



IPWSO
International
Prader-Willi Syndrome
Organisation

IPWSO Caregivers' ECHO abstract summary

Sept 22nd, 2021: Transition

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: Norbert Hödebeck-Stuntebeck (NHS), Hubert Soyer (HS) and Patrice Carroll (PC)

Presenter: Neil Gumley (NG)

[Video link](#)

["Transition for People with Prader-Willi Syndrome"](#) Neil Gumley, Melbourne, Australia

PDF of the PowerPoint is available [here](#).

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

NHS: Thank you so much Neil for the information you gave and the brilliant experience and examples you describe. It will be interesting to see how it works in other countries, and what the experience is out there, and if others have the same positive experience that you have. What I understood is that in childhood, it works very well because there is a good network around the paediatrician and all the people are working together. Do the other participants today have same situation, the same positive examples in childhood, and then later we can discuss if it typically becomes more problematic in adulthood.

Comment: Yes, I can speak to my personal experience with my son and here in the United States. It is like Neil described, a very good community when children are younger, there are so many specialists, available and knowledgeable providers, but then what I see in the States, is that it is difficult to find knowledgeable providers and caregivers in adulthood. Moving from adolescence to adulthood, it is very difficult to find providers, therapists, doctors, homes and schools that really understand Prader Willi syndrome and are able to support that. While we do have some transition services and planning, we don't have as much for the adults as we do for the young children.

Comment: Denmark is a very small country we are only 5.5 billion people and we have 140 persons with Prader-Willi and more than half of them are older than 18 years. The case is that we have two centers for Prader-Willi, and the children continue, when they are adults to go to the same doctors. So there's no transition. Well, there is a transition for endocrinology, but still they come to the same pediatrician once a year when they

are adults, and they are in the same clinic, and they have the same dietitian, as they had as children. On the other hand, I should say, I am not sure that we have so much physical therapy and occupational therapy for the younger ones and all this is not in the hospital but that's out in the community so it can differ. But contact with medical staff and dietitian and orthopedics and so on, that will be the same doctor when you are a child as when you are an adult, because they go to the same clinic. We have two clinics, one in the western part and one in the eastern part of Denmark.

NHS: What you describe is a model that they do not have to change, and maybe they have the chance to go on with the same care. Some of you know Constanze Lämmer, (Germany) she, does things in the same way. She's a pediatrician but she tries to work with people over 20, (25-30 at latest), but then they, they have to change.

Comment: The eldest one who is going to the pediatrician (in Denmark) is 70. We have more in their 50s, so they come to the same clinic.

NHS: So a model could be to have no transition. And the other thing you said is to have centers of reference, or competence centers where people can inform themselves, and have help for the transition process.

Comment: We have a Prader-Willi clinic it's in Sydney at the RPA and our boys have gone through the whole time with this same clinic. We've been able to use that one clinic right through the whole time with the guys that I've been working with. That keeps it really stable with them when they go into visitor and everything's checked. That's in Sydney, we've got the one clinic with it. They've been going through right since child if you go into you'll see little ones right up to adults.

NG: And you get better outcomes with the same group and that transition. This is one of our key areas, whether it can be practical or not. I know we tried to get a clinic in Melbourne, about five or six years ago it didn't get off the ground, but it's important to have that stable transition. If you can get that model. Stick with it because it's going to help with the transition. But a lot of the times, we have to move from pediatrician to adult endocrinologist, and it can be a daunting experience. I guess if we could have one thing in Australia that would be fantastic would be a clinic that can look after multidisciplinary from birth all the way up, and then have an adult part of it where they can integrate and understand all those needs. That would be it, in a perfect world, which you do have, but unfortunately it's not like that all over the world.

NHS: So it's like a little bit the same as what was said to have these clinics or how these reference centers so that they can support the transition, because they know the person.

Comment: In Switzerland it's nearly the same as in Denmark like Susan told us and we have two specialist doctors and all the people with PWS go to these doctors and then they stay there, so long as they like. It's possible to stay into adulthood as well. The limit is only the age of doctors.

Q: I have a question, do you find differences in knowledge? In the United States for paediatric specialty care, they focus on just that snippet of time and adolescence and then you transitioned to adults. Is there a difference in knowledge base or focus for physicians that one system would be better than the other?

Comment: I can add that in the clinic we have in here in Denmark it's not for certain that you will always see the same doctor, but it's the same rooms, the same nurses there saying hello, you have the same dietitian, but sometimes it will be another doctor if one is on holidays. Also, especially about endocrinology, the adults in one of the clinics, they have another endocrinologist – an adult endocrinologist. But in the other clinic they changed the endocrinologist but they see the patient together so they see the endocrinologist, and also the paediatrician that they know for many years, and perhaps a dietitian, and a nurse who organized all visits. So they come to the same source to say “hospital home”.

And I don't think there's any difference between the knowledge of the doctors and actually we have a professional board in our association and we meet two times a year, and discuss, so I think it's more or less the same treatment that they are offered. But again, in our hospitals, we do not have physiotherapist, and occupational therapist that's out in the community where they live.

They can have stress because if they live far away from the center and if they have another condition like asthma or something that the GP could take care of, they do not go to the center there they go there for the endocrinology and for their specific Prader-Willi problems.

Comment: (via email) I just want to shortly describe the French situation.

France has set-up since 2004 a network of "competence and référence centers", led by Pr. Maithé Tauber and the situation is rather good for young children since birth. Diagnosis is made in the first month of life and most families are followed by medical multidisciplinary teams, with a very good expertise and knowledge of the PWS. There is of course geographical and social inequalities. A transition process is organized, with overlapping but there is a lack of resources in the adult sector, especially in the competence centers. Adult endocrinologists are generally available, but there are few psychiatrists.

There are also other reasons than lack of resources for this gap in the transition period. Families are more focused on the future of their child: residential problems, employment, autonomy and facing crises and rebellions of their child, than on medical aspects.

During this period, people with PWS move from a support mainly medical to a support by caregivers and preparing this transition is not obvious due to profession organisation (they are not prepared to work together) and cultural aspects. The need of a global vision is often lacking and essential anticipation insufficient.

Neil's présentation was really rich and highlighted the most important issues.

I just want to add a comment on the essential work to be done with families. It is very difficult to accept that our child is becoming an adult and will live his own life away from family home. Preparing the family and the child to an independent social life, but with needed rules and limits must be done early enough.

I also would like to point out that understanding specific aspects of this transition associated to hormonal, physical and social transformation is really lacking. Affective and sexual life, building a positive self-representation, etc seems to be important.

Q: I was curious about how people manage the transition from home to adult services with regards to technology, so movies that they're able to watch, internet access. So when they're home they're really well supervised on the internet, as far as what they can watch on TV and movies, then they go to adult services and they have really full access to the internet, sometimes have a cell phone, can watch whatever they want to watch, and how do people manage that transition because for me that's a really hard one, to go from full supervision to a lot of independence around that?

NG: That's a really good question, and it's a difficult one. They are going from a world where parents have protected them, enveloped them, and they've actually been protected from the world. They might not be ready for things like that. Having a phone now and being able to Google.

I've got someone right now who is the greatest "Dr Google" person I've ever met. He's got every affliction in the world. He will go, "Oh my, I've got cancer, I've got this, I've got that." We're always going to the hospital.

And, and it is a tricky one, but one thing in Australia is we're governed under a framework. It's a professional board which is made up by the government and we work with them about autonomy and the decision makings that they have. One thing is key and that's education. We look at things that happen, and we look at an incident that might have happened. We look at if we have been able to educate them to a place where they can make a better informed decision, not straightaway, when they first get in the world, but that they can make an informed decision.

In our group home we have resident meetings. One day we brought in a police officer, because we thought, let's get them used to this idea because we know people with PWS have the ability to get into fights sometimes and they can cause a ruckus in the group - we've had police call outs from time to time. So why not get them used to the police and what their role is?

We have a group in Victoria called Family Planning Australia which talks about everything that you might not want to hear, but they start from language that might be easier for them to understand. They will have presentations on sexuality. All these different experts. It's continued education. I feel that we try to arm them over time with more education and understanding. People with PWS you can't stop them from

having internet access, they're going to find a way of getting it. So why not try and give them that little bit of autonomy and try and work with them, so they can access it in a safe environment in a safe way? It takes a little bit more time, but we've had some good outcomes from it.

And we've had some we've had some not so good outcomes but they do come back and say, "Hey, I remember that talk that we had with the police officer, and I know the next time if I do this, they're going to come and I might not be staying here." Just little things like that. They remember those things. The first time we had the police officer at the house they talked about the police dogs for an hour, so it takes time and education to me is the key.

NHS: How can we prepare for the new environment and how do we do this in the different countries? How do we support people? One of the things Neil said was training before starting.

HS: I think we look at how can we transfer from a dialogue with a child to a dialogue with a young adult, I think that's really a big question. Often adult people with Prader-Willi syndrome are treated as children. And that's due to their cognitive impairment sometimes, but they are adult people. And so therefore we have to find new ways how to communicate. It's necessary to find a dialogue on the same level, and not the dialogue from an adult to a child.

We have to start very early in education to give them small experiences they can deal with and not too much at one time. When the transition comes from an over protected family to a group home, and they have to make a lot of decisions for themselves, it's too much. I think we have to prepare them.

NHS: To have no cell phone and then suddenly have a cell phone for free use as much as you want is not being well prepared. We have to prepare the new environment, about the way how to communicate with these adult people. As Neil said in his presentation, often adult medical doctors have no experience communicating with people with cognitive impairments, and this is what we have to prepare in the way of communication in the new environment.

Comment: We have sometimes the opposite problem in that they're coming from a family home that is a free for all, there is no structure, and then they are thrust into a world of social care of group home that they're now thinking, "Whoa, I can't watch this, I can't play on my iPad 24 hours a day. There are rules."

I feel that a lot of times, at least for group homes, people enter, because there's a crisis, things are not working in the family home. There's not smooth transition planning because it's a crisis. So, to start prepping the family and the person with visits and realistic pictures, because they often have very fixed ideas of what my new house is going to be like. Again, education for everyone.

NG: I say this a lot to my staff when we have a crisis in the group home - there's a reason why they're there. Don't expect that everything's going to be going swimmingly. There's a reason why they've left their family home and they're with us, or there's a reason why they left their own place, and they're with us now.

This is why I said at the start, if you do a very simple literature search on transition in a social environment, in a group home environment, there's a couple of really good articles on some medical transition, and it's based all around having that connection between paediatric world into an adult world.

We're caregivers in a medical world. We're at the coalface, we're the ones dealing with the actual social changes, we're living with every day. Hopefully we can develop some more ideas around transition so when children are 8-11 and a family starts thinking about future steps we can advise. What's the problem with starting them on the internet early on so they're getting that socialization they're getting that taste of what it's like they're not dropped in an unknown world and they feel they can't fit in. Our job is to support them to find that place in life where they feel that they belong and they fit in.

We can't do that when their mind's racing, and saying hey, I want to do this I want to do that. I have this freedom now, or all of a sudden my freedom is being taken away. So how do we stop that? If we have some good literature on this, it's going to help parents when their children are young, these are the things you need to think about going forward, things that we have to start thinking about early. You might have a wonderful child, you might have a great experience till they're 20, 30, 40, that would be great, but unfortunately that's not the way it is. So, if we pre plan, it's going to help us.

NHS: So, one of the key things is to work closely with the family, and another thing is if there is a chance, work early with a family, so that we understand much more what the needs are.

Q: What do people do when there isn't an agreement with the family and what the adult wants to do? So you have a family, who is very much against their adult child, either accessing the internet or being in a sexual relationship with someone. What do you do, what do people do about them?

HS: First of all, we are responsible for the young adult with Prader-Willi syndrome and not for their parents. I think that's the center of our work, but it's always better to find an agreement, and I think we need a lot of talks with parents with anyone who they'll listen to, on how to mediate this process of transition to adulthood. As has been said, we have to start as early as possible to come in contact and provide guidance for the families and for the children with Prader-Willi syndrome. What are their goals? How do they want to live in the future? They will not stay always in one unchanging state, it will change, that's the problem.

Comment: For typically developing children, when I look at my children, I don't plan my other kids lives out, I haven't put that much thought into this whole thing. But I think, as a parent with a special needs child, I never knew, I still don't know, I really needed to give a lot more thought about preparing him for these specific stages in life that my other kids just understood. So it's really starting that preparation, and giving it thought throughout their lives and teaching them safe internet use, and having those sex talks like some of us did with our other kids. But also, what are healthy relationships and being a lot more thoughtful about teaching them this whereas the other kids just have friends and they just do their thing.

Comment: It seems that there are different situations - crisis situation and they all of a sudden move from home into a group home because with special rules. We can prepare the parents to do more in the years leading up to moving. So, preparing is the best but we have to be aware of these crisis situations where young adults and their families are in a very different and very hectic situations.

NHS: You're absolutely right and we have to be prepared for both situations when we have time and when we have no time. But the best thing is when we have time.

Where countries which have models that are helpful for this, the case management, the center of reference where parents can go and get information, what should IPWSO support and bring out in the world as a good model for this, and a helpful model for the situation when we have time, or a crisis situation, and there must be a switch from one place to another place.

The situation should be more and more that we have time, and then we should offer options for parents to go to these places. How can we manage this (in areas without good models?) Maybe a paper in this and IPWSO should make the recommendations.

NG: I think early intervention is the key, and I think early education is the key. It could be through IPWSO or it could be locally organised. When a child is born with PWS, the first thing parents will do is go to the internet and type in Prader-Willi and then they might look at the country they're in, and then that will give them a link to either their local states or area with information. And I think the lifecycle of someone with PWS is described a lot around food intake and all the medical needs going forward. There's not information about the transition and what's going to look like for an adult.

So if we're preparing that information early for parents then, part of that life cycle for them can be that option to start that transition and work through those steps, like you said, early intervention and early information would be key. It's another thing on top of a lot that they have to deal with, but they do we need to start planning what will life look like at 50 or 60? Where's my child going to be? How can I best prepare my child, so when I'm in a different space they're going to be in a social world where they're feeling safe, and they're feeling comfortable in their environment?

With all the things that are going on from birth, it's something that gets overlooked. But I think one reason why it gets overlooked is because there's no information out there. So we should potentially develop a paper, something for people to look at to give them another point of reference going forward.

Comment: I like the idea of a center somewhere a global center, or a center of excellence, as we call them here, because there's so much transition between providers and caregivers that consistency no matter what it is the is the key. Just this one house where you will have consistency, the house, the message stays the same, the people come and go. The message is the same.

HS: For this house, it's really necessary that they are always are willing to try new things, to have more research. As we learn we have to apply the things we learned. I think that's necessary, so this house, were it consistent over time with the same knowledge, that's not good. We have to develop our knowledge as we do in these discussions, and to bring it to the public and make it available for everyone over all over the world.

Q: You mentioned Neil that you brought Family Planning Victoria in to talk to people in the house. Did they come in just the one time or have they come in a number of times? My question really is around whether a single visit is enough for them to learn new skills or new ideas or whether in fact, an organization like IPWSO – we do a lot of thinking about how to train support workers. I wonder if we should be developing more programs for repetitious training of people with Prader-Willi syndrome - that ongoing education.

NG: We had them come in every time we've had someone come into the house, and it had to be several times because obviously, the learning capability. The first time is always the most difficult time; there are questions and answers. We needed that repetition. Yes, getting people in over time is really important, and them actually having an understanding of Prader-Willi is key because then you can communicate in the language that you need to tailor it to their way of learning, which is the most important to me, and having them come in, even if there isn't an issue, I think that's when it's best learned, not when we're having a crisis, when things are good.

Comment: I think that it is about training, it's about giving information. But I think it's also about challenges in life, challenges that you face. So, going to school, forming friendships, sexuality, and everything like that, it's about the challenges that you're just going to face in life. They are different, they are country specific, and even within the country they're going to be specific to different regions so an overall guide, of all the problems, the challenges that you can face, and then how to overcome them.

We've had problems with adults and the parents of the parents of young adults, and they've had a free for all (few restrictions at home) and then they go to the residential service just like you mentioned before, but if then guides were available to people before about what you might face going into a residential service or an independent

home and looking at all those challenges that people can face and giving information I think that would be a really good way to go forward.

NHS: As you said maybe there are centers and expert centers, but they have to have knowledge about the local specialties so then they can give better answers.

Comment: I think the take home message I've had from it all is that, possibly because PWS is such a relatively rare syndrome it's really a constant challenge to get adequate services, spread out far enough for everybody to access and even though I'm in Newcastle which is only two hours from Sydney where they have the wonderful Prader-Willi clinic, there aren't really a great number of specialists in all fields that specialize in Prader-Willi syndrome in my city. So, people have to travel to Sydney, and they don't do that all the time they just do it intermittently. So it's a real challenge.

As a psychologist I actually found that when I started to take on a couple of Prader-Willi clients that I actually spent a real lot of time educating myself via all of the wonderful resources out there on the internet and reading research papers, and going to a few talks in Sydney, Janice foster talked in Sydney, so I traveled to listen to her which was fabulous. But on the internet is a fabulous resource for building one's skills and getting up to speed on what we all need to know. So that's been terrific. so anything that you put on the internet, I feel pretty confident that people will seek it out and find it.

Patrice Carroll Summary

PC: This is no small task, we had a lot of conversation. Neil It was amazing, thank you. I will try to summarize as best I can.

Our topic was transitioning from child to adult services. This is a really important point - we go from, (in childhood) a large team with a lot of experience to (as an adult) a smaller team that's new to that person may not have the same amount of experience, and then we have to, as families and individuals caring for people with PWS have to rebuild that relationship with medical staff, and in most cases, train the medical staff.

We should start transitioning medical and health staff early so they can overlap with the children's team, using case managers and local PWS associations. As far as day programs and work, there's a need for staff training, because now it's a safety issue. It goes from being very important to vitally important because, as Neil used the example of working at McDonald's, that could be a major safety issue. So now the training goes from important to vital.

Be mindful about housing and the location, the mix of people, understand the person with PWS specifically and that they're on a spectrum so not all needs will be equal. Allow for a gradual move into the house, allowing for physical space, and routine from day one.

Managing expectations of the family is crucial to the social transition, developing relationships and friendships and new relationships with the families.

What helps: having role models, having early friendships, experimenting with independence in a safe way, learning the person's communication style and staying connected with support systems and to keep learning every day as much as you possibly can.

There was a discussion regarding continued care, education regarding the internet use, use of community resources and how do we prepare for increased independence and access, working with the families around their expectations, working with the individual around their particular goals

Being aware of how we're speaking to the individuals with PWS; if they're adults speak to them like they're adults. Meeting to prepare the person and the family for new structure and new freedoms is vital. The need to prepare parents as well.

Finally we ended on the topic of whether or not IPWSO take on this topic of transition, and I think there was a consensus that we should, bearing in mind that we definitely need more research and information regarding this topic before we can become a center of excellence.

NHS: Thank you so much, and the last point you made, as Neil said at the end of his presentation. this could be the start of a discussion about it and we will go on with it in Ireland and today is a very good basic for these.

Upcoming Caregivers' ECHO sessions

Weds 20 Oct

Young and clever: well educated people with PWS

Norbert Hödebeck-Stuntebeck, IPWSO PPCB Chair, Psychologist, Germany and Larry Genstil, Psychologist, Israel

Thank you very much to everyone who attended the session and participated. We look forward to seeing you on Session 5 in October.