IPWSO Caregivers’ ECHO abstract summary May 18th, 2022: The role of Diagnostic in supporting people with Prader-Willi-Syndrom

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

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Presenter: Norbert Hödebeck-Stuntebeck (NHS)

Video link

“The role of Diagnostic in supporting people with Prader-Willi-Syndrom” Norbert Hödebeck-Stuntebeck, PhD, PWS-InterNational, Germany

PDF of the PowerPoint is available here.

Q&A Following Norbert’s Presentation

HS: I don't know if, during this presentation, you have made your own cognitive, emotional and social profile, like I did during the presentation. But I have found that in some areas I would be not successful without the support of my friends or my wife. I think there a lot of things we found out with diagnostics, as Norbert already said. I remember for my PhD it was 60 people with PWS, on a neuro psychological test battery, and the WAIS is a part of it, and it's really helpful. That's my opinion, but now the discussion, what do you think about diagnostic tools and do you use diagnostic tools already in a systematic way in your services?

Comment: I think it's really important because it gives you the opportunity to look at, what's happening before an incident, what's happening in the middle of it, and then the reflection piece at the end is so, so important as well. Because I think it's the reflection piece at the end that will enable the team to support that person to future instances that are similar.

Comment: We use some diagnostic tools, I think a little bit different than what was presented today but definitely it's important I think that we utilize what we can to find out the best… but a little bit different than what was presented today but certainly using similar tools.

Comment: Yes, I really fully agree with what presented here, just wanted to add some comments. In French, we call that functional evaluation, which is really important and it’s a pity that it's very often missing. So, caregivers have to deal with people and they have no functional evaluation.

And the second point of view, in the training we often use a term challenging situations, because you know, it is not only a problem of the person and the behaviour, but it's also the situation which induces challenging behaviour and we see both with this ecological model in mind.

In your draft, Norbert, there is also one thing we discovered in several situations. This is spatial sensitive perception, and I just give an example of a young boy, who was presenting really high challenging behaviour, we finally through a very exhaustive evaluation found that he was sensitive to spatial audio. When we solve a problem, everything was okay after that. All those of carers and PWS specificity are not enough taken into account.

And so concerning john example I think that also we have to work on the theory of mind, because this is probably one very important thing, and I saw that in some papers that you can work to improve the theory of mind at any age, and this is very important to prevent situations.

HS: I think you summarized all the most important things about diagnostic. And, I think the examples Norbert gives us to make to make us sensitive for how important diagnostic is. I see written in the chat, “Thank you much it's so essential - more staff less persons together - but who can pay for this?” That's always a question, I agree of course.

NHS: Maybe I can answer to the last thing of who will pay the stuff. We cannot change the environment after a diagnostic but for the staff, it was helpful to know every time when we are less than three people in in the group home then we can have more problems, or if we see more than four or five people with PWS are together, then we know it could be that there is a critical situation. We cannot give them more staff but we could give them the information to be a little bit more aware of, be more preventative, when there are four or five or six together so this was the information we can use. We cannot always give more staff but I thought it gives them information to handle situations and be more preventative.

PC: Because you can also change programming. So you can have the person do something a little more independently, and not doing a group project with all six people. So you may not need to add staff, you can just be a little more creative about where you put people in the environment.

Comment: What I'm finding with the NDIS in Australia is the more evidence we can collect around behaviours, exactly on along the lines of what you're saying here, the improved likelihood of getting additional funding. So without collecting the evidence and understanding these tests, if you like, or understanding the ratios and the staff numbers, then you have no hope of getting extra funding. So, I hear what you're saying about who's going to pay for it, but if we don't collect the evidence and understand that, then we will have no hope of getting extra funding.

HS: You're right. We have two different sides, the one side is the financial side, and you have you have to collect a lot of arguments to get better financial support, of course, but also for the caregivers, I think it's necessary to have more information and therefore they can develop a better strategies for coping, the situation, together with with the people with PWS.

Comment: When I wrote this, who would pay, because I'm thinking about so many different systems in different countries, who is paying, and how many countries do have homes for PWS, and for example in our country it's the community who decide how much money they will pay for each person living in a PWS home. So, it is really necessary, Norbert's documentation here is so essential to test them and to know what they really understand. But I think the biggest hurdle perhaps is to persuade the community who pays. We really need so much documentation.

And you for the presentation Norbert because I always think, "What does he or she really understand", that is the first question to say to yourself.

NHS: Especially the last thing I present, these behaviour observation instrument or something like this. This costs nothing. This is only something what the team could do by themselves. They only listed, when was the conflict, who was there. And then at the end they make an evaluation but it costs no extra staff.

Comment: I know that, but when for example in the Danish group home, if staff, make all these observations and they go back to their boss or the community and say we need to have more staff, and less persons together, and then the answer is No, we do not have money for that. Therefore, we have to fight and fight.

NHS: But what I want to say is the staff who is there, we should think about what can they use and we have to find out which helpful tools which have for diagnostic, simple diagnostic tools we can give them to be used by themselves and not at additional cost. This is my idea and therefore I hope we discuss this over the next years to find out, simple things which could be very helpful by the resources they have by themselves. Maybe at the end parents too.

Comment: And education of staff is so important. Thank you.

Comment: I agree with your comment, Norbert. I think that it's very important just to collect facts and to try to build a picture. What I find in many occasion, the caregivers they try to interpret or understand too early, before having the global picture, and this is something the kind of tools you presented, and this is something which is in the ABA method for example, for crisis. This is something which just forces people just to look at the global picture.

HS: I think one of the things should be how to interpret the results. It's not easy, and caregivers need help. They collect a lot of facts but who is helping them to brings together an overview to make a global picture of all the results, and not to look only at one fact, I think that's very necessary. Norbert and I developed an instrument, which we tried out over many years, only for observation of people, and it lasts only two or three minutes for each person, three times or two times a day. And over the time we see a lot of development in our interventions, in our support, to find if it is useful or not. And that was really helpful. We presented it already, a one times in the Munich conference. But it would be really helpful to collect data to have arguments to convince people to finance all the things that are necessary.

Comment: I think that's very true, if you can validate the tools and prove their worth, include data and the published articles, and then use in that sense then that will affect the funding in a circular economy in that sense. So I think it's well worth piece of work doing and on two fronts. One is that the sort of National Front about how to change things and policies and things like that but just on the caregivers themselves, to have that tool to be able to change that behaviour and to better manage a behaviour, that'd be good. I don't see an issue with it.

Comment: Are the staff dedicated and paid for things like this, or they do other things and they are then asked to spoke about things that happened and be proactive or something like that.

NHS: You think that this is the job of them. Correct?

Comment: Almost like a job. I mean, extra job. Because, to participate in a study, let's say, it's something they must do extra. Because one job is to take care of someone or do some chores or something like that, then to intervene in in a study or theory principles it's something different.

NHS: So, therefore, I think, to make clear that diagnostic could or should be a part of the job. I think it is necessary to develop something in IPWSO and to bring it into the world and to implement it in different concepts in different countries. If we really do this, maybe in three years that we have a paper, I think it is an option to put it in the negotiations to the governments that this is part of the job of the staff in taking care of people with PWS. I'm sure not in all countries, it will happens and it will be successful, but if this is a paper from IPWSO I think it could be a part of the concepts of taking care of people with PWS.

HS: I think it's really necessary to have some standards. And if you think on growth hormone therapy, it starts in one country and over time it becomes standard. And I think also the diagnostic should come as a standard in caring for people with PWS.

Comment: One point which is important for me, before challenging behaviour and challenging situation, there often are many weak signals. And what we insist on, when we do training for caregivers is to share, to collect these signals, and to share it with all of the team. Just to prevent possible crisis and this is the kind of tool you suggested have something just to say how can we correct at these signals?

Second comment I wanted to do is that, certain behaviour is something very hard to live with for caregivers, and you know if you just help them to prevent these challenging behaviours they will just take them with no problem. This is not extra job, this is something which makes their life easier.

HS: Yes, I think you're completely right and I also think we should start at an early point. Start with the parents together to support parents with their children in the families, also to have a look at the systematic observation of the week signs. And therefore, we can prevent crisis and help them to stay in the window of tolerance.

Comment: In my opinion, this last thing is true because we have zero experiences in group homes and with professional caregivers, so I think that for avoiding crisis we must search for and note the triggers and the phenomenon that accelerate a crisis. We as parents and the nearest persons to the patient, are more focused on the person, and can see the slightest differences between normal conduct and an outburst. So in the difference by a professional caregivers, I'm reticent because they might strain to share.

NHS: In my mind, the things said are the same - parents and carers have the same needs. And as far as I described, we have to see the signs and the signals to understand and this is the same by parents or caregivers.

And the second thing which is necessary for both is, we have to know what to do. So we have tools for prevention, this is what we have to support or to train the professional caregivers, but also the parents, they are in the same situation. Some are at home, some are in group homes, but the need for parents and caregivers is absolutely the same. And therefore maybe we can support these things and how to find it out to parents, educators, and caregivers.

HS: And I think that we have also to involve the person themselves to explain to them what we have found out, and if it's all right, and I think they are really interested about the things. If you make a test with someone, he's very interested afterwards about the results and to discuss the results, and also if you give him the chance, what you have observed in weak signals and to the to make a feedback, and I think it's so necessary, to involve the person, even if they are a child.

NHS: Maybe I can ask about the comment at the beginning. When you say you do it, but you do it in a different way. Can you describe a little bit what you are doing?

PC: I can answer that. So we have an online database that we use that incidents are tracked, actually just daily shift notes are on there as well, and parents can see it and whatnot. But on that online database we're able to create a forum that tracks, anything that you want to track, bowel movements, aggressions, skin picking, anything you want, and then anytime that happens, it's checked off and then you have a pretty good list of when it happened, why it happened, what was happening beforehand. And you could then look at that we haven't done this, but you could then look and compare like what staff was on what residents for home at the time so you could do all that with that database that we have.

NHS: So you make a documentation every day in the computer and then you can collect the information.

Comment: For certain students too, we can collect a specific behavior so if we're tracking something specific, each person that's working with the person can track that behavior and then you can pull it to see the antecedent, what led to the behavior, the behavior itself that did happen, and then the consequence from that behavior. Starting with who was working with a student, what the environment looks like, where the person was, in order to see if there's a pattern to go back and review it.

NHS: And do you do extra or specialized tests like some of them I described, or totally other things?

Comment: I don’t know if there are specific behavioral tests that we do, in regards to the diagnostic there are more academics and things like that we would do, but not exactly as you described.

NHS: And what do you think, do you think it is helpful to do individualized standard of tests in the future, not only for behavior, it could also be speech or motoric or physiological things.

Comment: Absolutely. I think the more you can test, the more you can identify where issues lie or where the work needs to be done that's very important for all the individuals.

NHS: What do the others think about this idea to work on a draft or a paper about diagnostic system and PWS over the next three years. Should it be a goal of IPWSO or of the caregivers’ board? Is it realistic?

HS: I think if we start now, in discussions in PPCB, or in other countries with it, I think we can collect some things which should be standard to make a diagnosis on people with PWS, on the emotional and social and cognitive and motoric part. I think in medical part, there are already some standards and I think health and wellbeing, which is discussed in the next conference will make a big step in this, and therefore I think it's necessary to, develop it also for caregivers and parents, which might be helpful for them to get a better view on some things which they have in daily life situations, and which can help them to make a support which is helpful to avoid a lot of things we have in crisis situation. And I will support you in any way to get up these standards.

Comment: Yes, I fully agree, of course, with this project. I just want to add that it's also important to think about when to do and what. Because you will have a lot of evaluation. This can be a very heavy and needing a lot of time but there are some periods where we know that we need some specific evaluation, and this is also part of the project for me to identify what to do and when.

HS: I think you're completely right. We have to look, if you think on children, it's necessary to have developmental diagnostics. But it's also necessary, as Norbert mentioned CO for emotional development refers all to these things you have already said.

Comment: There is a good dynamic vision. Evolution in time is very important.

Comment: I think that we can do a great deal in helping people develop ways of collecting behavioural statistics. The bigger task is developing simple diagnostic tests along the lines that Norbert's talking about. So that I think is the big body of work to work out a simple way, more likely an online way, to develop and support diagnostics.

HS: I agree with and I think it's also necessary not only to have the diagnostic data collected, but also interpreted. Who can make the interpretation of the results? So not only to have data, we have to interpret the data in a way that the diagnostic will help us to have different and essential science on it. As caregivers we have documentation systems to write down each, what happened every day and, but it's not systematic and it's not interpreted.

Comment: So what's happening in the case of my son, to bring it back to a personal thing. Each support worker is supposed to fill out an online form - they just click on a QR code on the wall and fill out the form before they leave this shift. And then that information automatically goes into a spreadsheet, and from the spreadsheet, we come up with pie charts and bar charts that reflect behaviors, but I doubt that we've got to the point where we consider those deficits that Norbert was talking about and I think that would be useful addition to something like that.

NHS: And as I understand correct, not diagnostic only for negative things and for critical things. The most important is to find out where they are competent and start to develop with these things, absolutely right. But I will, I read the comment in the chat, *"I think that some country, some centers can start and implement and by time, others can admire and copy the model, but I don't even hope that all countries can implement programs with such quality norms."* Do you think that IPWSO could be the institution to start something like this and all countries or countries can use these information which are there? Do you think that episode can be the motor of this?

Comment: I think that it would be a useful project to try and seek a grant. I'm not sure who would be the most likely grant maker for a project like this, we often find that the whole area of intellectual disability is a bit underfunded. But it's obviously going to take time from support staff and volunteers to participate. But yes, I do think that IPWSO would be a good place to run it.

Comment: I need to know if there similarities between countries at all levels, because for example for hormone administration, this is an issue, because we have a program, a national program, but doctors they are only interpreters. We don't have a guideline for all terms of care. So if IPWSO can create some, I don't know, big document with many guidelines in every common situation, maybe we can show with all the person, which are involved so we can have an example of all guidelines for all in common for people with PWS.

NHS: And if IPWSO signs off on this, or the name of IPWSO is on it, you think it's easier to bring it to people, to bring into government, to bring to staff.

Comment: Yes, because if there are any other institution or officials that can assume their own position to make some of those guidelines. Maybe IPWSO can take the standards and create some something physical that we can show. Because doctors now are gaining experience but they are not so experienced. Also caregivers, also parents. Also if you think there are many undiagnosed, many without treatments or care at the minimum levels. So you're seeing the big picture of this. We all must bring all countries, all centers to the minimum standard.

NHS: So that many countries can use this. And you make the task a little bit bigger, you said we should not only collect things, and test, we also should describe for what it is good, and examples to use it.

Comment: Yes because you don't do nothing with the data as information if you don't give examples on at least one direction.

NHS: And this is what I tried to do today, not only to say there are different tests. So my idea was to give an example, how can we use, and how can this test be useful for the all day care.

Comment: Because anyone can do a little something to correct a situation or to prevent or to even see it as a crisis, or to notify somebody who was in charge, or who can take measures to correct the situation. But everyone must see the same things that can be corrected or taken care of. Because our job, all of us, is to make a better life for them so every aspect it counts.

HS: I think you're right it's really necessary every aspect counts to enhance the quality of life for people with PWS, and therefore it's necessary to have a lot of founded information and to make all who are dealing with the person to make aware this is the basis, we have to orientate for our support.

Patrice Carroll Summary

PC: We talked about the role of diagnostics in supporting people with PWS, with the premise that using diagnostics can improve care and improve quality of life.

We looked at the course of a crisis, the prevention, the crisis itself, post crisis and reflection. Looked at the level of arousal and the level of cognitive competence and high arousal equal to decrease in the level of cognitive competence. Understanding - to know as much as possible about the symptoms of the syndrome, to look at the environment, the bio psychosocial background diagnostic of the person with PWS, and examples of why diagnostics is helpful.

Understanding the behavior, to look at an IQ test but not just the number - to look at the sub tests, and that will give us a clear picture of what can cause the problems, to provide greater and more accurate support if we understand the competencies.

We saw examples of how interventions were more successful when there was a greater knowledge of competencies. Information can help us with daily schedules such as having the person work when the focus is stronger.

Documenting challenging behaviors can help pinpoint when incidents occur, and if there's a correlation between the incident and the either the staff who is working, or the number of staff who are working, or the number of clients present. Information gathered allowed for an opportunity to change the environment and ultimately decrease behaviors.

And then we went on to the discussion, which is what do we do now that we have the information? We can create an environment that doesn't antagonize deficits. When staff have a better understanding it can help them work more effectively. Individuals can learn a lot about themselves. The more information and documentation we have, the closer we are to be in a position to request increased funding. And ultimately, it gives staff and individuals a chance to learn and grow.

And in the States where we would say is, “When you know better, you do better.”

This was the final planned Caregivers’ ECHO for the 2021/22 programme. Please visit https://ipwso.org/how-we-can-help/project-echo/the-ipwso-caregivers-echo/ipwso-caregivers-echo-resources/ to view previous presentations. As new ECHO Programmes are announced we will share information via the IPWSO Newsletter. Thank you to everyone who participated.

Ends.