction of fatty acids in your group at the young age. I've certainly noticed a restriction of carbohydrate intake for parents restricting carbohydrates and there's a tendency towards a very modified ketogenic diet as the infants start to wean and in early toddlerhood. I just wonder if that's your experience also?

A: Yes, in the case of fatty acids, we recommend using the right amount of fatty acids. Infants need 40 to 45 percentage of their energy intake from unsaturated fatty acids. On the other hand side, we ask the parents to avoid sugar. That means no juice. We even call it baby juice, baby speeds or baby cookies, because we like the baby to get the good carbohydrates and also like the idea of conditioning the taste, so not to give over-sweetened mashed cereals.

Challenge presentation (Abridged)

GL: Susanne Blichfeldt, who I'm sure many of you know, is going to put forward a challenge on the management strategies to support the needs of the family of the child with PWS. Susanne is a paediatrician who specialises in Neuropediatrics. She's worked for many years with PWS and children with other developmental and neurological problems. She was involved in the growth hormone research in Denmark with children with PWS and is a co-organiser of programmes for parents at many IPWSO conferences. I don't think you could have an IWPSO conference without Susanne since 1991. She's a medical advisor and leader of the Danish PWS Advisory Board and a member of the IWPSO Clinical and Scientific Advisory Board and Famcare group. Suzanne also has a son with PWS who is 41 years of age and Susanne also extends her expertise to Greenland, several times in the year.

SB: Thank you so much for inviting me to do a case presentation. This is a very typical situation in many families that I want to present and perhaps you might think it's very simple and typical but it's a good thing for medical professionals to discuss.

A boy has 15 Q deletion. He was diagnosed when he was only 2 weeks old. He walked at age 2 and had growth hormone from age 1. His diet and weight is normal and he has blood tests every 4 months and for the hormones and everything is fine.

He was very ill once with severe gastroenteritis, but now is usually okay. His motor performance is OK, and his speech is a bit delayed. He recently started Kindergarten in a special needs school. He has a little sister. She's 3&1/2. Parents live together.

We call him Tim, in this case, and Tim is following the diet prescribed by the dietitian; there's no problems in the kindergarten or at home with food. The problem is when the family visits the father's family, either grandparents (or aunts and uncles). Tim has crying tantrums about food.

Aunt asks "can he have a small portion" (which Tim can overhear). Tim's father says no. Tim's mother is embarrassed that Tim is screaming and is tempted to give in and allow Tim cereal, which she has done on previous occasions as a result of Tim's behaviour.

Tim's mother and father discuss while Tim continues to cry and kicks his sister. Eventually the mother is very angry and wants to go home.

She tells me it is a situation that they have very often; when visiting certain relatives they always ask if Tim can have a small treat when Tim is able to overhear.

When visiting the mother's family, these problems do not arise; Tim is served only as requested by his mother.

Now the mother asks for psychological help.

What will you recommend for her?

[Discussion following Susanne Blichfeldt's presentation](#)

CL: I also think that this is a very typical situation. It seems like the other members of the family are not so familiar, or they have not accepted the Prader-Willi situation. It's always "only one cookie", or it's "only one ice cream", and for the parents this situation is very bad because they have to keep a hand on all these rules, every day, and they should also be the person who gives extra if it's possible. In this situation the parents are the bad guys and the other family members are the good guys.

Maybe they need, all together, a chance to discuss with an expert what is really helpful for the child. Often I think it's not possible to discuss it between parents and grandparents. They need someone else to sit at a table.

SB: I also think that the very bad situation for Tim here is that he is listening when they discuss, then he cries and then perhaps the mother gives in sometimes.

CL: He is learning the wrong thing because he's learning that if he cries louder and louder, he get what he likes to have. So of course he will do this.

Participant: I'm a psychiatrist in Ireland and I agree. I think it's sometimes the parents not being able to tell other family members or close friends. It is a very difficult thing for them to accept and be able to talk about. Food is such a basic thing I think most people struggle with. You know, even just normal kids, some families have more sweets when you go to a party so I think food is such a basic thing that it's quite complicated to negotiate every day all at the time.

It's a simple sort of behaviour strategy, "if I cry more, if I have more tantrums, I get what I want, I've learned that I just keep doing that and I get it". Now that's a very simplistic view but it does have a knock-on effect. I think it's being able to openly talk to family members, feel comfortable that your child has a specific disorder that needs specific supports.

CL: We try to keep in mind that it's not intentionally against the parents. (They want to see the child happy. But the way they go about this is) not good for the child. It's better to come up with things that are possible, rather than what is not possible. Write down *Our son is happy if he gets half of an Apple or some strawberries*, and not write down what is not allowed, because this list is so long.

This is a strategy for grandparents: to go and say "okay if we meet you, our son will be very happy if you buy some strawberries and not high caloric cake". To say what's allowed and what is possible in advance could be a strategy.

SB: 35 years ago my son with PWS was very prepared before we visited people and very often, he brought his own thing with him. Everybody was told that they could not serve him anything that we didn't agree on and we told people that it was like diabetes. At that time there was a great respect for diabetes. You wouldn't serve a child with diabetes anything that was not agreed with the parents so actually we did not have any discussions about food when we visited people because they knew.

I have visited many countries and often it's not respected. I really feel pity for these children because if everybody is prepared, you can avoid these situations.

Participant: Some of some of my families, some of my parents will bring a box of prepared items to, for example, grandparents with different options for the children and so maybe that could be that could be an option.

Participant: I'm a Clinical Geneticist in Malaysia. We deal with a lot of patients, children especially with PWS. We see the same problem here and sometimes it's worse because a lot of Malaysians live as extended family and can be the caretakers for the child with PWS. Despite getting them in during the clinical session, despite having them sit in with the dietitian sessions, we still have problems of educating parents. They feel that we're starving the child, that is the issue.

We had a Prader-Willi camp where extended family members were able to meet patients with (PWS complications) and see the seriousness of the problem. That camp

actually helped a few extended family members who were caring for the child during working hours.

GL: In Norway they have a similar situation where families with children of any disability can go and live-in for 2 weeks and learn all about the disability and I think it would be fabulous to have one in every country where families really learn what it's about. It's actually paid for by the government and they're there for 2 weeks. Their wages are paid while they learn everything about their child's disability. It works well.

Participant: We have a combined clinic sometimes with the dieticians. Malaysian families are quite close and a lot of the caregivers tend to be the grandparents or aunt or someone during the day where the parents go to work and so sometimes we actually invite these grandparents along to the clinic sessions so they are part of the management and on a day to day basis, get involved with the care as well.

Participant: From my view as a psychiatrist it's very important to underline what is possible and not underline what is not possible. This approach is important for the family members and it makes them feel more comfortable if they are not forced to forbid everything. Especially for the for the adults we treat here, it's important for them that they have for instance, a list of things that are allowed. It just changes the point of view.

Participant: I was going to bring up the adults. I mean, we see it all the time, in a number of different situations. It's not uncommon that there's marital breakdown and often one parent does the right thing and the other one doesn't. Grandparents some grandparents are better than the parents and vice versa. And then what's very difficult and very common is that the parents have a problem with their weight and they can't control their own weight, let alone their poor child.

In a group home, it's better, but if they are living with their parents not in a group home, they can have the initiative to go out and seek food and can be successful at it as we know.

TH: I wonder if I can raise a slightly different issue and that is, "Who takes the lead on helping with these problems?" we've heard from a geneticist who has been involved. Constance you're an endocrinologist. We've also heard from child psychiatrists. Who do you think should take the lead when there's a problem similar to what Suzanne has described? Do you think that is for the endocrinologist to try and manage? Is it for a dietitian?

There is, I think a danger, in supporting people with Prader-Willi syndrome that people fall between different disciplines and that no one takes up responsibility particularly when it's something that's quite difficult and you have to work quite intensely with the family. I'm just interested in your thoughts about who should take the lead, perhaps what discipline you think has the best skills here to manage this.

GL: I think really it's the responsibility who of whoever is seeing the client and their family and I think we have to not be afraid of what the family or caregivers think of us. If we know it's the best for the person with PWS we have to be their voice and their advocate.

Participant: I agree with what you're saying but I think the message also has to be consistent amongst the whole team. You can't have the dietitian saying "you can never do this" and then the endocrinologist saying "oh just once might be OK" because you've got to have absolutely consistent messaging. So it can't just really fall to one person.

SB: We have Prader-Willi centres where nurses have more time to talk with the families. But I think in a situation where a divorce (is imminent) in the family then perhaps a psychologist who knows both about Prader-Willi but also (conflict in extended families) should be involved too.

It's really important that families are not just told what food they should eat, but how to communicate the message of what Prader-Willi is, why is the calorie needs are so low. If people understand this they can respect it.

TH: To follow up some of those points, it does seem to me we are expecting the family to try and be consistent in their approach and of course, clinically, the person with Prader-Will syndrome may be seen by several different disciplines.

So it's also important for the *services* to have a consistent approach and those 2 things may be really quite difficult, to both get the families to be consistent, but also to make certain that the clinicians are talking to each other, and agreeing on an approach.

I think the other interesting point that was made is that failure of the family to be able to do that may be an indication of something else within the family and not simply an inability to do it. It's some struggle that's going on and therefore one might need to engage more fully with the family to address other issues than simply the issue of the eating..

Participant: That's reminded me of our situation. We had with a patient in our clinic who had joined a gym and got a personal trainer and the personal trainer did one of those body compositions scales and saw the muscle mass was so low and then said that the patient was eating not enough protein and gave the family, with all the best intentions, a very high energy diet full of protein to try and help the patient build up muscle mass. Even though we've done all this education, they came back because the personal trainer had said it was incorrect, so sometimes it's not just the people within your own service, you have to educate it's all these other health professionals.

Participant: You don't have to have a huge amount of intelligence to realise that the experts at the Prader-Willi clinic know better than some personal trainer. But there are some people who will cling onto whatever news they want to hear that fits in with life easier. I think the point about the whole family is really important. Understanding that

there's a lot going on, and to be a good therapist for the whole family is a very special skill.

Participant: I think it's very important to have a look at the whole family and some of my patients, I talk to them for 10 minutes when they come to my outpatient clinic and afterwards I have to talk to the parents for another hour.

It's so important to also understand also the needs of the families around our patients and to help them compensate and to help them just be good parents for their children, especially for the adult children. It's not easier when they grow up, for the parents.

Tony Holland Summary

The crucial message of Constanze's talk was the importance of a holistic approach and that there are different aspects to the care of the infant with Prader-Willi syndrome, not least the psychosocial, but the hormones, the nutritional, the motor development and so on.

As a psychiatrist and someone who works with adults rather than children, I was very struck by this point that you really have to follow the motor development because the motor development helps you think about the feeding and when they can be more independent.

The importance, therefore, of educating the family, of encouraging chewing because that also improves speaking and speech, the importance of physiotherapy, and I thought this point Constanze made, that parents get very little feedback from their infant initially, and therefore they need to be helped to understand and to recognise really small changes and see that as progress because that's a way of helping the family attach to their child and really feel that they are doing something that is of benefit to the child.

The issues of central thyroid insufficiency as well as adrenal insufficiency and then you talked specifically about growth hormone use, starting it from 6 months with an aim of reaching 95-98% of expected height. The importance of check-ups over time. The orthopaedic problems, to avoid passive sitting for example, and also you raised some other issues that are well known to all of us who work in the PWS field and are relevant across their life span; The high pain threshold, the problems of temperature regulation the fact that people with PWS rarely vomit, the excess daytime sleepiness.

I thought there was a really interesting issue too that came up in some of the questions and discussions about this tension between families being frightened of their child becoming obese so not wanting to feed them too much, but also then of not giving them enough or not giving them the right food, so that their development, muscle development, brain development and could be impeded.

So there's a real tension and a need for education for the families around that particular issue and that led very nicely into Susanne's presentation about that particular family and the tensions that existed in that family when they visited grandparents and other family members were involved. That brought out the need to try and involve the larger family whether it is through them coming to clinic or the example of actually going to a Prader-Willi camp.

I think there's always an interesting issue about parents of newborn children with PWS seeing other children with PWS and older children and particularly adults. When I first became involved in the field. Adults tended to be very, very obese and it was really very hard on children or parents of young children when they saw older children and saw that they were obese. At the same time that is perhaps a way of demonstrating to families, the importance of food management.

Then there was this discussion about how does one help the family? Who takes responsibility for helping the family and the challenges of that, so I've tried to pull out just a few of the points, but there were many more and each of you will have your own ideas that I hope have developed as a result of this, this discussion.

Upcoming Health ECHO sessions

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| Tue 20 th April | <i>Endocrinology</i> Charlotte Höybe, Endocrinology, Metabolism and Diabetology. |
| Tue 18 th May | <i>Understanding Hyperphagia</i> Tony Goldstone, Endocrinologist, Clinical Researcher. |
| Tue 15 th June | <i>Behaviour and Mental Health</i> Tony Holland, Clinical and Academic Psychiatrist, IPWSO President. |

Thank you very much to everyone who attended the session and participated. We look forward to seeing you on Session 3 in April.

Ends.