



The Proceedings of the 2008 and 2009  
International PWS Caregivers' Conferences:

## Best Practice Guidelines For Standard of Care in PWS

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## Forward

When Pam became president of IPWSO, she set out to visit all the places in the world that serve the needs of children and adults with PWS. She met with parents all over the world, listening to their problems and celebrating their successes. She discovered that because of early diagnosis, good food management and exercise, children with PWS were growing up and surviving into adulthood, creating a new expanding population of individuals requiring special services. She toured many residential programs serving individuals with PWS, meeting with professionals, providers and caregivers. From these experiences she could see that these programs had common features as well as strengths and weaknesses. She also realized that there was no network through which these programs could interact to share their experiences. Nor was there a method for older, more experienced programs to mentor newer, start-up programs. She was also aware that group home living was an especially important transition for young adults and their families. As the parent of a young adult with PWS, she knew the importance of staff training, food security and good behavior management to assure optimal level of functioning. She appreciated the need for exercise, leisure activities and social opportunities as well as sensory integration experiences such as dance, hippotherapy and asinotherapy. She also recognized that people with PWS should have opportunities for work and productivity to develop skills and enhance self esteem.

It was Pam's mission to share information about PWS with parents and professionals around the world. She was dedicated to establishing a "conference home" for caregivers around the world to share their experiences and to derive support from others who understand the stress as well as the rewards of working with individuals who have PWS. She envisioned that the outcome of such a conference would be: ***to develop a "best practices" model of care based upon caregiver/provider experience around the world.***

It is interesting to note that in the planning for the first Herne conference, the title was originally to be the First International PWS Caretaker's Conference. In the process of planning the content for this event, it became evident that the role of the caretaker was not static, but dynamic, ever changing to meet the needs of the person with PWS. The process of caring for the needs of a person with PWS was not *reactive*, i.e., looking after, but *proactive* and *interactive*. Subsequently, the title was changed to reflect the unique attributes of the role. Thus, the title of this conference was changed to the **First International PWS Caregiver's Conference**.

Pam said that most conferences offered a SMORGASBORD of ideas...but in the caregiver's conference, the attendees would work together with a leadership team to *prepare* the SMORGASBORD! The attendees would bring the experience (the food); the conference organizers would have pre-set the topics (the menu); the

experts would present selected “methods” (sample recipes are demonstrated); and the work group would decide the **Best Practice Guidelines** (the recipes for success)!

The Work Group consisted of a select number of attendees (caregivers and providers) plus the Leadership Team that included Presenter(s), Facilitator, and Scribe. The Presenter(s) provided an overview or different points of view on selected topics. The Facilitator stimulated discussion by soliciting or asking questions, focused the discussion by keeping the group process on track, facilitated consensus building, and summarized the discussion. The summary identified the key points endorsed by consensus: on what items do we agree mostly; what are the areas of greatest disparity and the reasons for these, if possible; identify problems or limitations and brainstorm potential solutions: what don't we know and what additional information is required. The Scribe took detailed notes of the discussion and developed the first draft of the written report.

The tasks of the Work Group were:

- To discuss pertinent issues related to the selected topics;
- To build a group consensus about those issues, i.e., what we can agree upon;
- To develop the **Best Practice Guidelines** related to the selected topics.

It was the task of the leadership group to synthesize the work and to present the guidelines to the entire conference as a poster in the **Market Place of Ideas**. The poster was presented orally and provided an opportunity to share ideas informally. A written product of this body of work was planned, and it was anticipated that the collected works would be published as conference proceedings.

There were two sets of 5 work groups who meet over the 3 days of the conference, and there were two sets of work group posters presented to the conference-at-large in the **Market Place of Ideas**. The format for the posters was as follows:

Poster Template:

- Title/Topic
- Abstracts of presenters
- Define the nature of the problem and why it is important
- Describe the impact of the problem on the following:
  - the person
  - the family
  - the caregiver
  - the provider (structure and organization of care)
  - the system of care (medical, psychological therapeutics, education, legal)
- Guidelines for best practice
- Discussion
  - Unanswered questions

- Issues outside consensus
- Challenges for future consideration

The process and content of the 2008 Herne Conference was evaluated. The attendees were asked to describe the nature of their work with PWS, and as expected, most of them were direct care providers. There were also “leaders” of group homes (team leaders, providers and administrators); medical, educational and psychological professionals working with PWS; parents and others involved in the care of persons with PWS. The structure of the conference was very favorably evaluated. The quantity of information provided and its relevance to day-to-day practice was very high. The competence and motivation of presenters and group leaders was rated as high. Of the 72 respondents, 68% indicated that the conference should be given again.

The planning for the first Herne Conference in 2008 occurred in the shadow of Pam’s diagnosis with advanced pancreatic cancer. Not even her chemotherapy regimen could keep her from participating in Herne I. She was so pleased with the format, the content and the process of this conference. At its conclusion, she began to make the plans for the next Herne Conference in 2009, a smaller conference designed to further develop the concepts introduced in the first conference and to finalize the best practice recommendations. Although she succumbed to her illness in November of 2008, Herne II did occur as planned with a smaller number of attendees and more intensive group work. This conference was dedicated to Pam’s memory, offering the first Pam Eisen Memorial Lecture given by Professor Tony Holland. Although Pam did many extraordinary things during her IPWSO presidency, the Herne Conferences are her legacy; she successfully focused the spotlight on the caregiver of the PWS person, the provider of PWS services, and the professionals who support the PWS person, their family, their caregiver, and their system of care.

This volume of work contains the written reports from almost all of the work groups from the Herne I - 2008 Conference plus the actual power point presentations of the posters. One additional work group from the Herne II - 2009 Conference (*Interpersonal Relationships*) and the Best Practice Guidelines for *PWS Behavior Management* and for *PWS Structure of Living* from the Herne II -2009 Conference are also included. Dr Holland’s presentation of the Pam Eisen Memorial Lecture has been transcribed for inclusion here. This conference book defines the **Best Practice Guidelines** for the *standard of care* of individuals with Prader-Willi syndrome. With her vision, dedication and persistence, Pam set the bar for us all!

Janice L. Forster, MD  
Editor

## **From Pam**

*(This is a transcription of Pam's welcoming speech delivered at the First International Caregiver's Conference at Akademie Mont Cenis in Herne, Germany in June 2008.)*

Willkommen! What a pleasure it is to welcome you to the International PWS Conference. It feels great to be alive and here with you in Herne for this historical meeting of PWS experts. I want to thank you for your get well wishes and encouragement while I battle cancer. As members of our global family, you have been the "the wind beneath my wings." This day has been a dream for a longtime, and now, with the help of many of you, it is becoming a reality.

I invite you to participate actively in this conference. The resulting consensus statements and suggestions on best practice standards and guidelines of care will have a global impact on many people with PWS and their families. Your thoughts based on knowledge and experience can make all the difference for families in emerging countries where they are just beginning to develop basic services. They are relying on you. For our developed countries where we have some services or even many advantages, there is still a lot to learn by sharing information across cultures. You may find solutions to challenges in the least likely places. Please take advantage of this opportunity to develop a tight network among colleagues to share and to learn.

For the first time in IPWSO's 17 year existence, we are holding a separate conference specifically for Caretaker's of people with PWS. We are giving prominent recognition to the professionals who face the daily challenge of giving people with the syndrome the opportunity to develop their full potential and to live a good life.

What is IPWSO? What is the significance of this conference? Why did we decide to hold this conference? What makes it so unique? What do we want to accomplish? What is our vision? In this presentation I will answer these questions by giving you historical facts about IPWSO. I will explain the origin of this conference, the format, expected outcome, and our vision in continuing the work we will accomplish in these few days. Our past and present board members are extremely excited about the possibilities this conference can offer to our members.

The International PWS Organization is an umbrella group. IPWSO is global in its scope, reaching into the far corners of the world. In 1998 IPWSO represented 21 countries. Now we are the voice for 80 member nations. Each member country of IPWSO consists of two delegates: one professional and one parent. We are proud and fortunate to have our office situated in Italy at the prestigious Baschirotto Institute for Rare Disease, fondly known as B.I.R.D. In cooperation with the Baschirotto's and other monetary grants, we are able to provide free diagnosis in countries where this is not yet possible. Imagine the difference this makes to our

members from emerging countries where there are no services or programs for PWS. Our diagnostic services and educational materials are precious to them.

When I first joined its board in 2002, IPWSO began to develop special services by distributing educational packets worldwide in many languages. Now we are recognized even by the UN as the *strongest worldwide educational network for a rare disease group*. Since 2004 we have continued our growth, distributing precious Medical Alert Booklets to be used specifically for visits to doctors and especially hospitals in times of medical crisis. Our DVD, **Food Behavior & Beyond**, with Dr Janice Forster and Dr Linda Gourash has been used around the world by parents as well as professionals in daily management of PWS. Many of our educational materials have been translated into other languages, and other professionals such as Dr Palma Bregani and Dr Irune Achutegui have shared their experience with PWS by developing and leading two courses for Spanish-speaking psychologists.

IPWSO has made a world of a difference to our families in emerging and developing countries. We have increased awareness of PWS by taking our display and educational materials to major international medical conferences. In addition to offering free diagnosis in countries where it is not affordable or possible, we have recently launched our most recent project, a course for clinical biologists and physicians teaching genetic diagnostics at B.I.R.D. Now that we have achieved early diagnosis and increased awareness of PWS management, we find ourselves with an expanding population of adults with PWS.

As a board member of IPWSO in 2002 and 2003, I traveled to Brazil, Mexico, and Chile for Latin American Conferences. Although the language and culture was different, the needs of these families were the same. The reality was a paucity of services. Families would gather at our hotel after conference hours begging us to talk about solutions. Many parents declared gratitude for the information we brought on the genetics of PWS, nutrition and other aspects. But, they stayed late at night and appeared outside our hotel rooms early in the morning, asking for help with management and daily solutions. They asked about group homes, respite programs, and rehabilitation services. Despite the cultural determinants of handicapped children continuing to live at home throughout their adulthood; one mother proclaimed that she needed a group home desperately. Then she said, "what *is* a group home? What does it look like?" Wherever I went, the same questions popped up. "What can we do for better daily management? Can you help us develop services based on experience and knowledge?" These questions were at the origin of the development of this conference.

In 2002 after being elected President, I set out to explore services for individuals with PWS including group homes, supportive living services, rehabilitation programs, respite care, and summer camps. There were many interesting exemplary models with different solutions to the same problems according to different cultures and realities. Among the many programs, I saw Frambu in Norway, a Rare Disease Educational Center, where people with PWS receive a

multidisciplinary evaluation while their parents are educated about PWS during a week long respite. I also visited the individual cottages on the island of Fejø, Denmark. In Germany I saw group homes with plans for elder people with PWS accommodations as well as a group home on a farm. In Switzerland a large facility is divided into small intimate living pods where each resident has individual living quarters. Through this study of successful programs, it was evident that there are basic universal perceptions about care-giving for this challenging syndrome.

While in Germany exploring group homes I met Norbert Hödebeck-Stuntebeck, a psychologist whose specialty was working with persons with PWS at the Diakonischen Stiftung Wittikindshof. I was particularly interested in the PWS curriculum that he was developing with Dr. Hubert Soyer, director at Regens Wagner Absberg, an institution in Bavaria. With the cooperation of PWSA-USA, I invited Norbert to come to the USA Conference in 2005. Here we spent a lot of time talking about developing global standards of care, and how we could make this a reality.

In 2006, Norbert Hödebeck-Stuntebeck, Dr. Hubert Soyer, and I met in Toulouse, France at the PWS Expert's Meeting. Here we established an IPWSO task force with the purpose of planning a conference to extend our present knowledge of PWS across country and cultural borders. Our goal was to utilize universal basic information to develop guidelines and standards for the caretaking of individuals with PWS. These basic components of care could be modulated to fit individual cultures. These standards would help countries develop services based on knowledge and experience. At the same time, countries with developed programs could improve their care by opening their minds to new ideas.

The first steps began at the International PWS Conference in Cluj, 2007 where caretakers across the globe developed topics for discussion at this conference in Germany. In 2007 I visited Norbert again in Germany where we worked assiduously for three days to develop our format and identifying our group leaders. In March of 2008, Norbert and Hubert joined our International (Exceptional) Board Meeting at my home in Pennsylvania (USA) to work further on our parameters for the conference. What were our expectations from this think tank conference? We wanted the answer to these fundamental questions: *What should we all be doing? What is the minimum we must provide? How can we build upon this?* We cannot expect complete consensus in each group and on each topic, but there can be consensus on suggestions and recommendations as well as topics for future discussion. At most conferences we are offered a *smorgasbord of ideas*. We go to the lectures and take what we need or want. This conference is different. Here we will work together with a leadership team to prepare our banquet. Based on suggestions made in Cluj, we have developed the "menu" with pre-set topics. All of you as attendants will bring the ideas (food). Our experts will present sample "recipes" or true and tried methods, and the work groups will decide on the final models for best practice, which are our *recipes for success*.

From this conference we will publish a document...a guideline of fundamentals...the “best practice” *standards of care* for people with PWS. Imagine the impact that this manual will have when it is translated into many languages and presented to politicians, nurses, doctors, dieticians, caretakers, parents, families! The guidelines we establish now will be basic and easily adaptable to meet cultural and reality differences. On some topics we may have consensus, and on other topics we may have suggestions for change; some aspects will need further discussion. We will need to continue our work and build on this foundation. Much of this effort can be accomplished on the internet through e-mail and SKYPE conferences. Through this technology we will continue our workgroups, perhaps adding more topics in the future. It’s going to be an ongoing dynamic experience!

The success of this conference depends on each of you. How can we meet our goals? How can we sort through our “recipes” to find the “cream of the crop?” Be constructive and positive. Look for affirmatives. Keep your mind open to new possibilities. You need to be willing to suspend your own belief system so you can explore new ideas and solutions. Bring your experience and knowledge and most of all your heart! The solution to world’s greatest problems begins with one determined person. I encourage you to be that person! John Kennedy *said* “If not us, who? If not now, when? Let us begin!”

**CARPE DIEM!**

## **Acknowledgements**

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## **Dedicated to Pam**

...who had the vision, dedication and persistence to make dreams come true...

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# The Pam Eisen Memorial Lecture

by Professor Tony Holland

Thank you very much for those kind words in your introduction. It is very difficult to follow Giorgio's singing of *Ich Auch*. Every time I have heard it – and I have heard it many times now – even being a rather reserved male Englishman, it still creates such strong emotions for me. It has been and continues to be a real pleasure to be part of this international Prader-Willi community over so many years. It is this combination of people with PWS and their families, clinicians, researchers and those who work in services that make up this community and is illustrated so well at this conference.

It is an extraordinary honour to have been asked to give this first “*Pam Eisen Lecture*”. Whilst of course moments like this are tinged with sadness I very much want my lecturer to be a celebration and for it to illustrate to you how partnerships like we see here today can achieve so much. Something Pam tirelessly worked to achieve.

I thought long and hard about how I could frame this lecture in a way that both enabled me to talk about research but at the same time honoured Pam and honoured IPWSO. In the end I decided to do it like this - Pam was born in 1948 and her daughter Gabi, who has PWS, was born in 1980, it therefore seemed appropriate to use these dates as a way of reflecting on what has changed for people with PWS during Pam's and Gabi's lifetimes. What would things have been like for a child born in 1948 or 1980 with PWS? And what is it like for families now? Has research and new understanding brought change?

This talk is a very personal perspective. Given that I am a psychiatrist, there will be a tendency to focus on mental health, but I will try and do justice to other aspects of research into PWS. In reviewing how knowledge has changed the message I want to leave you with is that these partnerships, which are illustrated here today, are absolutely crucial. Whilst research can result in new knowledge and eventually develop new treatments - the use of growth hormone perhaps being the best example - these advances are only of value if they are delivered to people with PWS and to their families. It is this translation of research from the initial stage of new knowledge and the subsequent development of new treatments into day-to-day practice that I believe IPWSO and all of the national PWS Associations are so important for. The struggle of course is that PWS is a rare disorder and new knowledge in such cases takes time to spread. Most doctors and other professionals will go throughout their working lives without seeing someone with the syndrome. So it is you as families and as paid carers, both through your experience supporting someone with PWS and through the

knowledge that you acquire, who can perhaps best inform the different professionals and new care providers.

Before reviewing how things have changed over Pam's lifetime and since Gabi was born I would like to say something about how Pam had an effect on me. Many of you knew her very well, and we have all learnt more now just listening to Janalee and hearing Giorgio speak. So this is a personal memory of her. I think of Pam first and foremost as a mother of a daughter with PWS. It was always clear on the occasions I met her that it was this fact which underpinned her passion and it was being a mother of a child with PWS that helped her continue throughout her illness and to come to the last conference here. It was being a parent that gave her the understanding and the sensitivity which informed other aspects of her role as President of the International Prader-Willi-Syndrome Organisation.



Perhaps the second way I saw her was as the person standing up like I am now behind the lectern talking to different audiences and informing them and telling them about the needs of people with PWS. She did that sensitively, she did it powerfully, and she did it in a way that I have no doubt has brought about change in many different countries.

The third way I remember Pam is the person who was small in stature and in size, was big in personality, in her smile, and who would always ask me what we were doing in Cambridge? She would always be so positive. That encouragement was really very important as research has its ups and downs. So during the course of this talk I want to reflect on these three different aspects of Pam's role, particularly when she was a President of IPWSO, but I also want us to reflect on how things have changed for people with PWS and what there is left to do.



In preparing this talk I asked myself, firstly, from my particular perspective and knowledge of PWS - What have been the key advances in our understanding? To what extent have these advances and in what way have they informed policy and



changed practice? Research is all very well but unless it brings about change and unless it is translated into practise then it is of limited value. When Pam was born PWS had not been recognised and, although by the time Gabi was born, it

was well recognised, there were many unanswered questions. Now in the 21<sup>st</sup> Century what can we expect in the way of new treatments and interventions that may change the lives of people with PWS?

Very early on in my preparation for this talk the international nature of research became very apparent – something Pam would have acknowledged and been very proud of. This next slide just lists some of the early publications in the scientific literature describing a group of people - sometimes only one or two people, sometimes more - who we would now refer to as having PWS. In the slide I have listed the dates of publication and the country. As you can see, the first is the famous one by Prader, Labhart and Willi. In the English language scientific literature, Bernard Laurance, a paediatrician who became the first President of the UK PWS Association, was the first to describe a group of children with PWS, but it was also described in many other countries - France, the USA, Sweden, the Netherlands and Belgium. I am sure that there are other reports I have not come across or cases published in journals of specific countries.

## Early reports of people with PWS

- Prader et al 1956 Switzerland
- Laurance 1961 UK
- Gabilan 1962 France
- Zellweger et al 1962 USA
- Prader and Willi 1963 Switzerland
- Sanchez Villares et al 1964 Spain
- Forsmann and Hagberg 1964 Sweden
- Monnens and Kenis 1965 Netherlands
- Hooft et al 1966 Belgium

The point I wish to make is that this process of describing a group of people with similar characteristics in a scientific journal is the way that research first gets started. Groups of children, who appeared to have certain things in common, are then reported by others who are asking themselves whether these observations that others have made are correct. Are there groups of people who have in common these particular problems and might there be a similar cause for their difficulties? What these observations, from many different countries, showed was that these children did indeed have similar problems to those initially described by Prader et al. The importance of this is that the observations from different

countries gave validity to Prader's initial observations acknowledging that there was something real here and we need to understand what causes it. This was a very important move forward and I'll come back to this particular theme in a moment.

From a different perspective the first PWS Association, (the USA PWS Association) was not established until 1975 - nearly twenty years after Prader's description. The next Association (the UK PWSA) was many years after that in 1981. So there was a big gap between when PWS was first described and when things began to move in terms of parents getting together and starting to advocate and argue for the needs of their children or adults with the syndrome. Subsequently other groups of parents became established in different countries - these parents being determined to see things improve and to ensure that in the future new parents would not have to endure the ignorance and late diagnoses that they as parents endured. This was the case in the UK and I am sure that this is true in all the countries represented here and those who are not. It is the group of parents who finally decided "We need to do something!" who then form an Association and it becomes established in a country and these Associations then help to push for research and services and ensure that policy development takes place for people with PWS in their country. Eighty-one countries are now members of IPWSO.

I want to focus now more on research and ask the question "Once PWS was described how did those doing research approach investigating PWS?" I will start by very briefly summarizing what is very obvious to you - that is the characteristics of people with PWS. What research has to do is to try to explain the fact that a group of children have been now been identified who have at birth particular features in common, however, these characteristics then change over time. After two, three, four years of age there is this switch from an infant who cannot feed properly, who is not thriving, to this infant who overeats - in addition, why is it that these children also have abnormalities in terms of their sexual development, their height, and they also have eating problems and they have a cluster of behaviour difficulties that seems to come together? The challenge is to explain these diverse observations. The way that research is then done is that specific aspects are investigated and groups with different expertise work away at understanding their particular bit - genetics, eating behaviour, endocrine function etc, One of the challenges, both for researchers and for IPWSO, is how to deal with this - how to bring these disparate pieces of knowledge together into a coherent whole.

When Gabi was born it had only just been established with certainty that PWS is a genetic disorder. Since then we not only understand much more about the genetics of PWS but also about some of the regulatory mechanisms of the body that control eating and those mechanisms that are responsible for growth and sexual development - and, as a consequence of this, the need for growth hormone supplementation for people with PWS is now accepted. We also understand more about the cognitive and functional development of people with

PWS, and we have identified some of the differences depending upon the genetic subtype of PWS. We understand much more about the behavioural difficulties and the psychiatric illnesses that are commonly associated with people with PWS. Importantly, as life expectancy has improved, we are now moving into an area of research concerned with some of the problems associated with age. What are the causes of death and how can we reduce the risks of life-threatening illness and premature deaths? Most importantly genetic advances now allow an accurate early diagnosis and together with these other developments described above we have more informed policy and practice. Much of Pam's contribution was in the dissemination of this knowledge to both families and to those working in services.

I want to consider these issues in more detail and here I am very much speaking as a researcher and reflecting on the processes that I think we go through when thinking about research strategies. This process is not exclusive to the research community. As Pam recognised we need meetings such as this. We need to bring together the different organizations and partnerships because it is at such meetings that ideas form and the research questions are developed. Someone has to recognise that there is a specific problem and then ask how we might investigate it. I can remember attending meetings of the UK Prader-Willi-Syndrome Association when Bernard Laurance would ask parents to report how often they had observed this or that in those with PWS. Did their child, for example, ever vomit? I remember that no parent put their hands up. This at the time seemed a bizarre and striking observation. But there were many other examples where the ideas arose through casual conversation and the thinking that followed then led to research and perhaps ultimately to new understanding. The task of the researchers is then to formulate and develop that idea and focus it down in a way that allows that particular issue to be investigated.

I think the second important point to make is that research requires a combination of different approaches. It requires both "fundamental research", that is, for example, asking questions about basic genetics or about the way the brain works. Such research may not have any immediate or obvious value in terms of changing the life of a person with PWS but ultimately it allows us to move to the stage of applied research where one is asking the question "How can knowledge that we have gained from fundamental research change the way we support people with PWS"? I know for many of you it can be frustrating that researchers ask for money for research and ask for people with PWS to participate and nothing much appears to happen. But a lot of research is about, in the first instance, gaining new knowledge. That new knowledge does not always change policy and practice but it is needed so as to lead to applied research which can then, hopefully, bring about change. So it is important to think about these two different aspects of research, and I will illustrate these in a moment.

The third point I would like to emphasise is that research can only happen if you have the necessary collaborations and expertise. PWS cuts across so many

different aspects of the biological and behavioural sciences, no one person or discipline can have all the knowledge. You have to bring together the expertise of different sciences and different scientists, with the knowledge of members of national associations. It is for this reason that the IPWSO scientific meeting every three years is so important. You also have to have the necessary technologies to do the research. Perhaps the best example of how ideas, together with new technology, can drive research is that of genetics. This discipline has advanced beyond all recognition. When Pam was born we did not know how many chromosomes there were in human cells. We did not know about the structure of DNA, the basic code of life. That was first proposed by Watson and Crick in Cambridge in 1953. So a lot of genetic research into possible genetic disorders just would not have been possible in the early years of Pam's life. However, as technologies and understanding developed ideas in genetics were really beginning to take off by the time Gabi was born.

So, technology is important. Another example: of technology is that of brain-scanning. I will illustrate this when we look at the issues around the eating disorder. But, of course, the other point is that, when it comes to clinical research, none of would be possible without people with PWS and their families being prepared to answer yet more questions, fill in yet another questionnaire, give yet more blood or whatever it happens to be. All these things are important, but it is sometimes very demanding and the process of research can be difficult because it raises complex and painful issues for people with PWS and their families. But no clinical research could happen without the participation of people with PWS and their families.

Research advances over time as technologies are developed and new theories are established. This has been very true with PWS but it is also true with other illnesses and developmental disabilities, such as Down's syndrome. However, as described, research starts with the recognition of the syndrome often many years before any meaningful progress is made – by Dr Langdon-Down in the case of Down syndrome and Dr Prader and colleagues in the case of PWS. You started with these case reports and clinical descriptions and then you try to identify the cause. Is it genetic? Is it due to some environmental factor? Having done that it is then possible to begin to systematically look at groups of people with the syndrome or undertake population-based studies. You move from describing individuals to describing all those with a given syndrome in a particular population, so that you can answer the questions “How much of what was found in small numbers of individuals with the syndrome is universally found - the overeating, short stature, the temper outburst etc – and if it occurs in many, but not all, how to explain that?”

These population-based epidemiological observations lead to a rather different phase. This is very much the phase that we are in at present, and that is what I call “investigating the mechanisms“. The challenge of research is to be able to explain How this particular genetic abnormality on chromosome 15 leads to a floppy baby; or to overeating or to having an intellectual disability or whatever it

is? For example is this fundamental a problem of the brain? Is there some other explanation? And in the case of behavioural problems how do biological factors, on one hand, and environmental factors, on the other, interact and lead to the sort of difficulties that we all recognize?

One important change is that we have moved away from this idea that somehow we will be able to cure PWS completely but rather to recognise that what we need to do is to look at the individual components that make up PWS and ask what can be treated? For example, growth hormone, corrects the short stature and the small hands and feet, and it changes the physical features of someone with PWS. So someone in the younger generation of people with PWS, who has been on growth hormone, may not have the recognized physical characteristics of PWS. The challenge of course is to find new ways for treating the eating disorder or ensuring that the psychiatric and behavioural problems are prevented or at least appropriately and effectively managed.

Now I want to move to looking at some individual advances. These are a very personal choice - in some areas I have more knowledge than in others. Genetics is a very difficult area and I struggle to fully understanding the genetics of PWS as it has proved to be far more complex than was ever anticipated when the chromosome 15 abnormality was first described. I remember attending an IPSWO meeting many years ago in which the geneticists were claiming that they would have the answer by the time of the next meeting. They still do not fully have the answer. What I have done, and this is not a complete list, is listed some of the research and some of the publications that have led to significant advances in this field of genetics as applied to PWS.

## Key landmarks - genetics

- Deletion identified on chromosome 15 at q11.13 - Ledbetter et al 1981
- Uniparental disomy of chromosome 15 in non-deletion cases of PWS - Nicholls et al, 1989
- Paternal origin of chromosome 15 in PWS - Butler et al, 1993
- SNRPN gene Glenn et al 1996
- PWS and Angelman syndromes – the importance of genomic imprinting - Glenn et al 1997
- Study of rare cases of imprinting centre defects in PWS - Buiting et al, 2003
- Characterisation of people with PWS and type I or type II deletions – Butler et al 2004
- The role of HBII-85 C/D box Sno-RNAs – Sahoo et al 2008
- Mechanisms of gender specific genomic imprinting - Horsthemke and Wagstaff 2008

Perhaps the most famous is the report by David Ledbetter and colleagues in 1981 identifying the deletion on chromosome 15. However, when people started to investigate larger groups of people with the clinical features of PWS they found a significant number of people did not have the deletion. So for many years there was a puzzle. Why is it that not everyone with PWS has the deletion? For a while it was argued that maybe the deletion is so small that it just cannot be seen. The technology at that time did not allow one to easily examine this hypothesis. It was essentially looking down the microscope at the stained chromosomes observed in the individual's white blood cells. But then there was the description of uniparental disomy (UPD) - a high proportion of those people with clinical PWS, who did not have a deletion, had both copies of chromosome 15 inherited from the mother and the chromosome 15 from the father was not present. A very significant advance was therefore the recognition of the role of genomic imprinting – maternal and paternal versions (alleles) of certain genes located on this particular part of chromosome 15 are unusual. Whether they are active or not depends upon the gender of the parents that the copy (allele) is inherited from. The copy inherited from the father is the one that is switched on, but the copy inherited by the mother is switched off. Observations based on a very different syndrome – Angelman Syndrome – which can also be due to a deletion at this site, added further support to this idea that the gender of the parent of origin influenced specific genes affecting whether the copy of maternal or paternal origin was active or not (normally both copies of any given gene are active and the gender of the parent of origin is irrelevant). This phenomenon was already recognised in other animals but it was a landmark observation in humans. Subsequently geneticists went on to describe what are referred to as 'imprinting centre abnormalities', a very rare cause of PWS. More recently, there was the characterization of the chromosome 15 deletion in more detail and the division into type 1 and type 2 deletions. Now, the fundamental research that the geneticists are undertaking is investigating the mechanisms of imprinting. Why do some genes become imprinted and how is imprinting controlled? This is the work of people like Bernhard Horsthemke - a world leader in terms of research on the imprinting centre on chromosome 15. So we now know much more about, but not all about, the genetics of PWS. I will come in a moment to some of the problems, but our understanding of genetics is very considerably advanced since Gabi was born and it would not have been possible to do any of this work when Pam was born.

So technologies have enabled remarkable advances in genetics moving from seeing PWS as due to a loss of genetic material (a deletion) of the PWS critical region (15q11-13) on chromosome 15, to recognising that the gene or genes concerned are unusually in that the maternal copy is imprinted (switched off) so that the absence of a paternal copy (due to a deletion or due to a maternal uniparental disomy) has a major effect (i.e. results in PWS), to now identifying individual genes – so called candidate genes. These genes may be the key in understanding PWS. So, the advances are first, PWS has clearly been identified as a genetic disorder. This seems obvious to us now but whether it was or not was uncertain when the syndrome was first described by Prader. Secondly, this

rare phenomenon known as genomic imprinting has been recognised as crucial to understanding the genetics of PWS. Thirdly, the possible candidate genes, such as *Necdin*, and *Magel2*, have been investigated and a group of genes, called the SnoRNAs, have been identified that may also be very important. But we still have some crucial unanswered questions. Is the full phenotype of PWS due to the absence of expression of just one gene or is the absence of expression of several genes at 15q11-13?

Although all cells of the body have the same sequence of DNA not all cells express the same genes. At present we do not know how the genetic abnormality characteristic of PWS results in the features of the syndrome. Some of our own work, led by Tessa Webb in Birmingham, is also beginning to look at the role of other genes located in that part of chromosome 15, perhaps causing some other aspects of the phenotype, particularly the mental health problems. It is an exciting time but to some extent there is a sense that the genetics has got a bit stuck – perhaps because of the complexity.

The next area of investigation I will consider and which I feel perhaps a little more comfortable with is that of the eating disorder. I just want to demonstrate in the same way that I have illustrated for genetics, how I believe things have changed. Whilst in genetics the advances have allowed an accurate and reliable diagnosis to be made at birth and that is very important because it allows families to be informed and interventions to take place, like managing the food environment. I think our understanding of the eating disorder has not, as yet, led to new treatments as such - obesity still has to be prevented by managing the food environment. In this context it is much more about the way we conceptualize, the way we understand the eating disorder. The point I want to make is that we have moved from seeing PWS primarily as a syndrome of obesity to instead a syndrome in which the potential for over-eating is the problem – I will illustrate this in greater detail.

The first systematic enquiry into the overeating behaviour of PWS was published by Silverstone and colleagues in the UK many years ago. The subsequent papers, on a similar theme, were those of Zipf and colleagues in the USA, who showed that people with PWS continued to eat and many did not slow down their eating or stop eating in the hour that food was available. Our work some years later added to what they had shown by asking people with PWS to rate whether they felt hungry or not. We were able to show that people with PWS did eventually rate that they no longer felt hungry and they felt full but they had to eat large amounts of food before this happened. On this basis we suggested that in PWS there was an insensitivity of the mechanisms in the brain indicating that enough food (calories) has been eaten to meet energy requirements – sometimes referred to as the satiety cascade. We argued that it was an insensitivity of the mechanism, rather than its complete absence, that was the problem. This concept of insensitivity informed some work that I will refer to later. To understand further the eating disorder and other aspects of PWS Dick Swaab in the Netherlands has investigated this tiny nucleus in the brain called the

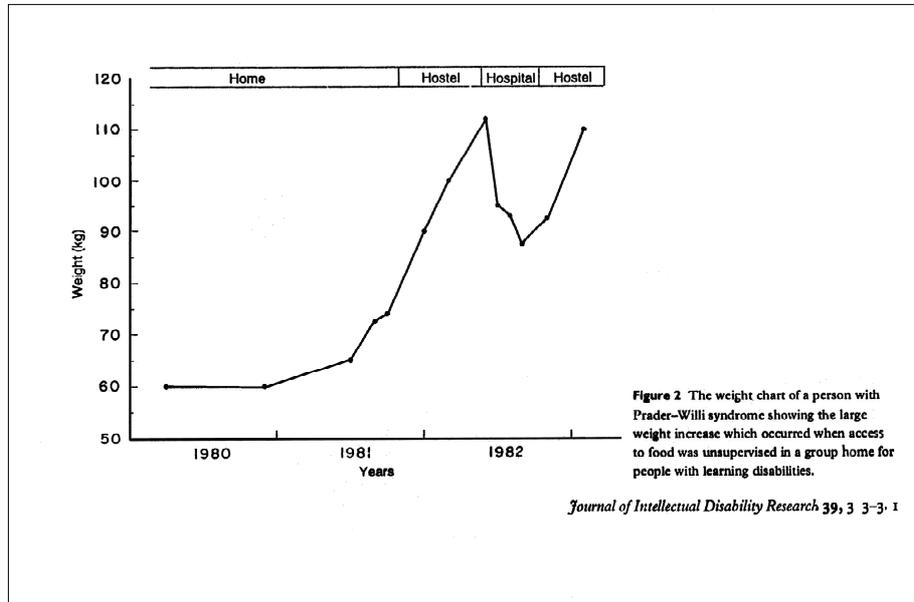
hypothalamus. To do this he has studied brain tissue of people with PWS obtained at post mortem. This work has been continued by Tony Goldstone demonstrating that certain control pathways in the hypothalamus would appear to be normal, although one abnormality identified was that the numbers of cells containing the substance oxytocin are reduced.

Outside of the brain, more recently there was the discovery that people with PWS had high blood levels of the hormone ghrelin – this hormone released by the stomach circulates in the blood and stimulates eating. If this was the cause of the over-eating the hope was that the effect of this hormone on the brain could be blocked and maybe that would switch off the eating behaviour. However, Tony Goldstone has shown that that this is probably not the case. By various means you can reduce blood ghrelin levels, however, when this is done in people with PWS they still overeat. You can see that research can have a bumpy and contradictory course. You think you have found something then it proves not to be the explanation for the problem.

PET and MRI brain scanning has been used by us and by groups in the USA to investigate the reasons for the uncontrolled eating behaviour. We showed that the control of eating behaviour and of satiety was clearly very problematic in people with PWS but it was not simply the functioning of the hypothalamus. It was the communication with other areas of the brain that are responsible for the conscious sensations of hunger and fullness that was problematic. There is a whole neural network that seems to be responding abnormally to food intake perhaps originating from a problem with the hypothalamus.

Below is weight graph of the first person with PWS I ever saw. At age 18 in 1981 she moved from her family home where food cupboards were locked and access to food was controlled, to a hostel where she had complete access to food. We now know that this extent of weight increase is exactly what can happen if you are not careful and if you do not continue to manage the food environment. This knowledge is important because you can make the case for the right support in adult life. If you

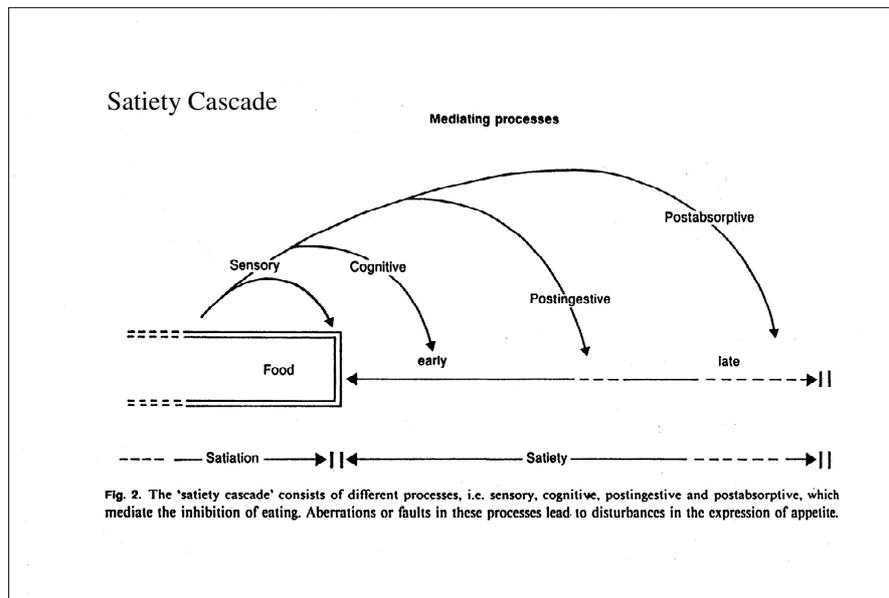
## Weight chart of young adult with PWS



allow free access to food weight can increase alarming (as shown in the weight chart) and this puts the person's health and even their life at serious risk.

Below is a schematic diagram illustrating the satiety cascade. As people eat, and as food enters the mouth and the stomach and into the intestines to be absorbed these feedback mechanisms of the body (illustrated schematically in the diagram) tell the brain that you have had enough to eat, by altering your sense of hunger and fullness – you don't feel hungry any longer and you stop eating. In PWS the behavioural and scanning studies mentioned earlier indicate that this failure to satiate or perhaps an excessive reward following food intake is fundamental what is wrong. However, why that is present is unclear - for example is it because certain types of cells in this tiny nucleus the hypothalamus have low levels of cells containing oxytocin as Dick Swaab has reported. Furthermore, we do not know how the genetic abnormality leads to a problem in the brain and in

turn to the well recognised abnormal eating behaviour.



Blundell, 1991

One major problem is that so much about PWS relates to how the brain functions and you cannot easily examine that during life. However, what has happened is that new ways have been developed, such as establishing genetic mouse models of PWS. Francois Muscatelli in France developed a *necdin* mouse model where the gene coding for *necdin* has been 'knockout' and is not functioning. She developed this mouse and looked at the behaviour of the mouse and of course with the mouse she was also able to examine the brain much more easily. So technology has allowed the development of ways for looking at the structure and function of the brain and of the chemicals in the brain by using brain scanning or the creation of these 'PWS' genetic knockout mice.

What are the unanswered questions with respect to the eating disorder? I think, we have moved from how Prader and others saw PWS has a model of obesity to seeing it as a model of overeating. Whilst there are some aspects relating to energy use, fundamentally it is a problem of excess eating and an impaired satiety response. It is for this reason that the biological factors that might be important in controlling eating behaviour are being investigated. In addition, this new way of thinking has helped us recognise the importance of controlling access to food in the living environment. You can now argue that here is a group of people who, through no fault of their own, cannot readily control their eating behaviour. You therefore need to take on responsibility on their behalf. In understanding the reasons for the eating behaviour one of the critical things we do not know is why we get this early switch at two years of age from an infant who cannot feed very well to a child that overeats. There is also a debate in the academic literature whether the over-eating is a problem of satiety or whether it

is a wider problem involving the reward systems of the brain. Such systems are often problematic, for example, when people develop addictions.

There is a lot of work still to be done here. Our hypothesis paper published in the *Lancet* has argued that we need to reconceptualise the eating disorder associated with PWS as a paradox. PWS is perhaps better seen as a syndrome of starvation, the problem is that the brain is fooled into thinking the person with PWS has not eaten and therefore understandably he/she wants to continue to eat. Such a model might also explain the low growth hormone because if you look at populations of people who have been starved their growth is stunted. The problem of course for many people with PWS is that they in fact live in a food rich society and in reality their bodies are far from being in a state of starvation – in fact quite the opposite.

I now just want to move to two or three other areas of relevance to PWS. Again these investigations illustrate some of the more general points about the process of research that are important – moving from description to investigating underlying mechanisms. For example, studies have characterized those behaviours associated with PWS in much better detail and have asked questions about whether there are differences depending on the genetic subtype or not. If such differences are found that is potentially very important because it may help us understand the mechanisms that cause such problems. For example, the purpose of some studies have been to understand why people with PWS have this marked propensity to repetitive and ritualistic behaviours or why they may become withdrawn or overactive, such as is characteristic of a bipolar mood disorder. It is now well recognized that the eating disorder is universal in people with PWS and that repetitive and ritualistic behaviours are very common, perhaps affecting 60% to 70% of people with PWS, with similar levels of people with PWS being prone to temper outbursts that continue into adult life. Skin picking is perhaps less common. From more detailed investigations some of the mechanisms that link the genetics to the behaviour are beginning to be understood. For example, I would argue that repetitive and ritualistic behaviours and the temper outbursts are fundamentally because people with PWS have arrested development - the characteristics of these repetitive and ritualistic behaviours are what you see as part of normal childhood development. Children aged two, three, four, five ask repetitive questions and so on. It is as if people with PWS get stuck in that phase. If this interpretation is correct it follows that such problems are not strictly an obsessive-compulsive disorder rather it is a developmental problem that leads to these repetitive behaviours. The work of Chris Oliver and colleagues in Birmingham, UK, have shown that particular cognitive problems referred to as 'set shifting' may be associated with these difficulties and with the propensity to temper outbursts. There is an interaction between biological risk and environmental factors that result in such behaviours.

Another example is the investigation of the risk for psychiatric illness. This work was based on our population-based study and resulted in surprising findings. In this study we tried to identify everyone with PWS in one particular region of the

UK. When we interviewed the 25 adults and their families we found that seven of the adults with PWS had had a serious illness characterized by hallucinations and delusions. Completely to our surprise we found that this was generally affecting those people with the UPD form of PWS. This led us into a completely different direction because it showed that it was not having PWS by itself that resulted in the occurrence of a psychotic illness, it was something about having UPD of chromosome 15. This then led to our work in which we tried to look at the effects of other genes on chromosome 15. This finding of excess psychotic illness in those with PWS due to a UPD has been replicated by Annick Vogels in Belgium. The importance of such replication is to ensure that such findings are indeed correct.

So what progress have we made from a psychological and psychiatric perspective? I think we have characterized the nature and extent of the problems themselves in greater detail. We have identified where there are differences, for example, with cognitive abilities depending on the genetic sub-type. Elizabeth Dykens, for example, has explored this further investigating the special skills with jigsaw puzzles found in those with a chromosome 15 deletion, not in those with UPD. Such observations remain unexplained at present. What is it that is different about those genetic causes of PWS that lead to these subtle differences? From a psychiatric perspective we are also aware of the risks of affective (mood) disorders and particularly the potential for psychotic illness more commonly in those with PWS due to the maternal UPD of chromosome 15. But the problem here is that, certainly when it comes to problem behaviour there is not an instant cure or an immediate treatment. Rather it is about the way, on a day-to-day basis, you support people with PWS – how you respond to people with PWS and how you manage the environment. The complexity of the behaviour problems and the risk of the development of additional physical or psychiatric illnesses means that proper assessments are needed when such problems occur – the expertise for such assessments may not be readily available. The important issue is to identify the nature of the problem – is it a long-standing behaviour problem or are these behaviours due to the development of an additional mental health or physical illness, or perhaps a mixture of several things? The real struggle is then how to get this knowledge into the care environment and change support practices. Where mental illness may be a problem then the careful and appropriate use of medication may be of great benefit.

As mentioned at the beginning of this talk I have, for obvious reasons, focussed on behaviour and psychiatric problems but I also want to mention growth hormone. The use of this has been one of the major changes for people with PWS. However, I am not an endocrinologist and my knowledge is limited. As mentioned earlier the use of growth hormone is a very good example of how having characterized the relative growth hormone deficiencies in people with PWS this then led to controlled trials of growth hormone replacement in the USA and in Sweden. We now realize that growth hormone replacement does more than increase height, it improves the body mass, it helps muscle strength, it

improves respiration and so on. The questions now are, at what age should it be started and particularly will it help the floppiness observed in infants, and whether it should be continued into adult life? Are the benefits and the necessary funding justified and are there any risks?

One particularly serious issue relating to PWS that has received attention has been that of life-expectancy and associated morbidity and mortality rates. There are reports of excess risks of specific physical illnesses such as scoliosis, diabetes mellitus, sleep disorder and respiratory problems. Based upon our population-base study we estimated a mortality rate of about three percent per year for people with PWS. Below is some data from a study examining the hospital admissions of people with PWS:

## Clinical and morbidity review of PWS in Western Australia

Thompson et al, 2005 JIDR

- 56 people with possible PWS (46 definite genetics) – Disability Database since 1953
- Birth Incidence 1:29,500
- 46 with definite PWS admitted to general hospital on 328 occasions (mean 7.1)
  - Respiratory disorders
  - Dental care
  - Musculoskeletal, cellulitis, ingestion of toxic substances, congenitalSix had psychiatric admissions

As with these and studies from many different countries identify the main causes of death the question then is how best can such deaths be prevented? One important approach is the development of good clinical practice guidance, such as those developed by a group led by Maithe Tauber in France. The American Association and IPWSO have also developed guidance that are really trying to help families deal with these issues and ensure that health professionals are aware of the potential problems. These approaches might prevent premature death and also ensure that proper treatment is provided if someone with PWS is ill. With improvements in life expectancy the question will then be whether older people with PWS have an increased risk for those illnesses associated with later life, such as dementia, osteoporosis etc?

The focus of this talk has been primarily about health but what about social aspects of support such as education in childhood and employment in adult life? Are there opportunities available? How can the lives of people with PWS be improved in terms of opportunities but at the same time how can they be

protected from the risk of overeating and all the things that then go with that? This is a major challenge.

So in conclusion I first really want to bring the talk back to Pam and our memories of here. I hope that some of the things that I have touched on and some of the points I have made during this talk would have been very close to her heart. These might have included the following: firstly, the international nature of work in this field, whether it is research or whether it is about care. We are all involved and any advances that are made in one country have relevance to people with PWS in another country. We are an international community. This is particularly important because of the rarity of the syndrome. Some research can only be done by bringing together people and families from different countries. It is about partnerships between different scientific disciplines as well as partnerships between national associations, people in research, social care providers and so on. Secondly, research is certainly multi-disciplinary. The complexity of PWS requires the skills of many different disciplines. Pam recognised this through speaking to interdisciplinary and diverse audiences. Finally, it is about having strong advocates – it is about ensuring that this knowledge and the findings from research are put into practice. I think that task has proved much more complex than was anticipated. I want to end by paying tribute to the work of all of the national associations and IPWSO and people like Pam and others who were and continue to be pioneers. Pam was a real advocate for research and for the dissemination of knowledge. Hopefully we will see the development of new treatments, such as for the eating disorder. There are effective management strategies for the behavioural problems and treatments for psychiatric and physical illnesses - we just need to know how to put them into place and to ensure that they are made available to everyone. I have tried to end on what is a positive but also a realistic note.

Thank you very much for listening.

# FIRST INTERNATIONAL PWS CARETAKER'S CONFERENCE

DEVELOPMENT OF GUIDELINES & STANDARDS

03. JUNE 2008 – 05. JUNE 2008  
HERNE, GERMANY

## Program

## 1. Conference day - 03. Juni 2008

<u>Time</u>	<u>Content</u>	<u>Moderator/ Speaker</u>
09.00 - 9.15	<b>Opening and first information about the conference</b>	Norbert Hödebeck- Stuntebeck (Organization committee)
09.15 – 9.45	<b>Greetings</b> 1. Horst Schiereck Mayor of the city of Herne 2. Dr. Dierk Starnitzke Director of Diakonische Stiftung Wittekindshof 3. Dr. Hubert Soyer Director of Regens Wagner Absberg 4. Michael Wedershoven Landschaftsverband Westfalen Lippe 5. Dr. Volker Holzkämper President of the German PWS association	
09.45-10.15	Lecture <b>„Idea, History and Vision of the conference“</b>	Pam Eisen President of IPWSO
10.15-11.00	Lecture <b>„Liberty and PWS – Contraries or Necessities“</b> <b>- Ethical questions in the care of people with PWS</b>	Prof. Dr. Leopold Curfs
11.00-11.15	Introduction and information about the workgroups	Norbert Hödebeck- Stuntebeck (Organization committee)
11.15-11.30	Break	
11.30-12.30	<b>Start Workgroups A1 – A4; S1</b>	Workgroup leaders
12.30-14.00	Lunch	
14.00-16.00	<b>Workgroups A1 – A4; S1 (Continuing)</b>	Workgroup leaders
16.15-16.15	Break	
16.15-18.30	<b>Workgroups A1 – A4; S1 (Continuing)</b>	Workgroup leaders
18.30-19.00	Break	
19.00-20.00	Dinner	
20.30	Culture program <b>„Circus Krönchen“</b>	

## 2. Conference day - 04. Juni 2008

<u>Time</u>	<u>Content</u>	<u>Moderator/ Speaker</u>
08.00-09.15	<b>Workgroups A1 – A4; S1</b> (Continuing and conclusion)	Workgroup leaders
09.15-10.00	Lecture <b>„Weight Management“</b>	Dr. Constanze Lämmer
10.00-10.15	Break	
10.15-11.00	Lecture <b>„Behavior Management“</b>	Dr. Janice Foster Dr. Hubert Soyer, Norbert Hödebeck- Stuntebeck
11.00-12.30	<b>Presentation of the result of the workgroups A1 – A4; S1 “Posters in a Market Place”</b>	Workgroup leaders
12.30-14.00	Lunch	
14.00-15.30	Start <b>Workgroups B1 – B4; S2</b>	Workgroup leaders
15.30-15.45	Break	
15.45-17.00	<b>Workgroups B1 – B4; S2</b> (Continuing)	Workgroup leaders
17.00-17.30	Break	
17.30-23.00	Leaving to culture program (Villa Hügel) with dinner in the region	

### 3. Conference day - 05. Juni 2008

<u>Time</u>	<u>Content</u>	<u>Moderator/ Speaker</u>
08.00-09.45	<b>Workgroups B1 – B4; S2 (Continuing)</b>	Workgroup leaders
09.45-10.00	Break	
10.00-11.30	<b>Workgroups B1 – B4; S2 (Continuing and conclusion)</b>	Workgroup leaders
11.30-11.45	Break	
11.45-12.30	Lecture <b>General health in adults with PWS</b>	Dr. Susanne Blichfeldt
12.30-13.30	Lunch	
13.30-14.30	<b>Presentation of the results of the workgroups B1 – B4; S2 “Posters in a Market Place”</b>	Workgroup leaders
14.30-15.30	<b>Expert panel</b>	Chairwomen: Dr. Susanne Blichfeld Experts: Dr. Tony Holland Dr. Janice Forster Dr. Linda Gourash Dr. Constanze Lämmer Dr. Hubert Soyer Barbara J. Goff Jackie Mallow Prof. Dr. Leopold Curfs Gorgina Loughnan Mary K. Ziccardi Norbert Hödebeck- Stuntebeck
15.30-16.00	<b>Evaluation of the conference and Perspectives</b>	Pam Eisen, Norbert Hödebeck- Stuntebeck, Dr. Hubert Soyer (OrganizationCommittee)

# Workgroupprogram

## I. Block:

**Workgroups: A1, A2, A3, A4, S1**  
**Day one (June 03) and day two (June 04)**

<b>Number</b>	<b>Title</b>	<b>Presenter, facilitator, scribe, translator</b>
A 1	<b>Environmental structure of living</b>	<u>Presenter:</u> - Karin Birkedal (Denmark) - Jackie Mallow (USA) - Winfried Schillinger (Germany) <u>Facilitator:</u> - Mary K. Ziccardi (USA) <u>Scribe:</u> - Marguerite Rubnow (USA) <u>Translator:</u> - Ilona Osana (Germany)
A 2	<b>Communication with people with PWS</b>	<u>Presenter:</u> - Dr. Linda Gourash (USA) <u>Facilitator:</u> - Anne Ogden (USA) <u>Scribe:</u> - Steve Drago (USA) <u>Translator:</u> - Andreas Bächli (Switzerland)
A 3	<b>Neurological, Psychological and cognitive aspects of behavior management</b>	<u>Presenter:</u> - Dr. Janice Forster (USA) - Dr. Hubert Soyer (Germany) <u>Facilitator:</u> - Dr. Tony Holland (England) <u>Scribe:</u> - Renate Scharfenberg (Germany) <u>Translator:</u> - Herr Schilder (Germany)
A 4	<b>Fitness, sports and motivation</b>	<u>Presenter:</u> - Georgina Loughnan (Australia) <u>Facilitator:</u> - Dr. Greg Cherpes (USA) <u>Scribe:</u> - Tina Silvast (Finland) <u>Translator:</u> - Dr. Susanne Blichfeldt (Denmark)
S 1	<b>Training for teachers</b>	<u>Presenter:</u> - Dr. Barbara J. Goff (USA) <u>Facilitator:</u> - Linda Thornton (New Zealand) <u>Scribe:</u> - Jeff Covington (USA) <u>Translator:</u> - Monika Fuhrmann (Germany)

## II. Block:

**Workgroups: B1, B2, B3, B4, S2**  
**Day two (June 04) and day three (June 05)**

<b>Number</b>	<b>Title</b>	<b>Presenter, facilitator, scribe, translator</b>
B 1	<b>Nutrition management specific for people with PWS</b>	<p><u>Presenter:</u> - Karen Balko –DVD (Canada)            - Dr. Glenn Berall - DVD (Canada)</p> <p><u>Facilitator:</u> - Dr. Constanze Lämmer (Germany)</p> <p><u>Scribe:</u> - Dr. Susanne Blichfeldt (Denmark)</p> <p><u>Translator:</u> - Herr Schilder (Germany)</p>
B 2	<b>Crisis management</b>	<p><u>Presenter:</u> - Mary K. Ziccardi (USA)            - Dr. Greg Cherpes (USA)            - Norbert Hödebeck-Stuntebeck (Germany)</p> <p><u>Facilitator:</u> - Dr. Janice Forster (USA)</p> <p><u>Scribe:</u> - Evan Ferrar (USA)</p> <p><u>Translator:</u> - Andreas Bächli (Switzerland)</p>
B 3	<b>Communication between parents and caregivers</b>	<p><u>Presenter:</u> - Anne Ogden (USA)            - Dr. Jorgelina Stegmann (Argentina)            - Jeff Covington (USA)</p> <p><u>Facilitator:</u> - Dr. Barbara J. Goff (USA)</p> <p><u>Scribe:</u> - Jackie Waters (England)</p> <p><u>Translator:</u> - Ilona Osana (Germany)</p>
B 4	<b>Environmental structure for work with people with PWS</b>	<p><u>Presenter:</u> - Renate Scharfenberg (Germany)            - Dortehe Pederson (Denmark)            - Marguerite Rubnow (USA)</p> <p><u>Facilitator:</u> - Dr. Hubert Soyer (Germany)</p> <p><u>Scribe:</u> - Jackie Mallow (USA)</p> <p><u>Translator:</u> - Monika Fuhrmann (Germany)</p>
S 2	<b>Aspects of psychological work with people with PWS</b>	<p><u>Presenter:</u> - Dr. Irune Achutegui (Spain)            - Dr. Palma Breganni (Italy)            - Dr. Tony Holland (England)</p> <p><u>Facilitator:</u> - Prof. Leopold Curfs (Netherlands)</p> <p><u>Scribe:</u> - Dr. Linda Gourash (USA)</p> <p><u>Translator:</u> - Mrs. Schmidt (Germany)</p>

**First International PWS Caregivers Conference**  
**Herne, Germany**  
**June 3-5, 2008**

Workgroup A1: Environmental Structure for PWS Living

**I. Workgroup Leadership Team**

**Presenters:**

Karin Birkedal – Denmark

Winfried Schillinger – Germany

Jackie Mallow – USA

**Facilitator:** Mary K. Ziccardi – USA

**Scribe:** Marguerite Rupnow – USA

**Translator:** Ilona Osana – Germany

**II. Work group participants:** Participants included caregivers, providers, and professionals from Germany, Scandinavia, and the USA.

**III. Abstracts:** A, B, and C

**A. Presenter:** Karin Birkedal – Denmark

**Title:** Fejo and Solvang

**The place “Solvang”**

Fejo is a small island in the southern part of Denmark. It takes 15 minutes by boat to get there and it has 650 inhabitants. If you walk around the island you will have made a trip of 30 km.

“Solvang” has 4 residents – 4 young women. Apart from Solvang there is one more institution for people with PWS it is organized differently and has 3 residents, 1 woman and 2 men. The difference between the two is that on “Marienlund” the residents are capable of being alone at night. They have their own houses and their meals are either put in the fridge or they meet in one of the houses and eat together. They have the same help as ours but they can manage many things on their own. There is one full time employee who takes care of meals, helping with shopping, cleaning and so on. Solvang and Marienlund residents meet at least once at week, where they eat together and enjoy each others company. This is very important for all 7, that they have a social life on the island with others that have PWS.

Solvang also consists of the farm “Solgarden.” Solgarden is placed 4 km. from Solvang where we have opened a shop. The shop is the place of work for 2 of our residents. In the shop we sell art, secondhand, furniture, gifts, tea, flowers and things produced by our residents. Apart from the shop there is a workshop and a stable where we have our horses. All of our young girls have their own horse. This is

where a lot of their spare time is used, riding and taking care of their horses. In the summer we also use our boat. All the residents enjoy going sailing and swimming. It is also possible to go on trips on the boat; there are bunks enough to stay the night.

### **The thoughts behind “Solvang”**

The concept of Solvang is that we are a family. Everyday life should resemble as much as possible of an absolutely normal family life. A life with a job, maintaining a household, cleaning, doing laundry, cooking, cutting the grass, taking care of the garden, painting and so on. Each and every one of our residents are individuals each with their different needs. That is why there are different rules and possibilities for each of them. Some do not need to be motivated as much as others. One is capable of having pocket money, one is able to work in the kitchen alone, some need to be transported everywhere others can take the bus or ride their bike and so on. We are honest about who can do what and why; we talk about it a lot. This is to make sure they know and understand why there is a difference in the possibilities we give them. At the same time it strengthens their friendships – because they help each other. If one can not do it, then one of the others help.

### **Weight – the main problem**

At Solvang we eat everything – “nothing is not allowed.” We care a lot about food – we make plans, read recipes, and shop with our residents. They are a part of the whole process, from start to finish. The concept we use is that we eat everything – it is the amount that counts! And the way it is prepared. There is a lot of effort put into making it look inviting and seem like a lot. They have 6 meals in a day. Breakfast, a snack, lunch, an afternoon snack, dinner and an evening snack. The size of the portions is dependant on the resident. Some get more than others because their weight is ok and they do not need to loose weight. Everyone has their own individualized weight chart and they get weighed once a week.

We do not have a physical training plan, but training is a part of everything we do. We walk, swim, ride, do gardening and so on. Some times they put on weight after a weekend at home, this is not punished. It is still not their fault that the opportunity has been there and we know, back at Solvang they loose it again, just by being a part of every day life again. It is my belief that if you live in a society that makes you feel safe and with as little insecurity as possible their system works better and they loose weight easier.

### **Working Together –**

In our concept we have few employees, because it is important that it is the same people they meet everyday. It gives them security and they trust us. Too many changes make them confused, stressed and scared. There are different kinds of conflicts, but we are always honest in our dialog with our residents if they conflict is about food – have they stolen or taken from the kitchen – then we blame ourselves!!! They are not to blame, food is their main handicap, - and we are to blame for not seeing the gap or opportunity and avoiding it. We avoid conflicts by always having

made agreements with each of them. So if we are going somewhere, they each know where the line is drawn and what is going to happen on the trip or outing. If there should come a conflict it is important that the employee that is involved in the conflict solves it. No one takes over a conflict no matter how long it takes – but there is always back-up in case it gets out of hand. But we normally solve them by talking “one on one”, giving them a chance and a “what now”. We use consequence – not punishment!! The parents will always be informed and brought in if it is a more serious conflict.

### **I believe...**

I believe that a person with PWS' world would fall apart if they do not feel comfortable and safe. If they do not know what the day brings or there are no rules and structures. It is important to take one day at a time. But most important is that you give your staff the right tools and understanding for the fact that it is never the resident with PWS' fault if they have the opportunity to steal food. A person with PWS will always have a plan and it our job to be one step ahead – ALWAYS.

**B. Presenter:** Jackie Mallow – USA / Prader-Willi Homes of Oconomowoc  
**Title:** Residential Supports - Key Components to a Successful Program -

**Key Components to a Successful Program** – In order to achieve a successful program which supports individuals diagnosed with Prader-Willi syndrome (PWS) several key components must first be established:

- Environmental Supports
- Group Plan
- Individual Plan
- Parent & Provider Relationship
- Staff Development

Each element is developed or designed to promote personal growth and development in order to enhance the individuals overall quality of life.

***Environmental Supports*** – When choosing a specific location for the program it is important to get acquainted first with the surrounding area to establish if there will be any heightened areas of concern such as walking distance to local convenience stores, or restaurants. Take the time to get to know the type of neighborhood where they will live and the surrounding neighbors, visit local businesses, assess area garbage storage and removal, and determine condition of roadways. Some things to consider when looking for a home/location may be: larger living/activity space, individual bedrooms, ample storage space, and the possibility to provide a closed off kitchen. The home should remain clean and organized, with a comfortable homelike atmosphere. It is extremely important to note that Individuals with PWS require a sense of personal safety, and in every environment from home to community, staff/family need to ensure food security. Safety can and should be enhanced with the utilization of locks and alarms. In a group living situation providing a sense of

security for their personal belongings and a sense of privacy for themselves will assist with decreasing their overall anxiety. Labeling personal belongings as well as the use of a bedroom locks and lockable storage trunks can provide an extra opportunity for the individual to have some sense of control over their own environment.

**Group Plan** - Due to behaviors and medical concerns inherent to PWS it is necessary for programs to have twenty-four hour awake supervision, which also provides another level of food security during the night hours which tends promote better sleep for the individuals being supported. Access to either internal or a well established external Medical & Psychiatric service providers is essential in assisting with appropriate attention to medical and/or behavioral/mental health crisis situations. Individuals with PWS like predictability and structure which can be met by posting daily routines, and activity schedules. To avoid problems surrounding fairness in a group setting a list of well established house expectations and guidelines should be reviewed and signed by all individuals who you are supporting. Maintaining a healthy diet through a nutritionist and by developing reinforceable exercise plans will promote weight loss and healthy personal choices. The program should provide a wide variety of opportunities for participation within the community and ensure the most success through pre-planned food security. Group meetings involving all individual who reside within the home to discuss activities, ideas and concerns opens doors for positive communication and strengthens good problem solving skills. Motivational programs assist with participation and add reinforcement to the skill development and task completion within the home. It is also important to celebrate group achievements through positive peer praise and recognition.

**Individual Plan** - Support and guidance is needed in assisting the individual with personal growth and development of their daily living skills, which will promote independence and an improved self image. Motivational plans with verbal and sometimes tangible reinforcers assist in obtaining follow through and task completion by the individual. Gainful employment or further educational opportunities should be provided for each individual taking into account strengths and limitations. Individual assessments and data collection assist in establishing clear objectives, active goals, and the development of a therapeutic approach plan which addresses the individual's specific needs. Opportunities for the individual to establish and maintain personal relationships with peers and family members can be strengthened through positive social skills training and verbal support and guidance. Open communication and allowing the opportunity for the individual to share their personal goals, dreams, likes, dislikes, issues, as well as one on one attention helps to establish the staff as a solid object. Taking time for rewards and recognition for personal accomplishments helps reinforce positive decision making and personal growth.

**Parent & Provider Relationship** – Every relationship develops and builds through mutual trust and respect. A solid unity between parents and direct care staff can be the make or break between a successful or failed placement. The individual diagnosed with PWS needs to see that everyone is on the same page. If there appears to be a hole in the relationship or in the communication structure they will

find a way to push right through and create what is known as the “PW Triangle”. In these situations they play upon vulnerabilities and create an unhealthy atmosphere for all involved. Open communication and agreed upon plans prior to sharing with the individual in care can create an environment more conducive to achieving positive or desired outcomes. When problem solving all participants need to listen and be willing to try new ideas or suggestions. Reinforce the relationship with positive praise and recognition.

### ***Staff Development***

Training – To develop essential skills prior to hands on support, staff should participate in a well established agency orientation and continue with ongoing mandated trainings in: PWS overview, Nutrition, Meal Preparation, Dietary Exchanges, Health and Medical Concerns, Activity Planning & Exercise, Resident Rights, Professionalism, Individual Service Plan, Behavior Management Strategies & Techniques, Crisis Intervention and Prevention, and attend offered PWS Association Trainings.

Support – To assist with structure and continuity amongst all team members team it is recommended to have at least bi-weekly team meetings and at least once per month individual meetings with all direct support care staff. This allows opportunity for networking, skill development and information gathering and dissemination. Each team should be able to recognize areas requiring growth and develop goals and objectives to accomplish each in a timely manner. Due to the stressful nature of the job environment, it is equally important for all teams to take note of the over all successes within the program and celebrate each success through positive praise and recognition

**C. Presenter:** Winfried Schillinger - Germany

**Title:** Environmental structures for adults with Prader-Willi-Syndrome (PWS) at Regens Wagner Absberg (Germany)

The holistic multidisciplinary care and education for people with PWS at Regens Wagner Absberg is aligned with the PWS symptoms and the individual clients' needs. It's essential to enable self-determined life decisions as much as possible according to the concept of empowerment.

**Main basics of the PWS-specific environmental structure:** to achieve a psychosocial stability, an enhancement of satisfaction with life, reduction of body weight and reduction of deviant behaviour are:

1. Daily structure
2. Syndrome specific treatments
3. Psychosocial support and care
4. Assistance in realizing individual aims and ambitions

## 1) Daily Structure

- ***PWS shared-living:*** 10 people with PWS are living in each of our PWS-group-homes. There is heterogeneity in gender, age and skills among the residents. The structure resembles to family and there is a reference caretaker system. Food is kept under lock and key at all times. Each person has a single room with an on-suite bathroom. Besides there is a dining room, kitchen, shared living rooms in each group home. Assistance is available 24 hours.
  
- ***The PWS-group home as predominant area of life***
  - Living as part of a social system facilitates encounter, relationship, and social skills.
  - Living with peers, people with similar interests and similar difficulties
  - Living a self-determined life with responsibility as opportunity to develop according to age, to become independent, to grow up and to partially disengage from the parents
  - Incorporation of individual resources and skills in the community boosts self-esteem
  - Living between self- and agent-determination
  - Participation in communal space decoration, voting rights in community rules, and communal spare time activities facilitate satisfaction with life.
  
- ***Therapies and accompanying programs***

The participation in programs and therapies occur according to individual needs, especially proven for people with PWS are creative programs and therapeutic encounters with animals in the form of asino (donkey) therapy.

  - Drama class
  - Dance class
  - Holiday and weekend programs, culture and hiking
  - Art therapy
  - Music therapy
  - Pastoral care, spiritual programs
  - Relaxation training
  
- ***Employment in a sheltered workshop as second area of life following the norm principle: living and working***
  - Inclusion and integration in heterogeneous groups instead of separation
  - Occupation according to individual ambitions and skills
  - 13 different jobs: toy assembly, metal works, computer work station
  - Daily structure to support psychosocial stability
  - Increased self-esteem through occupation and income opportunity

## 2) Syndrome-specific treatments

- ***Food management***
  - Balanced diet (1200-1500 kcal maximum)

- 7 regular meal times
- No food deprivation
- Food is kept under lock and key at all times
- Communal meal preparation once a week
- **Health education**
  - Teaching of nutritional knowledge
  - Information about long-term consequences of overweight and obesity
  - Teaching of alternative action patterns and self-modification strategies
  - Personal weight charts
- **Motion programs and fitness training** - a broad variety is necessary
  - (Nordic-)Walking
  - Cycling
  - Donkey-Trekking
  - Swimming / Aqua-Jogging
  - Aerobic, Dance and gymnastics
- **Health care through specialists and external therapists** integrated in the structure of daily life coordination and escort are executed by care staff.
  - Psychiatric treatment
  - Diabetes therapy
  - Orthopedic treatment e.g. of scoliosis
  - Occupational therapy / physiotherapy
  - Speech therapy

### 3) Psychosocial support and care:

Every PWS-client has a reference person to turn to in his group home. On top of that there is psychological and educational care and counseling available at the case manager. The case manager takes over case planning and the PWS-specific consultation of the care teams. In single case meetings the individual plans for assistance, facilitation and therapy are drawn up.

- **Aspects of psychological and educational care**
  - Support of self awareness
  - Conversational therapy for psychological conflicts
  - Assistance in relationship problems and emotional difficulties
  - Crisis intervention
  - Behavior modification intervention to decrease deviances
  - Social skills training, health programs / educative groups

### 4) Assistance in realizing individual aims and ambitions

Assistance and monitoring of PWS-clients in regards to realizing their goals, ambitions, and life perspectives (diagnostics, assessment, resource analysis) It's essential to enable self-determined life decisions as much as possible.

- **What are the needs, ambitions and aims of people with PWS? And,**

### ***what kind of environment do they require?***

It is crucial to ask the persons on following topics - as they are their own experts:

- living arrangements
- job
- spare-time / hobbies
- friendships, relationships, and sexuality
- self-determination
- nutrition, weight and health

- ***Discussion:***

- Are self-determined decisions and PWS mutually exclusive?
- Is agent-determined life the norm?
- Is it valid to say (according to Prof Prader): The measure of individual freedom correlates with body weight?

## **IV. Define the nature of the problem and why it is important:**

Throughout multiple environments and across all cultures, the workgroup participants agreed that individuals with PWS require consistently applied environmental supports. Anecdotal and clinical evidence suggests that, at a minimum, a plan for securing food and a structured daily schedule is absolutely essential. Furthermore, the caregivers' failure to provide minimal supports often leads to maladaptive behaviors, an increase in anxiety and life-threatening weight gain.

Environmental supports can be successfully provided by others (i.e. twenty-four hour awake staff supervision) or via mechanical means (use of locks and timers, for example). The supports must be clearly outlined and communicated to anyone who comes into contact with an individual with PWS. Regardless of how the environmental support is provided, all agreed that consistent use is the key.

## **V. Nature of the Problem**

### **1. Deficit in PWS specific care services - quantity and quality**

- There are still not enough residential and vocational services for people with PWS.
- Not all providers have a specialised knowledge about the management of PWS which is necessary to implement adequate, safe and accepted practices that provide needed environmental supports.
- Therefore many people with PWS do not really have the needed and accurate specific structures for a positive development and perspective of life.

## **2. Requirement of a holistic and multidisciplinary care to achieve a continuum of care**

- The environmental supports need to have a minimum standard in every setting that the person with PWS is supported in, e.g. a predefined weight management.
- Environmental supports need to work together in order to enhance the quality of life in all aspects of the person with PWS. There is often a lack of assessment, coordination and case management, which leads only to partially optimized therapies and health care. The future development of a person depends on coordinated planning of adequate therapies and care including all involved persons.

## **3. The basic necessity of individualized care besides a syndrome specific care**

- Environmental supports are manifested by group guidelines. However they may need to be individualized to meet the specific needs of the person with PWS. The danger is only to focus the PWS specific deficits and instead of comprising the individual resources and needs. Without having a perspective of personal development according to individual needs a person with PWS will not reach a psychosocial stability at all.

# **VI. Impact of the Problem on the Person, their Family, the Caregiver, the Provider and the System of Care**

For an individual diagnosed with Prader-Willi syndrome (PWS) the inability to provide them with the necessary environmental supports increases their anxiety which manifests in a lack of psychosocial stability exhibited through acting out/challenging behaviors, and decreases their ability to have a meaningful and productive quality of life. These individuals who must live in unsupported environments experience deterioration in health up to and including the risk of death.

Families who are faced with the realization that they can not find long term housing to provide adequate environmental supports for their child, may become dysfunctional affecting all interpersonal relationships. In addition, this fear of knowing that there is no long term security for their child, creates anxiety and apprehension and places unnecessary burden upon the family to make choices which may not be in their loved ones best interest.

Caregivers working in environments without the necessary training and supports increases stress which leads to poor choices and ineffective approaches to dealing with challenging behaviors. This inability to provide quality of care increases the risk of caregiver burnout, turnover, and an unstable living environment for the individuals.

Providers who are not allowed to develop programs with adequate environmental supports due to licensing/governmental restrictions, limited funding or training have a higher risk of providing poor or ineffective programs (homes). The inability to meet the client's individual needs creates a negative image up to and including possible

legal or licensing sanctions. Without consumer trust the negative feedback may reflect the future growth of the agency.

When meeting the multifaceted ever changing needs of an individual diagnosed with PWS, it requires a continuity of services through a system of care (medical, psychological therapeutics, education, legal etc.). A failure in the system occurs when the information is not presented in a thorough manner to the treating specialist or consultants. If an informed staff does not accompany the individual with PWS in the area of health care misdiagnosis may occur leading to health deterioration. This would also true when seeking other community services as their needs are not always accurately portrayed and inadequate environmental supports are not secured in a manner that would promote safe and healthy life choices.

## **VII. Discussion Summary**

Workshop participants easily agreed on many aspects of environmental supports and controls. These areas are summarized in the group's recommendations in the "Guidelines for Best Practice" section.

One discussion point of particular interest to the groups is the debate over whether people with PWS benefit most from living only with others who also have PWS. The workgroups participants agreed that, while some of the recommended best practice guidelines for environmental supports can be met in a heterogeneous environment, the majority of the critical environmental controls are most effectively implemented and accepted by those with PWS in this specific programming context. The workgroups participants expanded on this even further by endorsing the anecdotal successes and clinical evidence of immediate use of an environmental design that is specific to meeting the challenges of supporting those with PWS.

The beliefs of the workgroup participants varied as to the emphasis placed on a specific topic, but at the conclusion of the discussion sessions, no issues outside general consensus were identified.

## **VIII. Guidelines for Best Practice**

***Note: All guidelines should take into account the assessment of the individual to assure health and safety to promote quality of life.***

The individual's opinion should be taken into account when following these guidelines whenever possible.

The preferred form of living arrangement is in a PWS specific environment. This allows for consistency in treatment and a sense of fairness to the individual. All attempts to create a family like environment including their own space regardless of group size is important for the person with PWS to have a sense of belonging as well as a place to disengage from group living whenever necessary. It is vital for the

individual to have a choice of vocational opportunities. Enjoyment and fulfillment of one's own day is crucial for anyone to feel productive and an important member of their community.

Optimal success for the person with PWS will be based on the type of supports that are in place. Those supports require an understanding of the unique needs associated with PWS. All areas of a person with PWS' day should have proper supervision to assist with those unique needs. There needs to be a 24 hour type of supervision in place in either the form of direct caregiver support, alarms or security systems that alert caregiver or monitoring agency that the individual has either left the designated area(s) or that they are in need of assistance. A combination of the above may also be used.

At times additional support may be needed due behavioral issues or need for assistance from another caregiver. Each supportive environment, both home and vocational, should have a system in place that allows for quick response from another caregiver to assist in the need at hand.

Routine and consistency is essential to the successful living for a person with PWS. Inclusion of their ideas and preferences should be part of the planning of the structure of their regular routine. Meals should be managed and structured in a formal way to ensure that everyone knows the plan of how meals are to be planned, monitored, and served. It is necessary for every person with PWS to have a diet plan that can be followed by all. Exercise should also be a part of their daily routine including some levels of movement or fitness every day with the amount of time to be determined on individual basis. Healthy weight management is also an important part of monitoring the progress of supporting a person with PWS. Weights should be taken at a minimum of once per week based on history.

Structure of the home for a person with PWS should have some form of house rules for living with others. This allows for everyone to understand and remain consistent with those guidelines, expectations, and boundaries that are needed when in a group living environment. Another area is the complete security of food, money, and medication which should be locked and managed by caregivers.

To assist in overall self esteem it is important for everyone to feel a part of their community. Inclusion of those activities within the community should be individually assessed to determine environmental supports that may be needed. Understanding the person's own level of independence will assist in maintaining required needs for safety. Living in a community should include good neighbor relationships, it is important to create systems explaining about PWS.

It is essential to have individual behavior management plans ranging from positive motivation to crisis management. This once again allows for consistency and proper understanding of that specific individual's needs when that person may be in a crisis or to simply avoid or redirect the situation.

Training of caregivers is essential to the success of the individual with PWS. Caregivers need a good understanding of the complexity of the needs of the syndrome as well as the compassion that is needed when supporting a person with PWS.

Maintaining healthy relationships once the person with PWS is outside their family's home is important for the person with PWS. Contact and involvement with family and friends may need additional support and guidance from caregivers. That support may consist of teaching relationship building and in some incidences may require additional counseling. Human sexuality may also require support and education.

## **IX. Conclusion**

Across cultural lines all participants agreed that in order for an individual with PWS to succeed and live a healthy productive quality of life that a base line level of supports conducive to promoting safety and personal growth, must be in place prior to providing care. It was determined that environmental supports were non-negotiable, including but not limited to; food security, daily schedules, personal growth and development plans, trained caregivers, and a continuum of care throughout their day. It was also agreed upon, that it will be imperative that as care providers, that we continue to explore this cohesive collaboration of efforts through on-going education, training, and awareness.

## **X. Challenges and items for future consideration:**

Two specific areas emerged as priorities for future considerations. A thought-provoking dialogue occurred regarding the definition and application of self-determination, and how the concept can and should be specifically applied with people who have PWS. Areas of specific topics, which demand further exploration, include living arrangements, employment, hobbies and recreational activities, friendships, relationships, and sexuality, nutrition and weight, health choices, and levels of support required for a safe, healthy life. Similarly, the topic of the pros and cons of legal guardianship was identified as needing further exploration among the workgroup participants.

***NOTE: Please refer to the Herne II proceedings: Environmental Structure for PWS Living – Best Practice Guidelines for a consensus statement.***

QED

First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workgroup A2: Communicating with Persons with  
Prader-Willi Syndrome

**I. Workgroup Leadership Team**

Presenters:

Larry Genstil, PhD. (Israel)

Linda Gourash, MD (United States)

Facilitator: Anne Ogden, LMSW (United States)

Scribe: Steve Drago (United States)

Translator: Andreas Bächli (Switzerland)

**II. Workgroup Participants:**

Workgroup participants included parents, caregivers, providers and professionals from Germany, United Kingdom, Switzerland, and the United States.

**III. Presentations:**

**A. Presenter (in absentia):** Larry Genstil, PhD – Israel

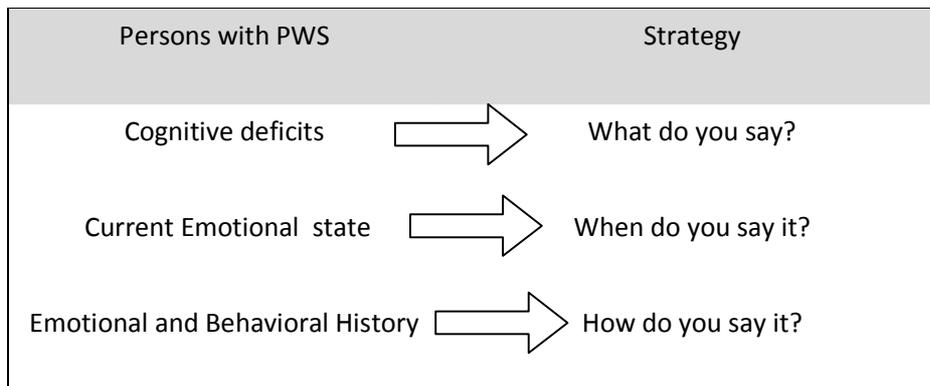
**Title:** When Communicating with Persons with PWS

- It is recommended that all communication with people with PWS be a combination of verbal and visual communication. For those who read, writing a summary of what was said is helpful. A simple drawing can also improve what was understood.
- Using sign language or other hand gestures are ideal as an adjunct to verbal communication for people with PWS.

- It is recommended to check with the person with PWS exactly what he understood. This is easily done by asking him what he heard. He should be able to repeat back what was said.
- When it is desired to communicate instructions, it is best done by clearly stating what is wanted in short, concrete sentences, accompanied by the appropriate visual prompts for that individual. When a person with PWS begins verbalizing or arguing in a way which indicates increasing rigidity, continuing to make demands of the person only increases rigidity. While there may be incidents which cannot be ignored (i.e., self-injurious behaviors), in general, calming, supportive communication is best at reducing the rigidity and preventing a possible tantrum.
- The method most recommended is similar to assertiveness training, in which a behavior that has been problematic is role-played as it previously occurred, and then role-played again as it could occur with better judgment. The entire session is video-taped. Then the participants in the group view the video and the person critiques his/her behaviors. The staff asks which role-play made the person look better, which person he/she would prefer to be, which person he/she would like to show to the world. The person is then given a homework assignment until the next group meeting for behavior rehearsal. The homework is to “practice” the behavior that was found by the person himself to be the best one for him. The combined visual and verbal way he learned of the need for change, together with the multi-sensory way of changing it (by seeing himself/herself on TV, hearing himself/herself, and from the experience of acting it out and rehearsing the new behavior), all create the conditions necessary for improved, pro-social behaviors.
- Use simple, concrete language.
- Keep communication short.
- Always use visual cueing in addition to verbal language.
- Develop a communication plan for every person, which defines the verbal and visual communication appropriate for each person.
- When giving instructions, use the recommendations above, plus demonstrate the instruction and have the person demonstrate back, thus showing he “got it.”
- When teaching new behaviors or improving old ones, using a method similar to assertiveness training with video feedback is quite effective. The new behavior is role-played or the problematic old behavior is role-played and then an improved version is also role-played. At the end of the session, the person is asked which behavior he/she preferred, which person he/she would prefer to be, etc. Then each person in the group is asked to “practice” the new behaviors until next group.

**B. Linda Gourash, MD - USA**

**Title: Communication with PWS Persons**



**Response**

***During* Disruptive or Shutdown Behavior**

**ALWAYS**

- Say as little as possible
- Use a calm relaxed, firm voice
- Act concerned but not emotionally involved.

**NEVER**

- Give IN
- Bribe
- Apologize
- Argue
- Try to Reason
- Make Threats
- Talk about the past or the future
- Tell him how desperate you are
- Talk about how he is inconveniencing others.
- Appear angry or shocked

**SOMETIMES**

- Reassure
- Redirect
- Remind\*
- Of next activity
- Of planned preferred activity
- Of planned incentive, if not already lost. (e.g. at the escalation phase.)

## IV. Nature of the Problem

Communication includes listening, talking, facial expression, tone, body language, and written communications. There can be difficulties in all of these areas. Persons with PWS have many of the following:

- Difficulties with speech production lead to “listener fatigue,” frustration or misunderstanding.
  - **Speech disorders** include
    - poor articulation
    - dysfluency
    - apraxia
- **Language skills deficits** result in difficulty putting thoughts into words and difficulty identifying how to present a narrative in response to a question.
- **Cognitive rigidity:** Persons with PWS may have a fixed idea or have great difficulty moving from topic to topic. They can be extremely perseverative.
- **Emotional dysregulation:** persons with PWS have difficulty regulating their emotional display in response to minor disappointments or frustrations
- **Mismatch between language skills and cognitive deficits** that sometimes lead us to overestimate their real understanding if their concreteness and capacity for fantasy is not taken into account.
- Frequently there is a **willingness to lie**. People with PWS can lie:
  - To tell the truth as they understand it;
  - To get out of trouble;
  - To trick people in order to get something (triangulation/manipulation).
- **Confabulation:** Sometimes persons with PWS tell fantastic stories for no apparent reason. While some of these stories can be amusing other times they are serious as in allegations of abuse.

## V. Impact of Communication Problems on Persons with PWS

Persons with PWS can be very anxious or frustrated as a result of communication problems. Their explosive behavior limits their ability to participate in community activities, transportation and work opportunities. Frustration and anxiety can lead to self injury, severe and prolonged tantrums, aggression, property destruction, running away, and involvement with law enforcement. Persons with PWS can be unhappy and ashamed when they do not feel proud of themselves and when people are angry at them.

## **VI. Impact of Communication Problems on the Caregiver**

Caregivers are also subject to frustration when communication is poor. Caregivers must provide much of the energy and motivation and constantly set limits. They must therefore communicate constantly in a positive way which is very fatiguing. If persons with PWS are successful in manipulating or triangulating staff, conflicts among staff can take place; e.g., when caregivers make promises that other caregivers must answer to, or when persons with PWS make claims that cause misunderstandings and conflicts among caregivers leading to stress on the job. As a result caregivers may not be able to continue in their profession.

Persons with PWS may calm down very quickly and appear to forget an angry incident, while caregivers may need more time to recover. Caregivers may need assistance to learn how to recover from intense experiences of emotional outbursts or insults or aggression. The team may need to step in and relieve caregivers temporarily of contact with someone with whom they have had a frightening or dangerous experience. Caregivers may experience anxiety on return to working with the individual involved.

Among caregivers, it is common to withhold sharing of information until they are certain that it is true. Also, it is common to postpone telling good/bad news until everyone is in a safe place.

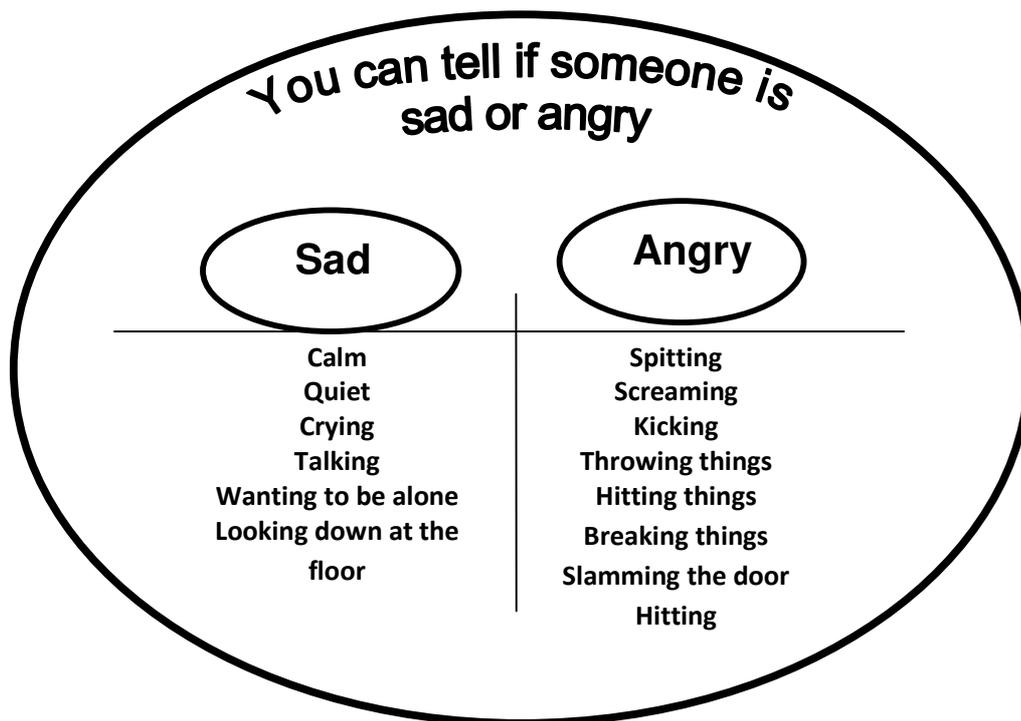
Sometimes boundaries cross when a question is too personal.

## **VI. Best Practices**

- Caregivers should be trained in active and diagnostic listening. This means they must discern the person's ability to communicate and to assess their emotional state. They should be aware that there can be a large discrepancy between a PWS client's apparent understanding and their actual grasp of the big picture, or long term goals.
- They should show concern without being emotionally reactive. Caregivers must be on guard against not making the client feel scolded or shamed; it is important to allow them to save face and to preserve their dignity.
- Care givers should learn to listen without agreeing or disagreeing. Many comments do not require a response, and there is no need to directly contradict false statements, fantasies, claims, dreams.
- Caregivers may need to allow extended periods of time to gradually redirect false beliefs and look for opportunities to provide for the emotional needs with other positive but realizable experiences. Individual counseling can be used very

effectively with persons with PWS if the counselor is patient and understands their need to cling to ideas as long as they need them.

- There is agreement on the value of proactive communication. It is necessary to choose the timing and the opportunity to communicate important information. Weekly meetings and designated communicators are used.
- Proactive communication includes teaching coping skills and scripts to help the person learn in advance ways to be disappointed and angry and to ask for help or to take action to calm down on their own. This learning takes place through discussions when the person is calm, role playing how to cope when angry, and practicing written scripts that may be a part of a contract for how to behave when stressed or angry.
- Schedules can establish expectations and avoid disappointments.
- Persons with PWS benefit from many verbal reminders and frequent praise.
- Caregivers should learn how important their choice of words can be. In particular person with PWS may overreact to negative statements or the word “No”. It is a skill to learn to turn a “No” statement into a positive statement, e.g., rather than say “No, we are not going out today.” You might say (with enthusiasm), “Yes! We are going to go next week!”
- Caregivers must be trained on how to communicate during emotional outbursts: giving physical space, knowing when to remain silent, waiting for the person to calm, intervening only as needed to keep everyone safe. It is effective to limit verbal comment to making suggestions for alternative and safe behavior.
- Caregivers must be trained on the importance of their tone of voice. Persons with PWS are very sensitive to scolding or blaming tone. Use enthusiasm or upbeat tone when appropriate. Redirect in a matter of fact or even supportive tone. If you find someone is trying to provoke, shock or upset you, the best response is to look and sound “bored.”
- Using humour is a valuable strategy to defuse a tense situation. ***Sarcasm is never appropriate.***
- Caregivers must work in a supportive team model with frequent communications among the team members to prevent misunderstanding and manipulation that leads to inconsistency. Inconsistency, even when caused by the client, creates anxiety in the client.
- Proactively, clients can learn from rehearsing how to cope when they are angry by practicing words and actions that will be acceptable.
- Counseling can help clients to distinguish their own emotions, for instance, the difference between mad and sad and the need to take time away when they are very angry in order to calm (see diagram).



## Helpful Visuals for Persons with PWS

When I am sad, I need someone to comfort me.

When I am sad, I can stay in community or family space.

When I am angry I must go into my own apartment or room.

If I feel sad, but I am doing things on the angry list, I must go into my own apartment or room.

When someone else is angry, it is good for me to move away.

## **VII. Further and Future Topics for Development**

- Assigning one or more staff persons to each client with the function of conveying all important communications; this has been helpful in some circumstances and problematic in others.
- How much venting is acceptable? When does a rant or a tantrum without property destruction or aggression require intervention only because of the noise level and duration?
- How much choice can people handle may depend on how much time they have to consider it. This may be true even of low importance choices.
- Best practices for talking about food
- Communicating about other specific difficult topics: death, serious illness, life changes, rights, grievances, privacy. Should a professional caregiver ever lie to a person with PWS?

QED

First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workgroup 3: Neurological, Psychological and Cognitive  
Aspects of Behavior

**I. Workgroup Leadership Team:**

**Presenters:**

Janice Forster, MD – USA

Hubert Soyer, PhD – Germany

**Facilitator:** Tony Holland, MD – UK

**Scribe:** Renate Scharfenberg – Germany

**II. Workgroup Participants:** Parents, professionals, providers, and caregivers from Scandinavia, UK, USA and Germany attended this workgroup.

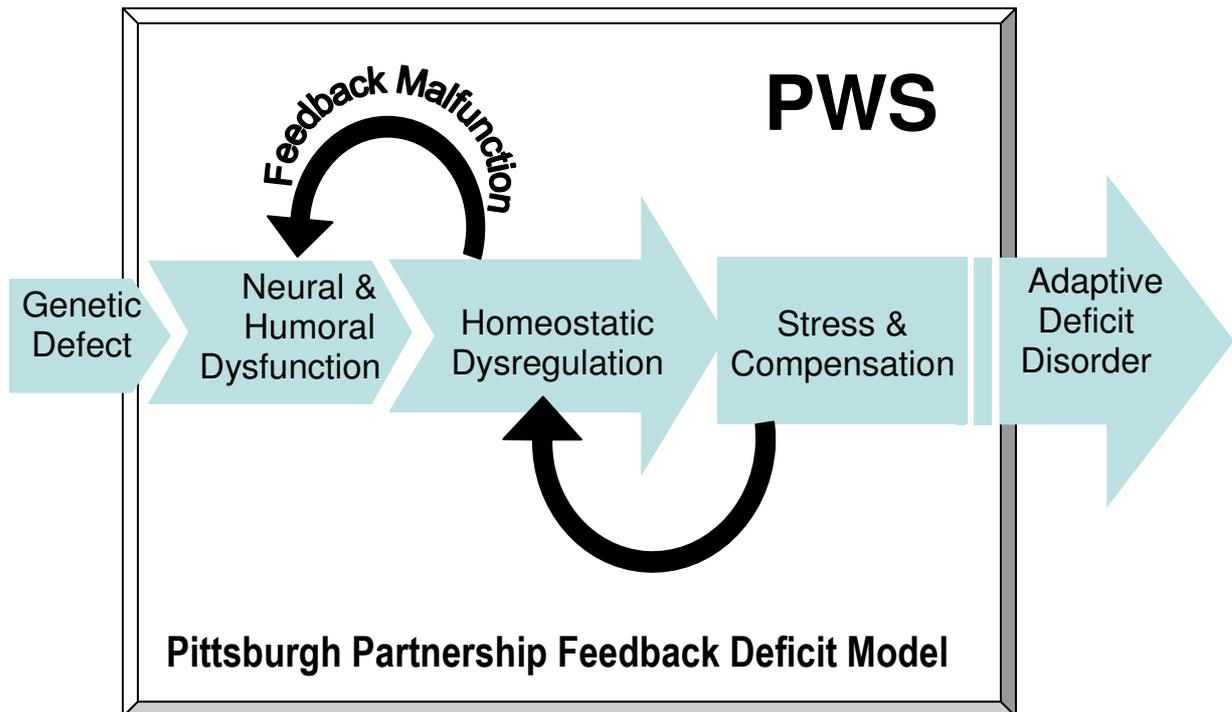
**III. Abstract (A) and Presentation (B):**

**A. Presenter:** Janice Forster, MD

**Title:** Neuropsychiatry of PWS

PWS is a genetic syndrome resulting from the failed expression of paternal genes originating from chromosome 15 q11-q13. The core features of the syndrome are food-related behaviors, hypotonia, growth deficiency, and hypogonadism. Additional features of variable severity include cognitive deficits, emotional dysregulation, behavior problems, communication problems and social skills deficits. Abnormalities of brain structure and function have been described through neuropsychological measures and functional imaging studies, as well as some studies of post mortem brains. The scientific community is just beginning to understand the development of the brain in PWS, the dynamics of its interconnections, and the impact of interventions such as growth hormone supplementation, gonadal hormone replacement, and use of psychotropic medications.

This presentation focuses on the phenomenology of PWS cognition and behavior from a neuropsychiatric and a neuropsychological perspective. Here, the phenomenology that is described is what is seen and experienced, not necessarily what is measurable. Examples of brain structure, localization and function provide the context for understanding “why they do what they do.” A neuropsychiatric model is presented consisting of **drive dysregulation**, **repetitive motor behavior**, **affective instability**, strengths and weaknesses in **learning and memory**, and universally **impaired judgment**. This neuropsychiatric model underscores that PWS personality traits are predetermined, and the absence of environmental controls, life style choice and self regulation have minimal impact on cognition and behavior in individuals with PWS. Environmental management provides hygienic stabilization, while trained staff support provides auxiliary cortical function to enhance learning potential and adaptability.



**B. Presenter:** Hubert Soyer, PhD

**Title:** Neurological, Psychological & Cognitive Aspects of Behavior in PWS

**Neuropsychology** is an interdisciplinary branch of brain research and seeks to understand the relationship that exists between behavior and the brain, that is, it intends to explain how brain activity manifests itself in observable behavior.

Neuropsychology deals with questions such as:

- What mechanisms are responsible for thought, learning and emotion in human beings?
- How do these mechanisms work?
- What impact do changes in the physical aspect of the brain have on a person's behavior?

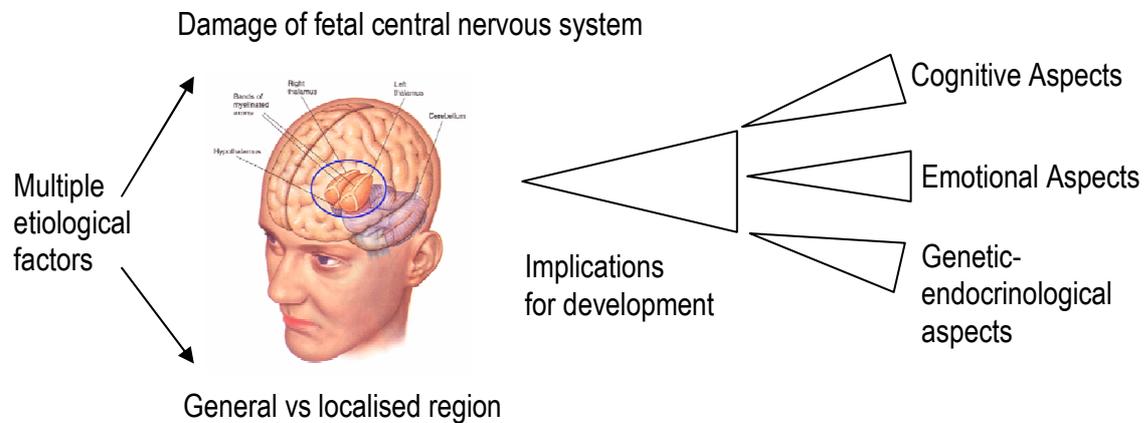
**Diagnosis:** Neuropsychology and neurology use a variety of tests to explore brain function and to understand the neurological basis of behavior (EEG, CT, MRT, fMRT, PET). Neuropsychological tests, such as The **Halstead-Reitan** Neuropsychological Test Battery and The Luria-Nebraska Neuropsychological Test Battery provide very valid data. The Halstead-Reitan Neuropsychological Test Battery consists of:

Category Test  
Seashore Rhythm Test  
Finger Oscillation Test  
Strength of Grip  
Reitan-Klove Sensory Perceptual Exam  
Finger-tip-Writing  
WAIS

Tactual Performance Test  
Speech Sounds Perception Test  
Trail Making Test A and B  
Reitan-Klove-Tactile Form Recognition Test  
Tactile Finger Recognition  
Reitan-Klove Lateral Dominance Exam  
Color-Word-Interference test (Stroop-Test)

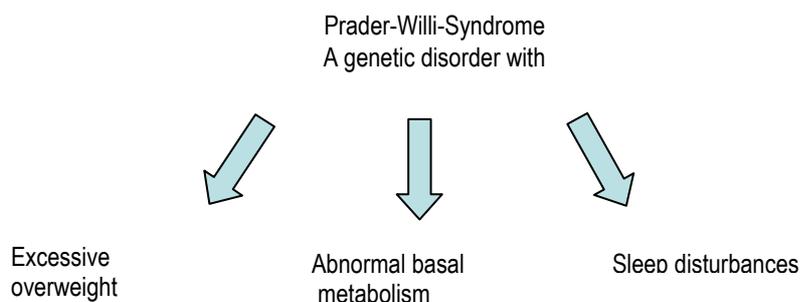
The Halstead-Reitan Neuropsychological Test Battery is a procedure requiring a great deal of time to complete and, in the case of PWS patients, can thus be carried out in a period of several days and not in one haul.

Etiology:



PWS presumes the existence of multiple etiological factors that are associated with damage of the fetal central nervous system. We now differentiate multiple genetic subtypes of PWS: two deletion subtypes, UPD condition and Imprinting Center Defect. Until recently the belief was that these factors had an effect on a narrowly-defined section of the brain, the hypothalamus and the pituitary gland in particular, but we now assume that this condition is related to a multiple impairments of the central nervous system. In this lecture, I would like to spell out the consequences this has for the patient's development by looking at three major aspects: the cognitive, the emotional and the genetic-endocrinological aspects.

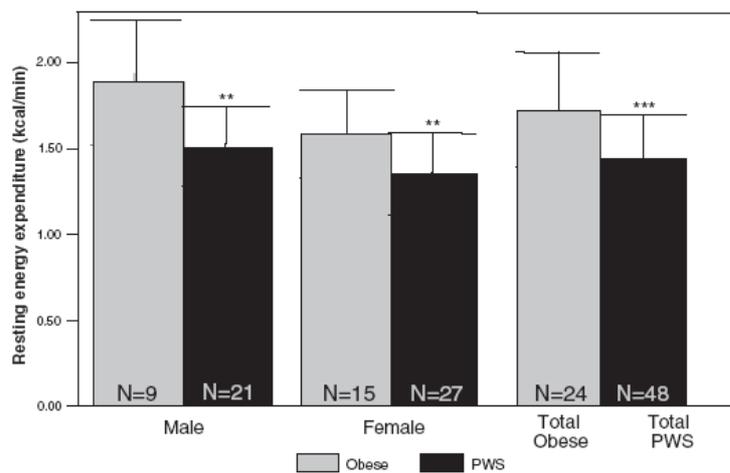
### The Genetic-Endocrinological Aspect



Weight problems represent a lifelong struggle for most PWS patients. Although the ensuing hyperphagia is considered to be the main symptom of PWS, there are many other factors that affect the behavior of PWS patients to the same extent.

#### *Abnormal Basal Metabolism <BMI>*

A study by Butler clearly shows that the energy expenditure at rest exhibited by people suffering from PWS is significantly lower than that in obese people. This results from a diminished basal metabolism, diminished activity and a diminished fat-free mass.



Resting Energy Expenditure (REE) significantly diminished by:

- Diminished metabolic rate
- Diminished activity
- Diminished FFM/FF Ratio

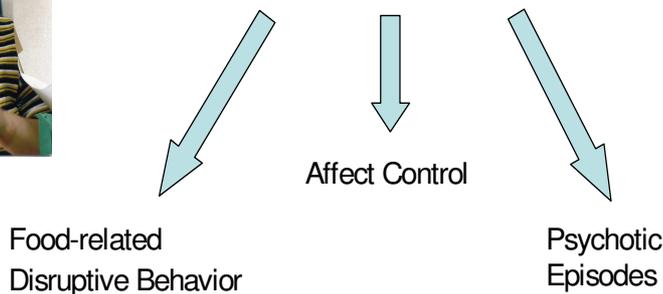
### Sleep-related respiratory problems

- pronounced snoring
- upper airway resistance syndrome (UARS)
- obstructive sleep apnea syndrome (OSAS) with hypoxaemia
- obstructive and central hypoventilation with consecutive hypercapnia
- (disturbances of sleep architecture with restless movements)
- daytime fatigue as a result of these malfunctions and other sequelae of OSAS

These disturbances are partly – but not exclusively! – due to obesity. In PWS, carbon dioxide provocation leads, for example, to a diminished ventilatory response, this in turn is ascribed to a malfunction of chemoreceptors. A heightened arousal threshold (more difficulty to wake up) in the presence of hypoxia and hypercapnia is associated with this situation. The result is an absence of a regular ventilatory response and evidence of impaired arousability.



### Behavioral Aspects in Prader-Willi Syndrome



### Behavioral correlates of these aspects

Obesity as well as the diminished resting energy expenditure and sleep disturbances all have an effect on the visible behavior of PWS patients. The disturbed sleeping patterns often cause fatigue during the day; the strong obesity curtails the patient's ease, desire and confidence to move around and the diminished resting energy expenditure makes it more difficult for the patient to lose weight through reduced calorie-intake alone.

PWS patients exhibit a number of behavioral patterns which are closely related to food, but which are not exclusively caused by genetic predisposition, such as:

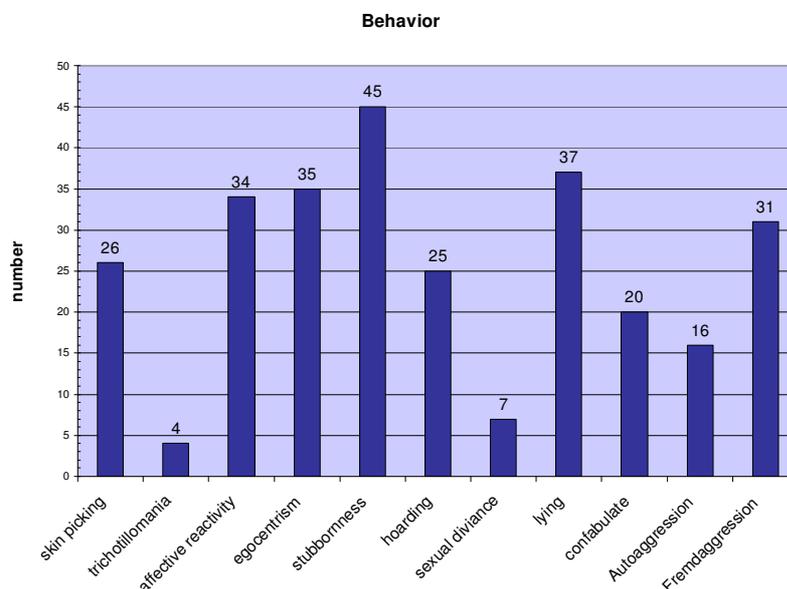
- Binge eating disorder
- Food foraging, hoarding and bargaining
- Illegal procurement of food
- Unusual food preference

We conducted two studies last year (2007) that demonstrate the behavioral aspects of PWS.

Affect Control: Everybody who has to work with PWS will be pushed to his or her own personal limits again and again due to the patient's deficiency in controlling his emotions. For this reason it is very important to know very well the resulting modes of behavior and understand them within the context in order to act and manage the situation correctly. Such modes of behavior are:

Temper tantrums	Ritualistic behavior
Strong mood changes	Compulsive and rigid behavior
Stubbornness	Obsessive behavior

There is a very wide range of these and other disruptive modes of behavior that differ as a function of the patient's cognitive abilities and genetic subtypes, as some studies have shown. Statistics provide evidence of the range of disruptive behavior in our institution:

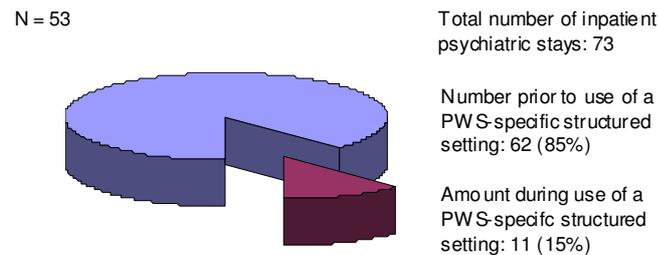


Psychotic Episodes and Requirement for In-patient Psychiatric Treatment:

Overall there seems to be a heightened predisposition to psychotic symptoms and psychotic episodes, especially in the case of UPD. The number of in-patient psychiatric hospitalizations can be an indication of whether the patient is vulnerable to these psychotic episodes. We can assume that stressors in the living and work environments play an important role in the onset of symptoms that require psychiatric treatment. We looked at the number of hospitalizations before and after the patient moved from the community to live in a PWS-specific setting. In a group of 53 PWS

patients, there were a total of 73 psychiatric inpatient hospitalizations: 22 PWS people did not have any stays for psychiatric treatment, 11 people had one, 8 people had two, 7 people had three, 2 people four, 2 people had five stays and one person even had seven hospital stays for in-patient psychiatric treatment. After the patient's admittance to a PWS-specific setting, only 11 (15%) of the in-patient psychiatric stays were necessary at our facility, and the reasons for those psychiatric hospitalizations were *not* PWS-specific in 85% of those cases.

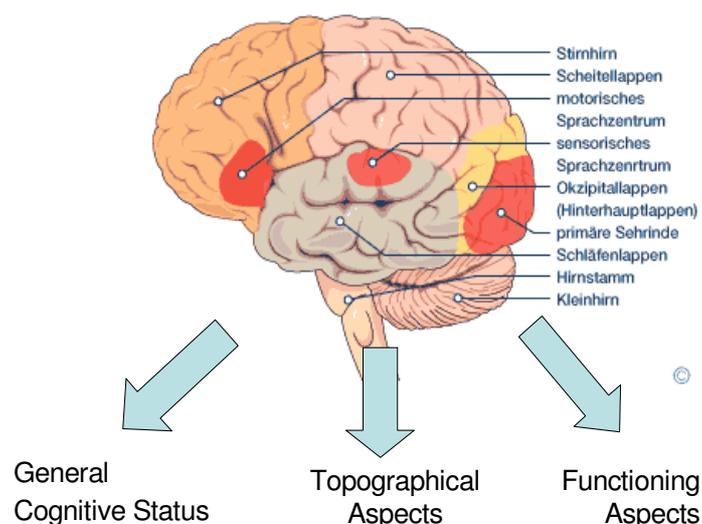
### Psychotic Episodes with in-patient stays:



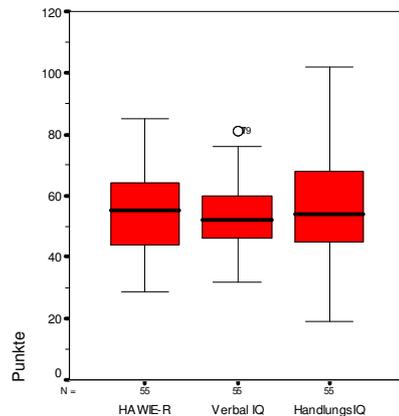
Stressors in the living and work environments have a negative effect on the psychiatric condition of the patient.

When neuropsychology looks at cognition, in addition to looking at overall intellectual performance which is always referenced to a normal population, it also looks at the regions and areas of the brain that are affected as well as at the content-related functions that are associated with them. We have used the Halstead-Reitan-Battery to test 50 people with PWS, and every subject was tested in several sessions.

### Cognitive Aspects



## General Cognitive Status Among 50 People with PWS:



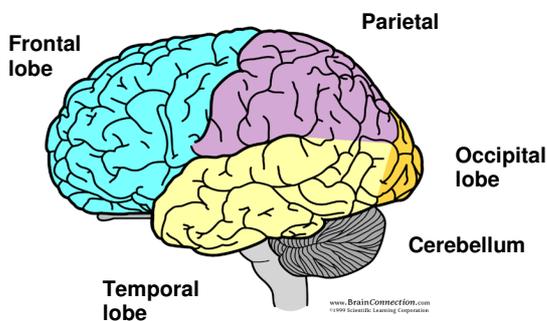
Total-IQ:  
Minimum: 29  
Maximum: 85  
Median: 55

Verbal-IQ:  
Minimum: 32  
Maximum: 81  
Median: 54

Performance-IQ:  
Minimum: 19  
Maximum: 102  
Median: 55

This corresponds to a level of intelligence of a light to moderate mental handicap. Overall we can assume that there is an impairment of the general brain functions. The sensitive neuropsychological procedures of Halstead-Reitan Test Battery also point to moderate to strong deficiencies and allow us to assume that evident functional deficiencies exist.

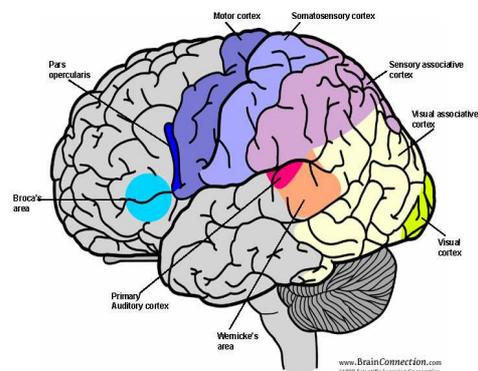
## Topographical Aspects



Brain research as well as data obtained from studies of clinical brain damage has shown that different regions in the brain specialize in different functions. Disturbances in specific areas strongly influence how our thought processes and behavior work.

Areas such as the Broca's language center or the Wernicke's language center are located on the left hemisphere of the brain. The visual cortex is located in the occipital lobes. The center for motor function lies in front of the central sulcus; the center for sensory perception lies behind it.

The question is whether specific areas are especially affected in the case of PWS patients.

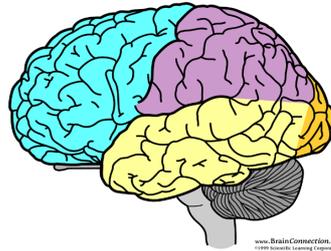


### **Prefrontal Cortex:**

- Reduced Performance in
- Trail Macking Test
  - Numbers-Symbol Test
  - Category Test

### **Motor and sensory cortex:**

- Pathognomonic signs in
- Tactile Form Recognition Test
  - Finger Oscillation Test (Bilateral)
  - Tactile Stimulation



### **Temporal Cortex:**

- Deficits in Speech Sounds Perception Test
- Suppressions in acoustic stimulation
- Problems with sequencing of actions (picture arrangement).

### **Hemisphere dominance:**

- No anomalies in
- Finger Tapping
  - Strength of Grip
  - Name Writing
  - Sensory Perceptual Exam

## **Motor and sensory cortex**

Sensory and motor breakdown of basic functions in the sense of pathognomonic signs can be detected through the Finger Oscillation test, the Tactile Form Recognition test and the Bilateral Tactile Stimulation test. It is necessary to take into account that in the Prader-Willi syndrome there is a lower sensitivity to pain, something that is also associated to skin-picking and to a problem that frequently arises such as pulling out of finger and toenails. When interpreting sensory anomalies it is advised to consider which part of the suppressions are possibly due to the peripheral and not to the central nervous system. Suppressions detected with the Finger Recognition, Bilateral Finger Recognition and Fingertip Number Writing tests point to functional disturbances in the region of the frontal area of the parietal region.

## **Temporal Cortex**

Suppressions in acoustic stimulation, impairments in Speech Sounds Perception Test and problems with sequencing actions - which is required in the Picture Arrangement (Bilderordnen) Subtest in HAWIE - can all point to an impairment of the temporal region. Great care should be taken when interpreting the failures appearing upon visual stimulation as evidence of a disturbance of the occipital region since a congenital nystagmus is frequently present in people suffering from the Prader-Willi syndrome and no anomalies can be established regarding the recognition of objects.

## **Pre-frontal Cortex**

The little differentiated naming abilities in the vocabulary test of the HAWIE suggest frontal damage. Performance in the Numbers-Symbol Test and in the Trail Making Test goes hand in hand with a speed component suggesting that areas in the anterior left hemisphere are affected.

The general slowdown exhibited in the Number Symbol Test and in Trail Making Test A, as well as reduced performances in processes that require coordination (Trail Making B and Tactual Performance Tests) and the high error rate in the Category Test that includes planning components, all point to pre-frontal damage. The pre-frontal brain damage is surely related to the difficulties that people with the Prader-

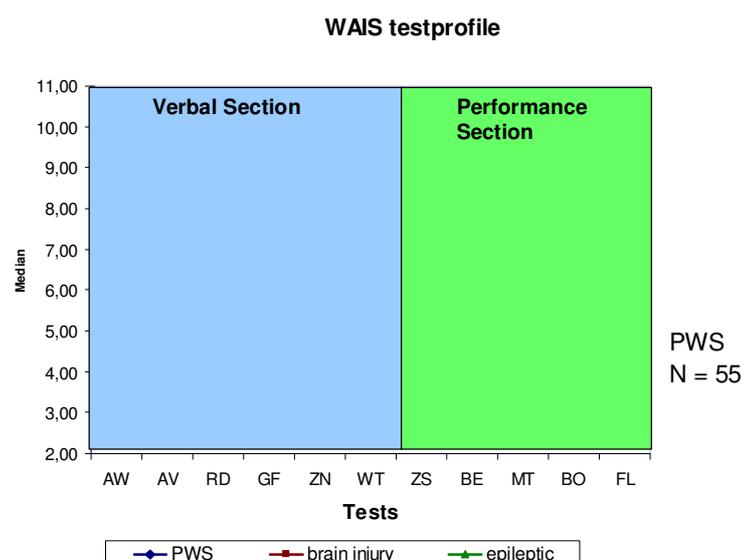
Willi syndrome experience to find their bearings in their social interaction with others and adapt to changing situations in their daily routine.

### Hemisphere preference

No indication of the dominance of one hemisphere over the other can be determined when we compare the procedures regarding hemisphere preference in the subtle tests of the Halstead Reitan Battery. These tests have been especially designed for this purpose and to calculate right-left differences. Such tests are Finger Tapping, Strength of Grip, Tactual Performance Test, Name Writing, Tactile Form Recognition, Tactile Finger Recognition, Finger-tip Number Writing, Sensory Perceptual Exam. Nor is there such indication of dominance when a difference between the verbal section and performance section of the WAIS is present. Overall we have to assume that there is damage in almost all regions of the brain. Focal damage can only be established on an individual basis.

The subtest profile of the 55 people with PWS shows a relatively homogenous pathway. A feature that stands out in the performance section is that there is a relative performance peak in the Picture Arrangement sub-test - something that is linked to the inclination patients have to do jigsaw puzzles. There is a wide spectrum of cognitive impairments, as can also be found in patients with brain damage. We know from brain research that the performance profiles of brain-damaged people differ from those shown by their normal control counterparts. The same is true of epileptics as a further etiological subgroup. The profile exhibited by patients with cranial-brain damage and epileptics differ from the profile of PWS test persons primarily regarding the height of the performance level. A striking feature and in great contrast to the comparison curves: there was no negative peak in the numbers-symbol test and the PWS test persons reached on average the highest score in the Picture Arrangement task. These results also coincide with statements in medical literature that report relative performance peaks in Picture Arrangement tasks.

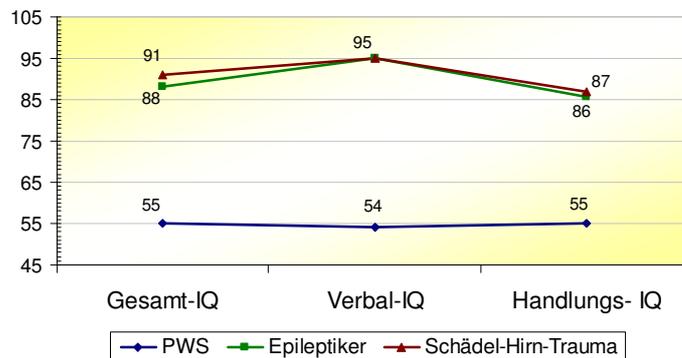
### Functioning aspects



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Verbal intelligence components are affected less by a neurological impairment than the non-verbal ones. This effect, however, cannot be observed in PWS test persons.

Obvious disturbances appear in non-verbal problem-solving tasks such as those in the Category Test that additionally requires cognitive flexibility.



A lack of flexibility and regulation of behavior was evidenced primarily in a tendency to perseverance. The test participants were not able to give up the patterns of problem-solving they had adopted and preferred to repeat tests rather than to face new tasks. In the daily routine, this behavioral pattern becomes evident, for example, at work, where people with the Prader-Willi syndrome often don't want to give up a task they have already become adept at.

An impairment in attention span tasks as well as shortcomings in memory also became manifest. Performance in the Stroop Test, in Trail Making Test A and B, and in the memory and localization components of the Tactual Performance Tests gave clear evidence of this.

In sum, we can say that people with PWS show signs of pronounced disturbances of perceptual organization, of disturbances of short- and long-term memory functions as well as disturbances in alertness and selective attentiveness. The more complex the tasks, the more strongly test performances suffered.

#### IV. Nature of the Problem

- Managing the tension between encouraging and supporting autonomy vs vulnerability of people with PWS
  - Eating behavior
  - Sexual behavior
  - Self-injury
  - Risk for exploitation and abuse
- People with PWS have unique neuropsychiatric vulnerabilities that put them at risk for maladaptive behaviors and mental health problems
  - Overeating and life threatening obesity
  - Repetitive and ritualistic behaviors
  - Increased liability to anxiety/mood disorder and psychotic illness
  - Stress sensitivity
  - Skin picking
  - Awareness for mental health problems
- People with PWS have unique neuropsychological strengths

- Long term memory
  - Visual processing
  - Shape discrimination
  - Attention for preferred activities
- People with PWS have unique neuropsychological strengths and vulnerabilities:
    - Short term memory deficits
    - Deficits in dichotic listening
    - Cognitive inflexibility
    - Fine motor deficits
    - Sensory perception/integration problems
    - Deficits in simultaneous processing
    - Poor time sense

## V. The Impact of the Problem on the:

- **Person:** difficulty with social relationships
- **Family:** need for support/information
- **Caregivers:** supervision, training, burn-out
- **Provider (structure and organization of care):** provide stable, life-long programmatic structure
- **System of care (medical, psychological therapeutics, education, legal):** need for informed decision making

## VI. Discussion:

The question we now have to ask is: what significance – if at all – do these neuropsychological aspects have in the day to day life and management of persons with PWS? There is almost no doubt that hypothalamic dysfunction causes a functional impairment of the brain since birth. But taking the neuroplasticity into account, we know that growth and differentiation of the brain is not only genetically but also epigenetically determined. The organic microstructures of the neocortex are mainly influenced by interactions with the environment. Genetically-induced maturation processes are combined with social shaping. Considering these aspects it seems necessary to initiate implicit learning processes on a pre-reflexive and preverbal level. Explicit learning processes verbally conveyed add to the development of selective attention and language. Genetically caused hypotonia especially disturbs early interactive experiences of a child with PWS and thus inhibits the development of prototypical experiences with others and causes a lack of "schemes of being-with" (Stern). Also explicit learning processes are affected due to the delay of acquisition of language. In my opinion it's absolutely imperative to offer support for a better development of motor skills in a social context which helps to improve the cognitive and communicative development. An important aspect of our work may be the impairment of executive control functions which are related to an impairment of the prefrontal cortex (PFC). As we know the maturation of the PFC starts rather late at the age of three and is completed not until the mid-adult life. This long lasting maturation process makes psychological and pedagogical interventions

possible in order to affect phenomena like egocentrism, deficient impulse control, inertness, tendency to depression. However, it's very important to intervene as early as possible to achieve the best effect. Of course, we can't eliminate genetic determination, but I am convinced that the score of impairment can be diminished. We've made the experience that also people in early adulthood can benefit from psychological and pedagogical interventions that particularly aim at attention and memory deficits, power of concentration and the difficulties of strategic and problem-solving thinking. The training works out best if it's embedded in everyday life situations. Memory for example can be trained by writing a diary, attentiveness can be increased with playing parlor games, and the ability to plan and to solve problems can be improved by creating a daily schedule or preparing a shopping list. Most of our men and women with PWS like to work with computer programs. These programs have to be playful and mistakes shouldn't have consequences in order to avoid frustration. Our people with PWS also like to communicate in writing putting an emphasis on the content and not the syntactic and orthographical correctness. To a certain extent tokens can be helpful, but - as we experienced - only if they refer to controllable and clearly defined tasks with not too high requirements. These are only a few examples for methods of improving cognitive functions. From a holistic view all interventions will influence cognitive, emotional and behavioral aspects of a person.

## VII. Guidelines for Best Practice:

Any discussion of guidelines for behaviour management in PWS must be informed by knowledge of the neurological, psychological and cognitive characteristics of people with PWS. As a group, **people with PWS have unique neuropsychological strengths and vulnerabilities**. For example, strengths include good long-term memory, visual processing, shape discrimination, attention and perseverance for preferred activities. On the other hand, there are vulnerabilities like overeating, stress sensitivity, obsessions and ritualistic behaviours, skin picking, emotional problems with tantrums, and an increased liability to mood and anxiety disorders, to psychotic illness and other mental health problems. Social relationships, therefore, are not very easy for staff and parents. The environment has to consider that people with PWS have short-term memory deficits, deficits in dichotic listening, fine motor deficits, sensory perception and integration problems, deficits in simultaneous and sequential processing, poor time sense, cognitive inflexibility and rigidity, and they have poor coping strategies.

**1. Appropriate food management:** Appropriate food management is not only essential for weight management but also helpful for mood and behavior hygiene. Management of food also requires management of money. ***When people with PWS have food security they are able to pursue other interests for a meaningful life.***

**2. Creation of a unique environment:** Due to the factor of stress sensitivity, a unique environment for people with PWS should be created to reduce stress and the risk of occurrence of problems through prevention by:

- food management (with food security and food control; food management is essential for weight management and is also the basis for mood and behaviour hygiene);

- proactive forward planning (a daily structure with interesting and meaningful activities);
- consistency;
- an informed, trained and counselled staff;
- persons to talk to, to support and assist;
- empathy;
- low expressed emotion and no discussions *during* an emotional crisis; and
- opportunity for relaxation.

**3. Access to appropriate care:** An early diagnosis of PWS is very important, and families must have access to adequate supports throughout the developmental period. An individual evaluation of mental health and other disease problems by a professional who has specific experience working with people who have PWS is essential. This includes an evaluation of the person's life experience from informed sources such as families and other caregivers as well as consideration of the person's developmental stage. Access to appropriate mental health care includes adequate therapy and/or psychotropic medication to be used together with the appropriate assistance. Compliance of the individual person with PWS is essential; it is helpful to get them involved in their individual daily plans and activities. Active listening is a key way for getting compliance.

**4. Behavioural programming:** Behaviour management presupposes an individual behaviour analysis taking into account predisposing, precipitating and perpetuating factors of individual PWS problems. Alternative reactions should be strengthened. A consistent and consequent environment is necessary; persons with PWS need an environment that offers certainty for them. Whenever possible the person with PWS should be involved in the behaviour management plan with regard to compliance to enhance empowerment. Contracts are helpful in this regard.

The use of contracts includes a consideration of the person's capacity and competency for making decisions. Some individuals may demonstrate the capacity to understand a specific concept, but they lack the judgment and perspective to apply this knowledge to their situation. One should always work with the individual to gain cooperation and *assent*. Family involvement can assist with decision making through *gentle persuasion*. If the person continues to refuse a plan that is essential for their well being, a formal evaluation of decision making capacity is required. Realize that competency can be in some life areas and not in others.

- Rubric for obtaining consent:
  - Work with individual to gain cooperation and consent.
  - Family involvement can assist decision making (gentle persuasion).
  - If refused, seek formal determination of capacity and competency, realizing that competency could be in some life areas and not in others.

The positive behaviour of a person with PWS should always be reinforced with attention. One should take into consideration *not* to use food as a reward, but use other forms of reinforcement like preferred activities, enjoyment of competition or items of reinforcement that you know the person with PWS likes. Pictures can be used to enhance learning. It is always essential to consider *their* point of view.

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First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workgroup A4: Fitness, Sport and Motivation

**I. Workgroup Leadership Team:**

**Presenter:** Georgina Loughnan

**Facilitator:** Dr Greg Cherpes

**Scribe:** Tiina Silvast

**Translator:** Dr Susanne Blichfelt

**II. Work group participants**

Most of the people who attended this workshop were caregivers. Other attendees included program supervisors and health care providers.

The number of years of experience working with people with PWS varied from a few months to several years.

Many attended to share their positive experiences with their clients' response to exercise and physical activity, where others were keen to learn the exercise needs of clients with PWS. Most attendees were hoping to gain knowledge in the motivational aspects of encouraging people with PWS to become healthy through physical activity.

**III. Presentation Abstract**

**Presenter:** Georgina Loughnan

**Title:** Fitness, Sports and Motivation for People with Prader-Willi Syndrome.

Prader-Willi syndrome (PWS) is a genetic disorder demonstrating hypothalamic malfunctioning. The collection of traits seen in the young client includes, low muscle tone, neonatal failure to thrive, delayed motor milestones followed by hyperphagia and lack of satiety, a mild to moderate intellectual disability and several hypothalamic disturbances including growth hormone deficiency. Adults with PWS will develop severe obesity if their lifestyle is not weight loss orientated. Co-morbidities resulting from overweight or obesity accentuate the congenital limitations, seen in people with PWS, preventing them from maintaining a healthy weight and good fitness. In order to maintain optimum fitness and prevent obesity in adults with PWS caloric restriction must be accompanied by appropriate physical activity or exercise. The benefits of exercise seen in the general population are paramount to people with PWS:

increased metabolic rate, increased fat free mass, improved circulation, improved mood, improved bone density and improved cardio vascular and respiratory health as well as the management of obesity.

The promotion of appropriate exercise is a “duty of care” to be provided by those in the role of caretaker. Although people with PWS may find the introduction of new exercise difficult to embrace, initially, after consistent insistence of the exercise program, the adult with PWS will accept the routine when positive results are recognized. Exercise needs to be seen as a non negotiable part of life that is commenced from the young of early adolescence.

Exercise in the young child with PWS is introduced as play and therapy based exercise for the improvement of muscle strength to assist with mobility and joint stability. It is also essential for the development of strong bones to prevent the lowering of bone mineral density that may be seen in the child who has been calorie-restricted. During adolescence it is essential to include regular effective exercise and not just focus on the maintenance of a healthy weight by calorie restriction.

In our public hospital PWS Clinic we have seen that, in adolescents and adults, effective exercise for weight loss and the reduction of co-morbidities must be done 5-6 days per week with appropriate supervision. Clients with home-based aerobic exercise equipment, who exercise 4-6 times per week lose significantly more weight and reduce their health risk significantly more than other clients who exercise less effectively ( $p < .0001$ ). The clients who achieve the greater results are those who participate in home-based, supervised exercise sessions.

Being involved in sport and social physical activity is important to promote an active lifestyle and provide motivation for the inclusion of activity in their lives but in most cases it will need to be accompanied by regular, formal, home-based exercise to maintain optimum health and weight.

Motivating the client with PWS may initially be very difficult, especially if they are overweight and are not used to being active. Persistence and regularity are essential as well as commencing activity/exercise at a level of intensity that they are physically capable of achieving. Exercising before meals is the preferred time as their meal may then be seen as the “reward” for their efforts. Carers who participate in an active lifestyle and will join the clients in active outings may contribute to the acceptance of activity, by the client. Motivation is often enhanced by weight loss success so charting weight loss can be beneficial as well as highlighting the improved tolerance to exercise and physical capabilities – for example: being able to tie their own shoes.

All clients with PWS will require motivation to do something that is new and may require some effort but once the activity is routine and consistent the ongoing motivating factor can be that of good health and success.

#### **IV. Define the nature of the problem and why it is important:**

**A.** Exercise is an essential element of care for people with PWS, from birth to old age, in order to improve and maintain physical fitness. It may take on the form of physical activity, ordered exercise or sports. Low muscle tone and strength, poor exercise tolerance and lack of motivation contribute to poor fitness and health.

Motivation, on the part of the person and their caregiver will always be necessary to maintain a desired level of fitness.

- Obesity is often problematic before an exercise program is initiated
- Individuals with PWS are often excluded from the social aspects of organised sports
- There has perhaps been a biased emphasis on food intake as a sole issue in weight control and pursuit of health
- As the population ages, the impact of exercise or lack of exercise is intensified. Significant morbidity is associated with a lack of physical fitness
- Motivation to participate in exercise activity may be problematic for individuals with PWS
  - Difficult to initiate
  - Low initial exercise tolerance levels
  - Body image
  - Potentially limited role models
  - Poor self-motivation
  - Dislike of something new or a change to routine

#### **B. Why is exercise important for people with PWS?**

- Exercise is important for health as well as weight control.
  - Muscle strength and joint protection
  - Cardio-respiratory efficiency
  - Increased metabolic rate
  - Bone density
  - HDL-cholesterol
  - Treatment of co-morbidities such as diabetes, hypertension, obstructive sleep apnoea, interstitial oedema
  - Enhances overall well-being

#### **V. Nature of the problem**

People with PWS are born with low muscle tone, less muscle bulk and strength and increased fat. Coupled with a lowered metabolic rate and caloric requirement as well as hyperphagia they are at risk of developing severe obesity. With less stable joints and a reduced exercise tolerance they are also at risk of developing co-morbidities associated with lack of physical fitness.

## **VI. Impact of the Problem**

### **A. Impact of the problem on the individual**

- Lack of physical fitness can contribute to a significant impairment in the individual's quality of life.
- An individual with PWS may be resistant to participating in an exercise program.
- The individual is able to recognize the health benefits after the exercise program is underway.
- A positive impact may be derived from the presence of structured activity.
- Decreased morbidity and increased health throughout the lifespan.
- Increased longevity.
- Potential accessibility issues in home, workplace, and social experiences.

### **B. Impact of the problem on the Family**

- Worldwide, many individuals with PWS live with family members.
- The need for exercise is evident in early life, but may not be immediately apparent in childhood and adolescence and needs to be stressed by the health care provider.
- Limited resources within the home to enforce an exercise program
  - Dealing with arguments, tantrums, possibly acting out.
  - Difficult to implement a program with the regularity that is required

### **C. Impact of the problem on the caregiver**

- Level of fitness and attitude toward exercise of the care-giver may play a role.
- Motivation
- Exercise should be supervised by the care-giver
- Care-giver to encourage a physically active lifestyle by participating with the client whenever possible
- An active caregiver encourages more incidental activity.

### **D. Impact of the problem on the Provider**

- Some group homes are structured in ways which make it difficult for an individual to have time and/or the energy to engage in exercise.
- Belief that structure exercise program applies to individuals with PWS only.
- Lack of specialized group homes in many countries.
- Physical activity is a prescription for life for anyone with PWS – not an optional extra

### **E. Impact of the problem on the System of Care**

- Significant cost associated with treating obesity related illnesses
- Significant loss of productivity due to illness and decreased physical capacity.
- Significant cost associated with the need for weight-appropriate equipment.
- Meeting occupational health and safety requirements.
  - Hospitals, clinics, workplace, home, vehicles

## VII. Discussion

- Physical fitness is essential for an improved quality of life for people with PWS
- Sports, fitness and motivation are the responsibility of caregivers for people with PWS
- Fitness can be achieved through sports and structured exercise provided the sessions are regular and effective
- Caregivers' participation in sports and exercise is motivational for the person with PWS
- Sports and fitness has an obvious positive effect on behaviour of people with PWS.

## VIII. Guidelines for Best Practice

- Essential effective exercise:
  - 5 to 6 days per week
  - 30 to 40 minutes continuous effort
- An exercise routine is important to begin *before* the onset of severe morbid obesity.
- Routine part of day.
- Participation by an enthusiastic caregiver.
- Individualize the motivations
- Scheduling exercise session prior to a meal or snack.
- Activity and sports are in addition to essential effective exercise.
- Pre-exercise assessment by physiotherapist.
  - Cardio assessment, if indicated.
- Design of plan
  - Discuss issues of the plan with the individual in advance.
  - Build in expectation of increasing intensity.
  - Use frequent praise and provide encouragement.
  - 3-5 minutes “warm up” before and “cool down” after exercise
- Initial exercise
  - Based on current ability
  - Minimum of 10 minutes of continual activity (walking).
- Gradual increase in intensity of effective exercise.
- Home-based exercise may be the most effective way of ensuring consistent, effective participation:
  - Treadmill
  - Exercise bike
  - DVD
  - Dance
- Upper body strengthening exercises
- Enthusiastic care-giver
- Fitness record
  - Diary, photographs, weight, waist measurement
- Availability of companion/participant

- Positive reinforcers
- Increased incidental activity
  - House responsibilities / own room
  - Park car further away from destination
  - Use stairs
  - Walk the long way around
  - Active staff make active clients
- Points to remember
  - Good hydration of client
  - drink water before and after exercise (but not to excess)
  - High pain threshold
  - do not let clients exercise when unwell
  - do not allow clients to “overexercise” (too long or too intensely)
  - Be aware of injuries
  - clients with PWS are prone to having hyper-mobile joints / lax ligaments

## **IX. Conclusion**

The work group members represented viewpoints from varied nations as well as disciplines. The importance of exercise in the life of an individual with PWS was universally endorsed. The difficulty in motivating individuals with PWS to begin or maintain a lifestyle which includes effective exercise as a matter of routine was also identified by all group members. Although there was variability among group members regarding the best use of creative strategies to motivate individuals with PWS, the work group shared a common respect for the importance of gaining an understanding of the person with PWS as a unique individual. Through this type of understanding, the development of a therapeutic alliance with an individual with PWS becomes possible. This alliance provides the opportunity to be a key motivational factor in the pursuit of health. The work group concluded with an enthusiastic acknowledgment the importance of this topic and looks forward to ongoing discussion and refinement of the points discussed above.

## **X. Challenges and items for future consideration**

- Specific research into the physiological effects of exercise on individuals with PWS
- Development of minimal standards of available equipment within the residences of individuals with PWS
- Recognition of protected time for exercise as an element of the duty of care of all people with PWS

QED

First International Caretaker's Conference  
Herne Germany  
June 3 – 5, 2008

Workgroup S1: Training for Teachers

**I Workgroup Leadership Team:**

**Presenter:** Barbara J. Goff, Ed. D., United States of America

**Facilitator:** Linda Thornton, New Zealand

**Scribe:** Jeff Covington, United States of America

**Translator:** Monika Fuhrmann, Germany

**II Workgroup participants:** The workgroup that developed this narrative was very diverse. It consisted thirteen members; eight from Germany, two from the United States, one from Denmark, one from the United Kingdom, and one from New Zealand. There were six teachers, three parents, one special education professor, one administrator, one adult caregiver, and two trainers.

**III Presenter:** BJ Goff

**Title:** PWS for Educators

**Part A. Overview of the syndrome:**

***Medical Overview:***

- Hypotonia
- Hyperphagia
- Obesity related issues
  - Glucose intolerance and diabetes
  - Hypertension
  - Cardiopulmonary compromise
  - Stress on vascular and skeletal system
  - Sleep disturbances
- Scoliosis
- Bruising
- Lack of vomiting
- Dental Caries
- Strabismus
- Incomplete onset of puberty
- High pain threshold
- Osteoporosis

- Altered temperature regulation
- Gastric Dilation

***Physical Characteristics:***

- Almond-shaped eyes
- Triangular-shaped mouth/thin upper lip
- Short stature\*
- Narrow forehead
- Underdeveloped sex organs\*
- Underdeveloped secondary sex characteristics\*

\*Growth hormone therapy will affect these characteristics

***Behavioral Overview:***

- Hyperphagia and Food Related Behaviors
  - Insatiable appetite
  - Preoccupation with food
  - Food seeking/foraging
  - Sneaking, hiding, hoarding
  - Eating unusual food or food-related items
  - Illegal activities related to food acquisition
- Self-injurious Behavior
  - Skin picking
  - Nose picking
  - Head banging
  - Rectal picking/digging
  - Hair pulling
- Anxiety
  - Manifests in younger children due to frustration over being misunderstood
  - Notable increase in adolescence
  - Sources of anxiety include:
    - Access to food
    - Change
    - Unrealistic cognitive expectations
    - Unrealistic behavioral expectations
    - Lack of communication: inability to understand or be understood
    - Internalizing things not easily understood (especially grief)
- Obsessive Thinking and Perseveration
  - Need reassurance about routine and expectations
  - Need specific reassurance about changes around food
  - Obsessive thoughts may result in unpredictable explosive episodes

- Compulsive Behaviors
  - Hoarding
  - Possessiveness
  - Need to know, tell, or ask
  - Repeating rituals
  - Ordering and arranging
  - Concern with cleanliness, symmetry, and exactness
  
- Transitions
  - Provide warning time for transition
  - Provide written or picture schedules
  - Allow time for an activity to be completed
  - Practice scenarios of change
  
- Change
  - Forewarn student (and parents if necessary) of any change to schedule
  - Provide Plan A and Plan B
  
- Concrete and Rigid Thinking
  - No “outside the box” thinking
  - Once something is said, it is the truth
  - Limited ability to abstract
  - Sarcastic humor can be misunderstood
  - People have specific roles that are not interchangeable
  
- Perceived Unfairness
  - The alleged unfairness may or may not be legitimate in everyone else’s view, but is experienced as such by the person with PWS.
  
- Excessive Sleepiness
  - Sleep study should be conducted
  - 50% of individuals with PWS have been diagnosed with sleep disturbance
  
- Lying and Stealing (food, money, personal possessions)
  - Most lies and thefts involve food or access to food
  - Allegations of abuse may be food related or in retribution
  - Grandiosity is not uncommon
  - If the opportunity exists for stealing food or money, the student is being set up to fail
  
- Running Away (elopement)
  - Student is angry
  - Student is hungry

- Student wants to get out of existing situation
- Student wants an issue resolved and is seeking higher authority
- Student wants to go home
- Student is seeking attention
- Tenuous Emotional Control
  - Responses can be out of proportion to situation
  - Misunderstandings can quickly escalate leaving no opportunity for typical interventions or preventative strategies

## **Part B. PWS Student in the Classroom**

Here are some *typical behaviors* of students with PWS you may see in your classroom:

- Refuses to put away materials and move to next subject
- Suddenly gets up from desk and walks out of the classroom
- Keeps interrupting with the same question or comment
- Refuses to participate in activity or assignment
- Has verbal and/or physical outburst as reaction to schedule/routine change
- Refuses to take direction
- Sleepiness
- Skin-picking
- Removes clothing (appears to be attention seeking)
- Incontinence (particularly when stressed, over-anxious or confrontational)

### ***Learning Profile:***

- Relative Learning Strengths
  - Good long-term memory skills
  - Receptive language
  - Good at puzzles
  - Visual processing
  - Basic math skills
  - Reading skills
  - Social and friendly
  - Average IQ = 70 (mild-range of intellectual disability)
- Learning Weaknesses
  - Expressive language
  - Poor gross motor skills (including balancing skills)
  - Sometimes poor fine motor skills
  - Sequential processing deficit
  - Difficulty with abstract concepts
  - Poor short term memory

Learning and social gaps tend to widen during grades 3-5 (ages 8-10). The shift from concrete to abstract learning generally finds the student with PWS falling behind in most academic areas. As educators so well know, "...from kindergarten to fourth grade, students are learning to read; from fourth grade on, they are reading to learn."

Socially, typical classmates are maturing and becoming increasingly involved in more sophisticated peer relationships and activities. Appearances and peer approval become paramount leaving the student with PWS at a disadvantage in establishing friendships.

***Practical Tips:***

- Visual schedules
- Verbal reminders
- Plan A and B
- Once things are set in motion, it is difficult to be flexible
- No more than two-step directions
- Shortened assignments
- Controlled access to food and money
- Answer questions up to two times and then write it down if necessary
- Brief or no homework assignments
- Ask – don't demand
- Extra travel and set up time
- Keep tissues and adhesive bandages on hand
- Provide opportunities for student to work alone or in pairs
- Social Skills Program
- Provide opportunities for physical activity
- Empower all who are involved with the student to be an authority
- Communicate and write down consequences of food stealing
- Minimize discussion when student is upset
- Use a mirror to recognize and express emotions
- Use social stories to teach "life skills"
- Use flash cards and/or picture symbols to communicate wants, needs, and transitions
- Reciprocal communication between home and school when changes occur
- Always remember: Mom Rules!

***Given the significant role of hyperphagia and anxiety in the daily life of a person with PWS; these areas need to be addressed in very specific ways.***

## ***Hyperphagia:***

As we know, food-seeking, acquisition, stealing, and hoarding are common characteristics of individuals with PWS. Food is everywhere at school. Here are some possible sources:

- Teachers' desks
- Students' desks
- Backpacks
- Snacks in classroom closets
- Lunches
- Trash
- Programs and outings
- Edible reinforcers
- Cafeteria
- Unlocked lockers
- Coat pockets
- Vending machines
- Bake/Candy Sales, cooking programs, fundraisers
- Meeting rooms
- Staff offices
- Special events
- Bus drivers
- Friends

Each of these food sources must be carefully and consistently controlled for the student with PWS.

## ***Anxiety:***

Anxiety is a key concept to understanding the student with Prader-Willi syndrome. Many of the challenging behaviors displayed by these students are anxiety related.

Typical anxiety-producing situations include:

- Transition times
- Who do they go to for what
- Not sure what is coming next
- Changes in food menu or times
- Perceived unfairness from other students or adults
- Teasing/Bullying
- Unclear communication or instructions

Therefore it is important to do the following:

- Post a schedule for the student
- Give a warning when a transition is to occur

- Make clear who is to address their particular needs
- Communicate changes in food presentation, food related activities, or meal/snack times
- Communicate PWS needs to other classmates as appropriate

To best understand the student with PWS, it is good to look at the positive and negative characteristics in general and some of the influences as to why those students move from positive to negative traits:

<u>Positive</u>	<u>Negative</u>	<u>Influences</u>
Sense of Humor	Furious	No structure
Communicative	Aggressive	Out of control
Pragmatic	Cruel	Perception of unfairness
Common sense	Unfair	Emotional
Ideas on what to do	Other person is bad	Reaction to emotions
Dependable	Swearing	Hungry
Kind	Violent	Frustrated
Good memory	Repetitive talking	Anxious
Good worker	Mood swings	Panic
Word puzzles	Emotional	Overtaxed
Puzzles		Poor communication
Computer		
Charming		
“Stick-to-it-ness”		
Likes animals		
Likes children		
Bright		

As is evident, there are many positive characteristics of the student with PWS. There are also some influences that can turn those positive characteristic into negative ones in short order. The role of the educator is to focus on those positive traits and provide an environment that is conducive to reducing those influences and stimuli that will have a negative impact on the student.

#### **IV Nature of the Problem**

Prader-Willi syndrome is a low incidence disorder; as such it is unlikely that a teacher, even one trained to work with children with special needs, will encounter a student with the syndrome in their career. Given limited school funds and the demands of more common disability groups, training in working with students with PWS is generally not a priority.

Subsequently, few targeted resources exist and there is little emphasis on teacher training. Families of children with PWS find it quite disheartening to see their children enter a school system that is ill-prepared to teach them. These families find themselves in the role of primary resource for all matters concerning

their child's disability and his/her special needs. Moreover they find that, given the uniqueness of the syndrome, especially the behavioral challenges, their input is often questioned or even discarded in lieu of more traditional teaching and behavioral strategies which are often ineffective.

There is a need for families and teachers to have a shared knowledge and understanding of PWS so that effective communication and collaboration on behalf of the student can develop. It was the mission of this workgroup to establish content guidelines for the development of a "Best Practices" manual for educators.

Whenever a teacher feels unprepared, and a family feels unsure and distrustful of the quality of education their child will receive, the child will be poorly served. It is easy to misunderstand the abilities of a child with PWS, and to misinterpret their behaviors, as PWS is such a complex, multilayered syndrome. Such misunderstanding has resulted in unrealistic academic expectations and inappropriate behavior management strategies. Under these circumstances the student with PWS may become increasingly unmanageable, resulting in suspensions, alternate school placements, and more problems at home.

The families, when able, generally provide whatever information, training, and support they have available to school personnel. Sometimes it is well-received, other times it is seen as only the parent's perspective and not a realistic and honest representation of the child. In many countries written information and audio-visual aids are simply non-existent, so educators and parents are without a common ground of understanding from which to develop an appropriate educational program. Students with PWS can be quite challenging in an educational environment; yet those challenges can be addressed when there is a shared knowledge and understanding of PWS and how it is manifested by a particular student.

Toward that end, the following guidelines constitute our consensus on essential information for educators. It is our hope that each country will utilize these guidelines as a framework to build their own training program and resources appropriate to their school system and educational practices.

## **V Impact of the problem on parents and teachers**

A training curriculum on Prader-Willi syndrome will provide a general overview of PWS, which is an essential first step. However, both educators and parents recognize that each child, and therefore each family, is unique. First, each family exists within a culture and a language, as well as, a belief system about, and experience with, education. In addition, each child has their own personality characteristics, idiosyncrasies, personal needs and wants that must be known to the teacher. A sharing of information specific to the child must occur on an on-going basis. Some examples of important areas of discussion include:

- Those behavior problems associated with PWS and those that are not
- Physical, cognitive, and emotional strengths and weaknesses
- Individual learning profile
- Consistency of rules between school and home
- Student's vulnerability to manipulation by others
- The student's behavior triggers and how can they be avoided
- The how's of routine communication between school and family
- Changes at home that may affect behavior at school
- Handling of holidays and other special occasions within the classroom
- Medical history and current concerns
- Intensity of the food seeking behavior
- Developmental milestones
- Positive traits of the student
- Ensuring equal opportunity to experience the fullness of school life
- Need for additional support staff throughout the school day
- Agreed upon consequences for food stealing
- Support and resources needed outside of school

## **VI Impact of the problem on the educational setting:**

An unanswered question is "What type of educational environment is best suited to a student with Prader-Willi syndrome?" Educational programs and services for individuals with disabilities vary by country, so we cannot recommend any specific educational model. Individual characteristics of the student must be considered in determining an appropriate educational program in accordance with the policies, practices, and resources of the country and specific school system.

Each school must balance the needs of the individual and what is the most viable option for keeping them in school. Some typical placement options include:

- Integrated pre-school—children up to age five who are educated with typical children
- Full inclusion—students are educated with typical, same-age peers
- Resource room—take student out of general education classroom for specific skill or subject matter tutoring
- Substantially separate—students are educated within a school with typical peers, but in a separate classroom with other students who have special needs
- Special school for students with learning and other disabilities
- Residential school for students with special needs

## **VII Impact of the problem on the system of services and supports:**

The main responsibility of a school system is to provide services to all students as they are needed. Due to psychological and physiological differences, students with PWS are often in need of related support services and therapies. These include:

- One-to-one aide or paraprofessional
- Speech Therapy
- Occupational Therapy
- Physical Therapy
- Behavior management
- Nursing
- Dietary consultation
- Adaptive physical education
- Social work
- Psychology
- Socialization skills
- Counselor

## **VIII Impact of the problem on the community**

***Transition Planning:*** A key component to providing services to students with special needs is the transition plan from secondary education to adult life. A secondary education program should be designed to not only expand and enhance academic skills, but expose students to a variety of realistic vocational options with a focus on successful work habits and behaviors.

At a young age children may not be obese and parents do not have to use such strict environmental controls. However, once the student reaches adolescence, food-seeking behaviors and anxiety around food become more prevalent. This must be taken into consideration in determining vocational placement options, as the student with PWS may require more intense supports in certain situations.

To ensure a smooth transition from school years to adult life, schools should provide the student with a personalized program that offers essential services to meet the individual's needs as they navigate through this major life transition. While the school may not provide these services directly, they need to identify community or government resources that do, including:

- Case Management: support, resources, and advocacy
- Vocational Services, including a program of work experience while at school
- Residential Services
- Recreation and Leisure Services

- Transportation Services

Best practice for the person with Prader-Willi syndrome is to have 24 hour / 7 day week supervision whether that is in a residential living situation, at home with family, in educational or vocational programming or on a community work detail.

Much like choosing an educational setting, finding a meaningful day activity must be a very individualized process meeting the unique needs of the particular person.

Regardless of the living situation, the individual needs assistance in finding a productive and meaningful day activity. Many individuals with PWS are able to be competitively employed with natural supports and a job coach. It may be best to have one or even two part-time jobs. Individuals with PWS have demonstrated success in the following jobs:

- Clerical
- Assembly
- Cleaning
- Childcare
- Animal care
- Crafts
- Landscape Maintenance

Many individuals with PWS are motivated by money and responsibility. But, any money earned must be closely managed. The most common reasons for job loss are food or behavior related problems. Sheltered workshops provide the close supervision needed by some individuals and are good for their opportunities for socialization, but not for challenging or consistent work. Work experience offered at school, or day habilitation programs offered by a community-based service provider, can provide volunteer opportunities, educational opportunities, socialization, recreation, and supervision. However, no pay can be earned and it is often not challenging enough. Length of workday or workweek at a job site where there is an opportunity to obtain food should be carefully considered before putting the individual in a potentially stressful work situation. Transportation must be monitored for food-seeking opportunities.

It is important that teachers, therapists, and paraprofessionals empathize with the student and their family as much as possible. It is not easy to have PWS, or to support someone who has it, yet families, educators, service providers, and the individuals themselves are discovering (and inventing!) more and more opportunities for learning, living and working in their communities.

## **IX Conclusion:**

This outline is a compilation of areas that teachers feel they should know about and what parents feel teachers should know about. With such a comprehensive training approach for the educators, families can feel assured that their child's

teacher is well versed in Prader-Willi syndrome and has the tools necessary to make the environment as safe and conducive to learning as they can for the student with PWS.

We offer these educational training guidelines as a framework for your own creative work in educating individuals with Prader-Willi syndrome.

QED

First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workshop B1: Nutrition

**I. Workgroup Leadership Team:**

**Presenters:** Glenn Berall, MD and Karen Balko RD

**Facilitator:** Dr Constanze Lämmer – Germany

**Scribe:** Dr Susanne Blichfeldt – Denmark

**Translator:** Herr Schilder– Germany

**II. Work group participants:**

- International workgroup with representation from Germany, USA, UK, Finland, Sweden, Denmark
- Care-givers from group homes, medical doctors, family assistants

**III. Presentation Abstract:**

**Presenters:** DVD by Glenn Berall, MD and Karen Balko RD, Canada

**Title:** Nutrition Management in PWS

Dr Glenn Berall and Ms Karen Balko were invited speakers for this workshop. They were not able to come to present in person, so they sent a DVD that was presented to the participants at the workshop. They focused on management of the diet in PWS using the “RED-YELLOW-GREEN” system:

- RED: Food to be eaten rarely or never;
- YELLOW: Food, carefully included in the diet in calculated amounts;
- GREEN: Low calorie foods that can be served with less restriction in the PWS diet (see below).

With this system the different food items are divided into color-coded categories based upon their caloric content. This model can make it easier for families and caregivers to manage and administer food. It is also a useful to educate the person with PWS about their diet.

Important points from Dr Glenn Berall’s presentation:

- Without help and treatment the hyperphagia in PWS will cause severe obesity and morbidity (diabetes, heart disease, liver disease etc.)
- Healthy food combined with physical activity is essential.
- Food supplements as fatty acids, iron, calcium and vitamins are often needed.
- Access to kitchens and food should be avoided.
- Education of all involved staff (home and at work) and family is a must.
- Consistency prevents anxiety and outbursts.

The ideal weight depends on the persons muscle mass and level of activity and can vary between persons of the same height.

Important points from Karen Balko's presentation:

Using the RED- YELLOW - GREEN SYSTEM makes food security easier:

- RED: Can be fried meat, fat, chocolate, etc.
- YELLOW: Essential food but to be served in fixed amounts per serving. Examples are bread, meat, milk, and fruit. Bread, milk and meat can vary concerning the amount of calories per gram, as the amount of fat per gram can vary a lot.
- GREEN: food with 25 kcal or less per 100 grams. This includes all vegetables. These can be served at all meals and give more volume.

How many calories? Individual needs vary a lot. If weight loss is wanted, start with 7-9 kcal/cm height, and follow the weight, as the needs for each individual can be less. If stable weight is wanted: start with 9-10 kcal/cm height: example: 1500 kcal/day for a person being 150cm.

From Dr Constanze Lämmer's plenary talk about diet and nutrition in PWS:

Even with a low body mass index (BMI),\* adults with PWS can have a higher amount of body fat than typical persons. So, examination of body composition together with BMI is important when estimating the nutritional needs for a person with PWS.

A medical evaluation together with the dietician is essential. Special diets are not recommended. Low-glycemic-index carbohydrates are preferred; they prevent hyperinsulinemia and make weight reduction possible. The diet should contain 2-2.5 liters of free-caloric drinks per day. In most cases (adult) daily supplements of calcium (tablet) vitamin D, iron and fatty acids are recommended and are calculated by the dietician.

From the work in our group:

- In the group discussion we talked about the food and especially the many challenges in how to provide information to everyone involved in the care

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\* Body mass index is determined by assessing the ratio between body weight (kg) and height (m<sup>2</sup>); it is the best way to estimate body fat percentage. For most of us, a BMI > 25 indicates overweight.

- if the person with PWS and how to manage the diet administration in daily life.
- We focused on “food security” (“no hope, no doubt and no disappointment”) and the need of calorie management for weight loss, stabilizing weight or gaining weight.
  - We talked about special conditions in PWS that makes the diet different from other persons (low calorie needs and the reasons for this).
  - The diet should be individualized.
  - In conclusion we find that guidelines or recommendations are needed both about the food itself and about daily management. Communication between all the involved persons is essential and includes the person with PWS, the family, the caregivers, the staff at a group home, and the system of care in general, e.g. work and school placement.

## **II. Nature of the problem:**

- People with PWS need a special, lifelong diet. The calorie needs are based upon their body composition, BMI, and their daily physical activity.
- Even though most adults with PWS need a low calorie diet administered by others, there are considerable individual variations in calorie needs.
- Management of different meal sizes in groups of people with PWS can be difficult.
- Costs for healthy food rich in protein and vitamins and also low in fat are higher than typical food, and availability of some vegetables is often seasonal.
- Staff is often not educated enough about nutrition.
- A dietician is often not available, at least not on a daily or weekly basis.
- There can be a lack of communication between the people who buy and prepare the food, and those who know the diet recommendations for people with PWS.
- People with PWS are not able to manage the diet by themselves.
- Preventing overweight and or malnutrition is one of the most life saving treatments in PWS.

## **III. Special situations for people living in group homes**

- Sometimes there are very big differences between the way the diet is administered in the group home and in the family, when the person with PWS is on “home visits.”
- Weight gain that is seen after family visits and during holidays is not always talked about with the family.
- Sometimes the person’s weight goes up and down many times during the year.
- It is reported to be difficult to manage different meal sizes in the group homes. This means that people living there are at risk of either overweight

or underweight or malnutrition if all have the same amount of food every day.

#### **IV. Impact of the problem in the person with PWS**

- Many people with PWS need more information about their own disorder and about their diet.
- They need to be listened to about their concerns about food, and their special food preferences (taste) should be taken in consideration when the food is prepared.
- They need to know that they will be taken care of.
- They need to be aware about what is decided for him/her; they need to know that their needs will be met.
- It is important to feel safe about the food.
- The person with PWS feels safe when there is good communication, contact and agreement between staff and family.
- From a medical point of view, the person is in a great health risk if overweight is not prevented or treated.
- Registration of weight (daily or weekly) is recommended
- Weight loss too quickly can be dangerous for the body.
- The up and down fluctuation of weight is a health risk problem.
- The person with PWS might feel guilty if their weight goes up.
- Overeating at holidays can be dangerous (risk for stomach rupture).
- Plans for managing food over the holidays can be helpful for everyone.

#### **V. Impact of the problem on the provider**

- The staff is responsible for the quality of the food. Planned meals (menu) are recommended.
- We propose workshops for providers where the focus is on the food in PWS, on recipes and how to prepare the food.
- We propose that one of the staff is the responsible for regular contact to the dietician, for getting new information about PWS and diet and for giving this information to staff.
- We propose that one of the staff plan the menus and also get more education about PWS and diet (most optimal a person with special education in dietary management.)
- Also needed are weekly meetings for the staff also about the diet for PWS and other health issues.
- There must be time allotted for the staff to learn about food!
- Continuous education is needed; education of new staff is especially important.
- Sometimes the cost of the food for the management of the diet in PWS exceeds the budget when prices are higher than expected.
- The staff can educate the parents.

## VI. Impact of the problem on the family

- The family needs ongoing education about the needs of their child or adult child.
- Many families feel helpless about the situation at home and have lost the control of the diet.
- There should be a contract with a dietician who also works with the family with an adult who does no longer live with the parents.
- There is a need to plan daily activities during home visits that take away the “focus on food”.
- Older parents can have very little energy. The staff can help them by giving exact information about what the person with PWS can eat on home visits: if possible, also arrange meals to be served in the parents’ homes.

## VII. Guidelines for Best Practice

- A person with PWS needs a lifelong diet that is individually adjusted based upon body composition, BMI and daily activity.
- Families/staff must have detailed information about:
  - Food items
  - Daily management of food
- Good management means that there must be good communication between all of the following people involved in the care of the person with PWS:
  - The person with PWS
  - The family
  - The caregiver
  - The system of care in general
  - Regular meetings between staff and family
  - Regular feed back from dietician to family
- The use of the system of the RED – YELLOW – GREEN diet (food divided into special categories) can make the administration of food easier for people with PWS, their families and/or caregivers.
- Food is not a reward and not a punishment.
- Fix time of meals by sequence of activities.
- Use rituals to introduce expected behavior; for example, drink water before starting a meal.
- Write rules down on a board.
- The importance of **food security must be stressed.**
- **An individualized diet and ongoing education** of all people involved in the care of the person with PWS is required.

QED

First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workgroup B2: Crisis Management

**I. Workgroup Leadership Team:**

**Presenters:**

Norbert Hoedebeck-Stuntebeck

Mary K. Ziccardi

Dr. Greg Cherpes

**Facilitator:** Dr. Janice Forster

**Scribe:** Evan Farrar

**II. Work group participants:**

The majority of the individuals who attended this workshop were caregivers; there were 6 managers/administrators, 2 teachers or workshop instructors, and 2 professionals. Their level of experience with persons who have PWS was as follows:

Years of Experience	<1	<5	5-10	10-15	>15
Workshop respondents	4	5	7	1	2

Their expectations were to learn/share information about how a crisis evolves, how to manage it, how to prevent/ avoid it. They also wanted to understand the nature of the settings in which crises occur, the standards for group home practice, and how to avoid physical restraint, if possible.

The workshop attendees had different attitudes, knowledge and skills reflecting the context and level of experience that they had working with persons with PWS. They also represented different nations with unique standards or regulations for the care of developmentally disabled individuals.

In the course of discussion, it became apparent that each individual's understanding of a crisis was personalized based upon his or her unique experience. Some examples of crisis behavior reflected typical PWS behavior problems in general and did not define the leadership's experience of critical incidents.

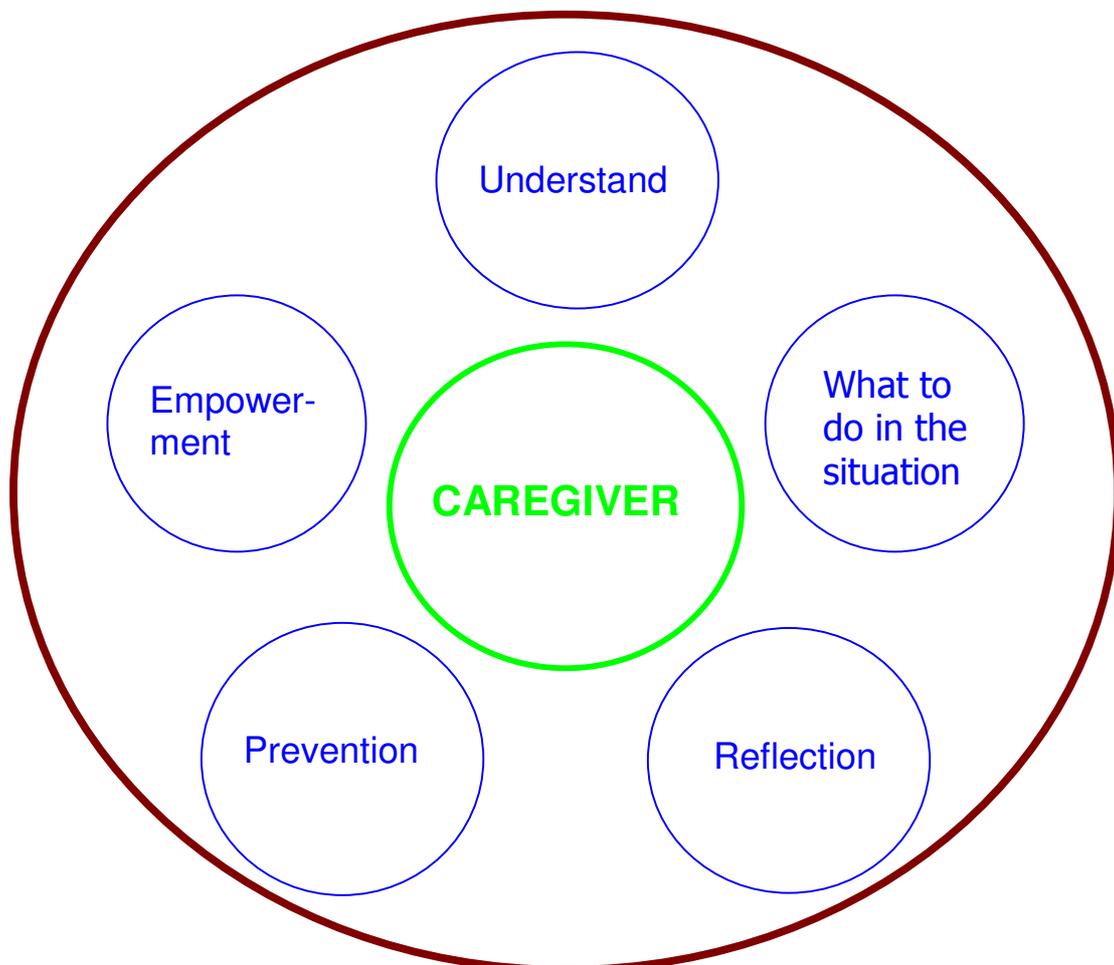
### III. Presentation Abstracts: A, B, and C

**A. Presenter:** Norbert Hödebeck-Stuntebeck

**Title:** Crisis management in support of people with Prader-Willi syndrome:  
Understanding, acting, reflecting and preventing crises

Every person who is involved with the care and management of a person with Prader-Willi syndrome will eventually experience a crisis situation. And each crisis experience offers an opportunity to learn how to better manage a crisis in the future. Crisis management is best understood as a circular rather than linear process as demonstrated by the diagram below. Note that the caregiver is at the center of what is called a circle of support.

#### People with handicaps (PWS)



#### Behavior problems

This model suggests that successful management and prevention of crisis situations depends on the completion of several steps.

First, caregivers must understand the crisis. Some strategies for understanding include:

- Asking searching questions to gather information about what the person in crisis is thinking and experience.
- Observing the person's physical reactions, expressions, and gestures.
- Making eye contact with the person so that communication remains open throughout.
- Examining any environmental factors that contributed to the crisis.
- Noting who is present and involved in the crisis.
- Establishing what happened before the crisis erupted.
- Determining if this is new behavior.
- Exploring what the client hoped to gain from this behavior.
- Recording any consequences (negative or positive) that were applied and to what effect.

Caregivers can then explore what meaning can be derived from information gathered, while completing additional steps in the process.

Second, caregivers must know what to do when a crisis occurs. It is important that caregivers understand their own goal in a crisis situation, and how they will act as a leader in the situation to restore control to the situation. This foreknowledge will allow caregivers to present a clear and stable personal plan for the person with PWS to follow out of the crisis. Specific strategies can be invoked such as:

- Allowing space to the person in crisis
- Use clear and calm speech
- Delay consequences
- Stay out of power struggles
- Help client to get "unstuck" from behavior or thought contributing to crisis
- Know limits

Third, after the crisis is resolved caregivers should take time to reflect with the client involved. Some issues for client reflection include:

- What did the client do well during the crisis or in resolving it?
- Encourage the client, as able, to explore the perspectives of others involved in the crisis.
- Develop, if possible together, a new behavioral plan to encourage improved behavior.
- Build bridges with the client through the reflection process. Even if the client is resistant to reflecting allow their participation.

This step should also included reflection of the caregivers to identify what they learned and experienced. A significant portion of time should be allowed for this

step and no restrictions should be placed on what is said or shared. The goal is to see the crisis honestly and accurately and with some distance and perspective.

Fourth, caregivers should use information and insight gathered in the three previous steps to inform prevention. Prevention can include making changes to:

- The daily schedule
- Opportunities for people with PWS to expend energy through the day
- The number of available caregivers
- Qualification of caregivers (i.e. would additional crisis response training help?)
- Crisis response protocol
- Level of support the institution provides for crisis intervention
- Utilization of external support (clinics, psychologists, etc.) to help in crisis situations or prevention planning.

Fifth, caregivers must feel empowered. This model is demanding of caregivers and requires strength to complete. However, if implemented properly and continuously, these steps allow caregivers to draw strength from their own experience and the support of those around them. Only empowered caregivers can create a process of crisis management and prevention that is reliable.

**B. Presenter:** Mary K. Ziccardi

**Title:** Management of crises in community settings: Educational and legal

The ongoing support required to manage an individual with Prader-Willi syndrome must occur in every environment and in all situations. A caretaker's failure to anticipate and respond to potentially difficult situations will either immediately, or eventually, result in a crisis requiring more significant intervention, along with the potential for an undesirable or even irreversible outcome.

For children and young adults, the challenges present in an educational setting may be significant, and certainly problematic, if ignored or incorrectly addressed. Delayed intellectual functioning, coupled with behavior patterns that draw negative social attention, can lead to a difficult, and often socially isolating educational experience. These factors, coupled with the anxiety of a food-laden environment, require both the competency and commitment of all school personnel to assure a successful educational experience.

Intervention by law enforcement presents special concerns. When summoned, law enforcement personnel are typically trained to "respond"; many times, a wait and see approach, along with verbal reassurance, will result in a favorable outcome and avoidance of an escalated crisis situation. Educating law enforcement prior to needing their assistance is recommended. Further, law enforcement personnel must understand that, contrary to their training, the

implementation of a consequence may have absolutely no effect on whether the same “bad act” will be repeated. Lastly, use of physical intervention must be a last resort, and only if imminent danger is perceived or present. Law enforcement personnel would benefit from advanced information about the risks associated with PWS and restraint, especially positional asphyxia and increased risk of fractures. When necessary, a cooperative partnership between the caretaker and local law enforcement authorities will increase understanding and reduce risks.

**C. Presenter:** Gregory L. Cherpes MD

**Title:** Prader-Willi syndrome and crisis management: Inpatient treatment setting

Individuals with Prader-Willi syndrome are at risk to experience physical and behavioral health difficulties that in the absence of adequate environmental management may reach critical dimensions. Certainly not all difficult behaviors are considered crises. A crisis could be defined as a situation where the supports available to an individual are less than the individual’s needs at that time. Crises may be short lived or chronic in duration. They may be brought about by factors unique to the individual, factors associated with the PWS phenotype, or factors associated with the environment’s response (or lack of) to the individual. When the individual’s current environment cannot adequately: promote necessary steps toward health, sustain physical or emotional well-being, or prevent the recurrence of issues leading physical or behavioral health emergencies, consideration can be given to treatment in an inpatient setting. (Note: in the United States, “inpatient treatment” refers to care provided in a hospital where the patient stays both day and night until he is discharged to home).

The Prader-Willi Syndrome/Behavioral Disorders Unit of The Children’s Institute is one such program that offers a model of inpatient treatment for the management of crises related to physical or behavioral health, including crises related to obesity, life-threatening conditions associated with morbid obesity, intense food seeking behavior, skin picking which reaches the level of self-mutilation, person-directed aggression, destructive behaviors, and identifiable psychiatric disorders. Whether “medical” or “behavioral,” the presenting difficulties that warrant the use of an inpatient management setting typically share the qualities of imminent threat to safety (self or others) and/or the presence of severe impairment in the quality of life. Alternatively, a specialized treatment unit may be considered when other treatment settings cannot meet the needs of the individual with PWS.

Over the course of the past 25 years, a specialized milieu has been developed to meet the needs of individual with PWS in crisis. Key components of the milieu include: 1) A structured, predictable environment, 2) Absolute food security, 3) A restricted calorie diet based on the individual’s current need, 4) A daily exercise program supervised by a physical therapist, 5) An individualized behavioral

management plan, 6) Accommodations to meet the needs of individuals with severe morbid obesity.

While alleviating the immediate crisis is of great importance, an effective treatment program must also address the factors present or absent in the individual's home environment which may have directly or indirectly caused or failed to prevent the crisis. Family or other caregiver involvement in education and training throughout the course of the individual's inpatient treatment episode is a necessary, capacity-building component of effective treatment. Likewise, capacity-building in the form of education and support among the individual's health care providers is essential to increasing the individual's likelihood of a successful return to their home setting.

#### **IV. Define the nature of the problem and why it is important:**

- A. Definition:** *A crisis is a negative situation that is out of control with risk to health and safety.* Examples generated by the work group included:

**IMMEDIATE RISK** (High severity): A crisis that must be dealt with in order to prevent immediate danger to self or others.

- Self-injury
- Aggression
- Medical illness (acute onset or exacerbation requiring medical attention of admission to treatment facility)
- Psychiatric illness (psychotic or acute episode of new illness)
- Running away
- Property destruction involving damage that can cause injury to self or others, i.e. breaking a window)
- Tantrum resulting in aggression towards others or self)

**POTENTIAL RISK** (Intermediate severity): A situation that places the caretaker "on alert" because it could escalate to a crisis.

- Tantrum
- Threats of aggression/property destruction/running away
- Food acquisition
- Psychiatric illness
- Property destruction
- [INVOLVEMENT OF LAW ENFORCEMENT – either by the person calling emergency numbers or others calling law enforcement due to the severity of the incident –added by the committee]

**MANAGEABLE RISK** (Low severity): Situations where a crisis may be emerging, but it is not as yet at the point to require full crisis intervention. In these situations,

Implementation of lower level preventative measures might stop a crisis from developing.

- Shut down from programming
- Tantrum
- Food acquisition
- Breaking the rules
- Psychiatric illness
- Splitting system of care
- Impaired judgment reflecting inappropriate behavior
- Bereavement
- Property destruction
- Uncertainty or changes in programming

## **B. Why is crisis management important for PWS?**

- Proper crisis management provides security for a person with PWS.
  - Security is knowing they will be taken care of no matter what. This builds a relationship of trust.
- Crisis management provides a “skin/buffer” to protect a person with PWS from the stresses and pressures of the community.
  - They are not doing it on purpose; they cannot express themselves appropriately (i.e., anger, sadness).
- A crisis is a learning opportunity for prevention.
  - It provides an educational opportunity to clarify the underlying issues.
  - It gives perspective; a reminder that the little things can become big things very quickly.
  - It provides the tools to be prepared.
  - It reaffirms that it is important to be one step ahead.
  - It provides an opportunity to prevent frustration before escalation.
- Crises are important in order to get to know each other (a process of relationship development).
- Crises cause stress and interfere with personal development and competence.
- Crises are exhausting, demoralizing and interfere with ongoing relationships.
- Crisis management keeps everyone safe.
- Crises are universal; everyone who works with persons who have PWS has experienced a crisis.

## **C. What makes a crisis more likely when working with persons who have PWS?**

Persons with PWS are highly dependent upon environmental parameters for their adaptive functioning, regardless of their level of intellectual ability. Further they require a high degree of consistency and predictability in their daily plan. FOOD SECURITY is the hallmark of the environmental infrastructure. This provides *no*

*doubt* about their diet plan; *no hope* of deviating from this plan (and because of secured food access, there is no chance of getting any more than predicted); and having fulfilled both of these requirements, there is *no disappointment*, and hence, no emotional distress and no acting out behavior. This *no doubt, no hope, no disappointment* paradigm also applies to the daily schedule. The person knows the schedule of activities and tasks throughout the day. *Preferred* activities follow less preferred ones to assure smooth transitions. All activities are time-limited (even leisure activities) so individuals are less likely to *get stuck*. By providing structure and consistency in the environment, the stress sensitivity that is characteristic of the syndrome is managed, and behavior problems are diminished.

There are several personality traits (the behavioral phenotype) typical of persons with PWS that inevitably place them into conflict with the care-giving individuals who are responsible for securing their environment. Persons with PWS display cognitive rigidity, inflexibility and perseveration that interfere with changes or transitions. As a result of their single mindedness, they have difficulty taking another person's point of view; their capacity to understand the impact or the consequence of their behavior on others is diminished. They can be noncompliant, argumentative and oppositional. Their judgment is impaired regardless of their intellectual ability. They have selective interests and excessive/repetitive behaviors that often require limit setting. Their emotional response can be intense and reactive or unexpectedly minimal. They can display a high degree of persistence and refuse to end an activity, or become easily frustrated and withdraw from an activity. Some of the most challenging behaviors include: aggression, self-injury, property damage, running away, shutdowns, temper tantrums, withdrawals or refusals.

These behaviors elicit a response from the caregiver. The nature of the caregiver's response can determine the outcome of these behaviors. Appropriate management maintains harmony with the environment; mismanagement results in greater conflict between the person with PWS and their environment. This conflict increases the person's stress, and increases their acting out behavior. Thus, a crisis can be precipitated when a system/provider/care-giver is not aware of the unique needs and personality traits of the person with PWS or when typical PWS behaviors are mismanaged.

## **V. Nature of the Problem**

### **A. When/where do crisis situations happen?**

Consensus existed among the group that a *risk for crisis* was high among individuals with Prader-Willi syndrome. This consensus may of course reflect a bias among the panel presenters and group members who elected to participate in this topic.

In addressing the question “When/Where do crisis situations happen?” an extensive list was generated and appears immediately below in the order presented by the group to the facilitator:

1. Parent visits (before or after)	16. Too much or insufficient information
2. Special events	17. Food situations
3. Communication issues	18. Social context (including intimate friendships)
4. Changes within care team, rules, or staff	19. Being rushed or hurried (time management)
5. Before breakfast	20. ‘Planned’ crisis
6. When there is too much free time	21. Unknown situations
7. Meals with other residents	22. Fear of evaluation
8. When children come home from school	23. Unmet expectations
9. Workplace	24. Arguments in social context
10. Seasonal	25. Crisis never occurs when sleeping
11. Full moon	26. Low structured weekends-more free time
12. Menstrual cycles (of staff!)	27. After lunch
13. Task specific	28. Time before going home
14. Function of expectation	29. For some, when left alone
15. Anticipation/expectation (planning)	

A change of any kind in the life of a person with PWS can cause a crisis. Some examples include:

- Change of placement
- Illness
- Bereavement
- Life changes
- Uncertainty – especially with food

There are some interactional situations in which a crisis can occur:

- Splitting situations
- The need to set limits due to impaired judgment
- Attention seeking
- Impaired judgment
- Poor communication
- Call to a “higher authority” (this referred to involvement of law enforcement or other authority agency)

Ultimately, the above lists reflect the fact that a crisis may arise at any point in a person’s day or life when the available resources, both intrinsic and extrinsic, are inadequate to maintain an individual’s health and safety. Vulnerability to the wide variety of external stimuli noted above underscores the importance of establishing and maintaining predictable routines, including all aspects of meals. Items listed above that reflect an internally mediated locus of control serve to underscore the importance of developing an appreciation for the individual.

While certain characteristics are attributed to persons with PWS, an in-depth knowledge of the particular individual being served is of equal or greater importance in identifying specific triggers to potential crisis-inducing situations. Personality style, coping skills, temperament, self-esteem, and past experiences vary from person to person and all contribute toward a person's tendency to experience a crisis. The presence of identifiable affective, anxious, or cognitive disturbance will serve to further influence an individual's response to the above situations.

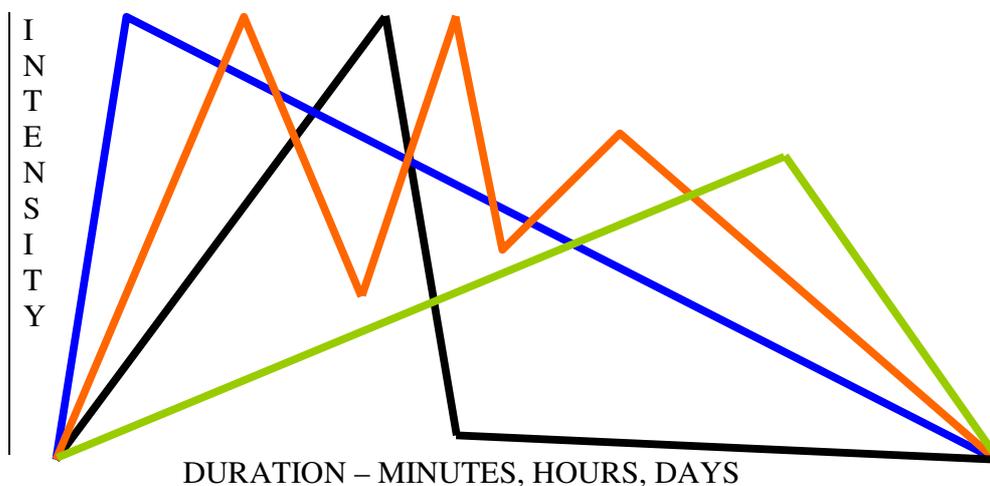
The group recognized the potential of caregivers to contribute to the emergence of crisis situations. A group member mentioned "menstrual cycles" (of staff) somewhat humorously. The spirit in which this comment was made was to emphasize the role of the caregiver's outlook, mood, and her own well-being. Poor communication among caregivers was also identified as an important contributor to potential crises.

While the broad list of potential triggers for a crisis may seem daunting, clearly individuals with PWS do not spend most of their lives in crisis. The list generated does reflect the powerful need for vigilance and mindfulness on the part of caregivers to help alleviate the risk for crisis. There is always reason to be hopeful that attention to, and anticipation of, internally and externally mediated triggers, which will certainly vary among individuals with PWS, will help to enhance the individual's well being.

## **B. How long does a crisis last?**

There is a high degree of variability in the intensity and duration of any given crisis. Some present unexpectedly, while others build slowly in evolution. Some appear to be resolving when they suddenly peak again. Some resolve quickly. Each pattern is unique to the person and to the context of the situation.

### **Anatomy of a crisis:**



Due to time constraints, the workgroup was not able to fully explore this topic. The two comments generated were: tantrums – 1 hour, or until the next meal. Certainly, a great variability exists in terms of the duration of a crisis. Duration is influenced by the cause of the crisis, the presence of potential perpetuating factors, as well as the resources of the caregivers to respond to the crisis. While “until the next meal” may reflect the experience of the workgroup member who offered the comment, the leadership team would like to emphasize the importance of caregivers not attempting to use food as a means of resolving a crisis situation!

### **C. What do you do when there is a crisis?**

**Caregiver response:** The presenters and the workgroup participants acknowledged that determining the appropriate response to a crisis situation is dependent upon knowing the individual with Prader-Willi syndrome. It is the caregiver’s role to ensure the safety of the person in crisis, as well as the safety of all others in the environment. This is most effectively accomplished through providing appropriate supervision and support. Some techniques that were identified by the workgroup participants include providing a distraction, using redirection to another conversation or activity, and effective use of active ignoring strategies.

- Provide a distraction: Use humor, play, or conversation; provide an opportunity for other activity (redirection).
- Change the situation; if in a dangerous situation – start up a conversation and move to safe place; the person with PWS will follow you, most likely.
- Ignore the behavior; take away the audience; do nothing – sometimes a crisis must run its course – “give room” – give space to the person – but assure safety of all.
- Provide 1:1 assistance, as needed.
- Clarify rules/consequences.
- Present a paradox (i.e., a response they wouldn’t expect): give the person what they want; surprise or shock the person.
- Allow the person to cry.

It was agreed that some crisis situations, when health and safety are not at risk, need to “run their course”, while space and supervision are provided. Based on the relationship between the caregiver and the person in crisis, humor and play may provide opportunities towards crisis resolution. It was noted by the workshop participants that often, during or after the crisis, the individual in distress might actually cry. As with all other interventions identified, support and reassurance should be given. Crisis response depends on the setting and knowing the person’s pattern of behavior.

A caregiver’s attitude and approach during a crisis situation is paramount towards ensuring a safe outcome. The caregiver’s ability to avoid argumentative

comments and use a neutral and reassuring voice are desired responses. Using brief sentences and clear directions may also assist in re-establishing control of the situation.

- Low key emotional expression and low voice volume
- Communication – short and clear
- Never argue back, but acknowledge what is said
- Have neutral affect and calm voice

On the topic of the use of physical restraint during a crisis, group consensus was not reached. This remained true regardless of the opportunity for participation in recognized physical intervention management training courses. Workshop participants did, however, acknowledge the use of techniques such as therapeutic holds and defensive strategies (i.e. passive block with an open hand) if the situation presented imminent danger. Use of a locked door as an environmental barrier was also considered in the discussion.

Protecting the individual from self-injury is a constant objective for caregivers. If a specific object is being used to cause self-injury, caregivers are in agreement to remove any dangerous items and to block head banging with a pillow, for example. Obviously, obtaining medical treatment for any significant injury is required.

Restrictive strategies that *were* used include\*:

- Locked door
- Hold the person if in dangerous situation
- Defensive strategies;
  - Passive block
  - Open hand
- If self-injury, offer medical treatment.
- If an object is being used to cause self-injury, remove it.
- Block head banging by the use of a pillow

\* Always document (critical incident report).

#### **D. What do you do *after* a crisis?**

**Post-crisis intervention:** A caregiver's role at the conclusion of a crisis is equally as important. Workgroup participants offered several suggestions during the post-crisis phase. Suggestions included that having the caregiver re-establish an environment of control and security, while accepting that the incident is over and "letting go" of it. Sometimes, it has been found helpful for the caregiver to accept responsibility for what occurred.

Some workgroup participants made suggestions regarding post-crisis communications. It has been deemed helpful to communicate something positive

to the individual following a behavioral crisis. Other post-crisis communications included agreeing to a (new) contract about behaviors and exploring meaningful and/or natural consequences. The use of consequences, as part of an overall management program approach, was not explored in depth within this workgroup.

- Re-establish control
- Accept the fact that it is over
- Just drop it
- If you are the person in charge, take responsibility
- Find something positive to affirm in the person
- Ask person what they think should be the consequence
- Consequences have to have meaning for the person
- Make contract regarding behavior

## E. What do you do for prevention?

**Planning for prevention:** Preventative strategies were discussed as the necessary key element in reducing the frequency and severity of crisis incidents. The workgroup agreed that is important to know the behavioral history of the individual receiving services, as well as information about his/her family, but recognized that continuity in communicating the information may be difficult due to staff turnover.

Advance planning is critical to preventing crisis situations. Some specific strategies discussed by workgroup participants were contracting for understanding of expectations, use of social stories, and clarification of expectations, outcomes and relevant issues, balanced with the individual's needs and desires.

- Advance planning
- Contracting based on prior crisis
- Social stories (using drawings or words)
- Clarify expectations/consequences
- Clarify emotional issues/needs
- Know the history of the person/family
- Know the person's behavior history

## VI. Impact of the Problem

- A crisis impacts the life of a person with PWS in significant ways. To begin with, a crisis calls into question whether or not their world is secure and their caregivers are trustworthy. If those questions are not answered affirmatively through the effective resolution of a crisis, then the increased anxiety of the person with PWS will likely lead to even more crisis situations. Conversely, if the crisis situation is handled appropriately and effectively, then proper crisis management can strengthen the bond between the person with PWS and their caregivers, leading to a more crisis-free environment in the future.
- Family members can also be dramatically affected by crisis because a crisis can trigger concerns that their loved one is in a living environment that does not safe guard their basic human need for safety and security. These family concerns can result in increased friction with caregivers and possibly lead to removal of their loved one from the program. Also, friction between family and caregivers can slowly erode the partnership between the caregiver and the person with PWS that is so essential to maintaining a healthy living environment.
- The impact of a crisis can have a profound affect on caregivers. Exhaustion, increased excitement, physical tension and a feeling of helplessness can accompany a caregiver's crisis response. Effective crisis management requires a high level of professionalism in the face of escalating stress; it demands clarity of focus and a neutral emotional approach. Caregivers rely on their supervisors to empower their response through the availability of necessary resources, such as adequate staffing patterns, training, and appropriate policies and procedures to support them before, during and after a crisis situation.
- Crisis situations are particularly difficult because, by definition, they are emotional events – both for the person in crisis and the caregivers who are responding. When asked how crises made them feel, work group participants shared 10 responses; only one of these was a positive response: focused and calm. The rest of the responses captured a range of negative reactions including feeling helpless, tense, a lack of emotional control, a sense of failure, indecisive, overwhelmed, and a need to lower expectations. Work group participants also indicated that in some cases their flight/fight impulse was activated during a crisis.
- Interestingly, frustration was identified as an issue for both caregivers and the person in crisis. For the caregiver, frustration builds because of the ability of people with PWS in crisis to push the caregiver's emotional buttons to cause hurt feelings. These hurt feelings potentially lead to anger for the person with PWS which can extend long after the crisis if not resolved in a healthy and constructive way. For the person with PWS, frustration can grow with the

caregiver's response to the crisis. In time, if that frustration is unresolved, it can turn into a long lasting anger towards the caregiver. This simmering anger can lead to behavioral problems – and possibly another crisis – in the future.

Crisis management challenges providers on both a person-centered and an agency-centered level. On the person-centered level poor crisis management can result in staff burnout; physical and emotional injury to both staff and people with PWS; serious disruption in daily activities and services; and a loss of the sense of community within the living environment. On the agency-centered level a provider can experience potentially expensive property damage, litigation, and staff turnover by not managing crisis effectively. Significant problems with crisis management can attract the attention of monitoring agencies resulting in additional oversight - which can be time consuming and demanding - and in some cases it can result in a decrease or elimination of funding. Frequent crisis situations also diminish a provider's reputation in the provider community and among potential consumers.

Crisis response has an impact on the system of care in society. Escalating crisis cases can exhaust local supports when community resources (i.e. medical, educational, and legal) cannot handle the level of care needed to respond effectively and appropriately to the person in crisis. When this happens, a person with PWS will often be placed in a situation where the primary responders (for example, the police) are not trained to react in a constructive manner. This can lead to a worsening of the crisis situation and an even longer disruption of the daily therapeutic programming so vital to the stability and health of a person with PWS. As a result of these factors, crisis interventions beyond the immediate caregiver level are costly and frequently ineffective. Thus, it is a priority to give primary caregivers the crisis management tools, training, and resources needed to respond immediately and effectively to a crisis situation so that it does not escalate to involve a local system of care that is often ill equipped to manage a PWS specific crisis situation.

Participants acknowledged there could be a difference between the perspective and understanding of Prader-Willi syndrome among frontline staff, their supervisors, and program administrators. This difference can be harmful to the effective functioning of an agency, as conflicting opinions and views can lower staff morale, decrease the value of programming, and prohibit the kind of consistency and structure necessary for effective PWS care giving. Therefore, it is very important for an agency to have a shared organization-wide commitment to working with people with PWS, based on available best practices. This might mean, for example, adjusting caseloads for people working with PWS clients who may require more time. In addition, most successful organizations have people in management with direct service experience working with people with PWS so they have an appreciation for and understanding of the challenges staff face.

This understanding will contribute to better communication between staff and management and reduce frequency of conflict and burnout.

## **VII. Discussion summary**

### ***Key points*** endorsed by consensus:

- Crises are a part of life.
- Crises are a learning opportunity for everyone.
- Crises may have a healthy role in organizing the system of care.
- Crises interfere with the psychological development of the person due to stress.
- Crises interfere with the relationship between the person and the caregiver.
- Crisis management requires accountability.

### **Issues outside consensus:**

- Accountability of the person with PWS for their behavior:

A primary issue that demands further exploration is the caregiver's assessment and understanding of our individual and collective philosophies regarding "accountability" (i.e. the personal responsibility that can be expected by a person with PWS for an individual was regarding their behavioral choices, including motive, forethought, the planfulness of their actions, and their ability to predict and appreciate the results of their actions. It follows, then, that the value of the application of consequences, as well as the short and long term benefits and risks, requires extensive and ongoing discussion.

### **Unanswered questions:**

- Cultural differences in expectations for the use of physical restraint and therapeutic holds (Autonomy vs Paternalism):

Individuals with PWS are rarely capable of making informed decisions, especially when they consider food access to be a variable. Impaired judgment, regardless of intellectual capacity, informs environmental management. The question remains: under what circumstances are individuals with PWS able to make autonomous decisions, and when must others who are knowledgeable about the person and the syndrome make decisions on their behalf (paternalism). There are cultural differences in our understanding of an individual's right to make choices versus their competence to make decisions. Competence is determined by the capacity to understand the potential positive or negative outcome of a decision. Caregivers can enhance the decision-making ability of an individual with PWS by limiting the number of choices, minimizing the risk of an unfavorable

outcome, and selecting an emotionally neutral time or situation to make choices. When an individual with PWS is too sick, too angry, too combative, too destructive or too self-injurious, their capacity to make even limited decisions is impaired, and the caregiver must act with paternalism. At times, this may require physical intervention in the form of a physical block, a therapeutic hold or physical restraint. There are cultural differences in the implementation of physical interventions despite the universal goal of maintaining personal safety.

## **VIII. Guidelines for best practice**

1. The top priority in crisis management is to protect the person, staff and others from harm.
2. There must be acceptance that crises will arise when working with a person with Prader-Willi syndrome, and the overall goal is to reduce the frequency and intensity of the incidents.
3. In response to a crisis, effective staff will be able to stay calm, think before acting, know what to do and display a capacity for self-reflection.
4. Staff orientation and training should provide ongoing PWS-specific information including crisis management.
5. Effective staff should be able to offer security and “unconditional love” despite the person’s crisis.
6. Effective staff will utilize a team concept with communication about advanced planning, structure, consistency.
7. Following a crisis a debriefing process should occur to find reasons for the crisis without blaming anyone.
8. Advanced planning is a strategy for prevention with two components:
  - Person centered planning includes understanding the history of behaviors, what motivates them, and how to communicate best with them.
  - Caregiver centered planning includes adequate training and appropriate staffing patterns.
9. Administrators must appreciate the seriousness and the challenges of working with crises occurring in persons with PWS.
  - Care-givers would feel more supported from their administrators and others by:
  - Providing opportunities for relationship building with the person through 1:1 staffing.
  - Well-structured teams.
  - Advanced training on PWS and crisis management.
  - Not working alone (no one-caregiver shifts).
  - Time for staff meetings and communication.
  - Professional mentors and support networks.
  - An informed organizational structure regarding PWS.
10. Online support group (international).

## **IX. Conclusion**

**Feedback on workshop:** Workshop participants were overwhelmingly positive in their response and almost everyone participated. As a result, many suggested hearing how others respond to crisis situations helped participants learn new strategies and think through their own responses to crisis situations. Participants also appreciated the opportunity to be part of a group that understands the inherent challenges (and joys) of working with people with PWS. And, while participants felt more discussion is needed, this workshop provided a new motivation for their work. It also empowered most participants to better handle crisis situations in the lives of people with PWS.

**Impressions of the leadership team:** The leadership team appreciated the participation and contribution of each and every member of the work group! The exercise was a learning experience for everyone. It underscored several facts. First, each individual working with a person who has PWS has a unique vantage point from which to view the person and the impact of the syndrome on that person. Second, cultural diversity and level of experience also affected the work group's knowledge, skill level and their point of view. Third, the participants selected the topic of "crisis intervention," suggesting that they had a personal bias based upon their experience or their desire for knowledge pertaining to this topic.

The leadership team perceived a sense of urgency to resolve the unanswered questions and issues lying outside consensus. These included issues related to accountability for their behavior, the use of physical restraint, and autonomy with decision-making.

## **X. Challenges and items for future consideration:**

Participants endorsed the concept of developing a global on-line support network for care providers that would build on the international information sharing of the workshop. It is believed that an ongoing opportunity to share insights, discuss challenges, evaluate strategies, and provide support to each other on a regular basis would be an invaluable resource for care providers around the world.

Participants left unresolved the challenging issue of what role, if any, does the practice of physical restraint have in providing care for people with PWS. Many participants had different personal philosophies and some participants work in countries where physical restraint is illegal. As a result, this issue requires much more international exploration before any meaningful consensus can develop.

QED

# First International PWS Caregivers Conference

Herne, Germany

June 3-5, 2008

## Workgroup B3: Communication between Caregivers and Families

### I. Workgroup Leaders

Presenters:

Jeff Covington, Administrator-USA

Anne Ogden, Administrator-USA

Dr. Jorgelina Stegmann, Physician-Argentina

Facilitator: Dr. Barbara J. Goff, Professor of Special Education-USA

Scribe: Jackie Waters, Parent/Advocate-United Kingdom

### II. Workgroup Participants and Dynamics:

There were twenty-four participants in this work group from diverse backgrounds. Sixteen were from Germany, three from the United States, two from Sweden, and one each from Israel, Argentina, and the United Kingdom. The majority were caregivers (15); in addition to three administrators, two teachers, a physical therapist, a special education professor, a physician, and a parent.

All participants agreed that this was a crucial topic and as Dr. Goff stressed, "It is at the heart of all we do." Many caretakers felt that this was not something they were specifically given training about, but very much believed was needed as part of their preparation programs.

### III. Presentation Abstracts

A. Presenters: Anne Ogden and Jeff Covington

Title: Communication between Caregivers and Families: A Comprehensive Approach to Building Trust between Providers and Families of Individuals with Prader-Willi Syndrome

Communication is one of the most basic components of being a human being. However, on a daily basis we all recognize that our communication is often misunderstood or taken out of context. While on a basic level communication is simple, on an application level, it can be treacherous.

The flow of communication between a caregiver and the family of the individual receiving care is a crucial component to creating synergy in a person's life. It is not unique to individuals with Prader-Willi syndrome, but does warrant substantial discussion within that community based on the all-consuming nature of care giving parents have had to provide prior to residential placement. The scarcity of the syndrome coupled with the lack of awareness and understanding in the greater community have left parents feeling as if they are the only ones who can care for their child with PWS. Relinquishing that responsibility to a professional caregiver can be an emotional and traumatic experience. It is therefore essential for the caregiver community to understand the unique nature of this important aspect of care for the individuals they serve.

This presentation will take participants through the process of building the relationship with the families based on trust. The essential elements to building and sustaining a positive working relationship between caregiver and family member include respectful interactions, prompt responses, demonstrated empathy, and a focus on problem-solving versus blaming. Once trust is established the family and provider must working on setting expectations. The presenters will speak about how to do that and what examples of those expectations should be. They will also delve into what to do if those expectations are not met by either party. Next the presentation will move into what providers can do if they are stuck on a particular matter; calling in recognized leaders and experts to assist, and how that can send a clear message to the families and establish their trust.

**B. Presenter:** Jorgelina Stegmann, MD

**Title:** Learning and Working with Parents

In Argentina and Latin America there are no institutes for people with PWS. The patients live with their families. The families are the caregivers for the individuals with PWS. Good communication between parents and health professionals is the key for successful treatment. The relationship must be based on trust. This trust building is based on knowledge about PWS and the dynamics of the families.

The successful treatment is based on:

- Diagnosing the situation in the family and in the patient
- Understanding the syndrome through EDUCATION
- The family and caregivers giving the same message to the patient.
- All speaking the same language

- Support for the parents (psychiatric diagnosis for the parents, meetings between the parents and professional, etc).
- Learning and working together (as a dynamic process)
- Seeking the **meaning of life** of each individual, working on his/her difficulties to achieve a social reintegration, with changes at the psycho-physical level.

These dynamics between parents and professional are a shared process toward achieving the well being of the individual with PWS.

#### **IV. Nature of the Problem**

Given the genetic nature and associated characteristics of PWS, it is essential for the caregiver community to understand the unique circumstances of the family of an individual with Prader-Willi syndrome. Research has shown that stress on the family with PWS is much greater compared to other conditions. It is not surprising that this is so given the life-threatening nature of the syndrome and the wide-sweeping life changes a family must make to ensure the health and well-being of the family member with PWS. Furthermore, it is considered a low-incidence disability and, as such, few professionals in any related fields have adequate, if any, knowledge of PWS. So, when families reach out for support and services, they are often met with questions and even skepticism about the specifics of the disability.

Those who have worked with families of individuals with PWS know that it is imperative to establish a trusting relationship with families built on proactive, responsive, and effective communication. The relationship is one of interdependence. The knowledge of parents about the child must go hand in hand with knowledge of staff providing PWS services and supports.

Anxiety is a hallmark of Prader-Willi syndrome and, as caregivers and families working together; we must keep that knowledge in the forefront of all we do. Individuals with PWS are best served when they are assured that all those who care for them are in agreement as to what and how services will be provided, who is responsible for providing those services, how decisions will be made, and that they are participants in that decision-making.

#### **V. Impact of the Problem**

The experience of individuals with PWS manipulating the relationship between their families and their caregivers is well-known. For example, the individual may claim that the parent gave permission for a special treat during an outing with a caregiver, or the individual may claim to the parent that the caretaker is somehow abusing him in hopes of obtaining some desired end. In some cases, it results in simple misunderstandings, but in others, the results can be devastating. If parents and caregivers fail to recognize and honor the role each plays in the life

of the individual with PWS, years of struggle, even a loss of services can be the result. Some direct care providers have reported their relationship with the parents of the individuals with PWS as the most difficult aspect of their job, while parents frequently report that communication with caregivers, inadequately trained staff, and high turnover make it very difficult to have a trusting relationship with those providing services.

A lack of trust and ineffective communication can be a disaster for any relationship. The bond between caregivers and families of individuals with PWS can, at times, be precarious and, if not tended to, become detrimental to the individual being served. Families often fight for years to receive appropriate services for their individual with PWS and become battered and bruised along the way.

The impact of this constant struggling on the family can be devastating. If they do not have trust in the provider, their level of stress and worry may be extremely high which can further erode the caretaker-family relationship. For the caregiver, not feeling trusted or supported can be very difficult to manage. Rightfully so, caregivers take a great deal of pride in their work. Being second guessed or undermined can have a negative impact on their delivery of care. Individuals with PWS may perceive this strain and exacerbate the conflict between the parties to achieve their own ends or it may simply increase their already heightened level of anxiety, in turn, negatively affecting their behavior.

What is the resolution to this relationship dilemma? How can caregivers engage with families so that they feel they no longer need to fight for understanding and essential services for their child? What do caregivers need from families to support them in their work with individuals with PWS?

## **VI. Discussion**

There was international consensus that the family/caregiver relationship is critical to ensuring a high quality of service, indeed a high quality of life, for individuals with PWS. Caregivers were unanimous in their desire for specific training in working with families. The discussion evolved into a brainstorming session on essential components to such a training program.

To begin, we found it important to identify specific family characteristics and consider their impact on the care of the child with PWS, both for the family and the professional caregivers.

Each and every family is unique. There are several characteristics that should be taken into consideration when working with families. While the task force was not able to explore each of these characteristics and their impact on relationship building and service provision, we urge providers to consider the role they may play in living and working with individuals with PWS. Some critical characteristics are:

- Parental age and health status
- Parental marital status: married/partnered/single
- Birth parents/foster or adopted parents/guardians
- Siblings/No Siblings
- Family income relative to family needs
- Extended family/Alone
- Other support systems
- Religion
- Language
- Culture

This is not an exhaustive list, but is a recommended start to thinking about families as unique systems, often quite different from the caretaker providing services. Frequently, the conflicts between caretakers and families have less to do with the actual care being provided than the dynamics of understanding and communication between parties.

The task force also discussed the many different settings where an individual may receive services and how each of these settings has its own characteristics, philosophy of service, and legal mandates. At times, the requirements and approach to service in one setting may be in conflict with those of another. This creates problems for everyone involved; the individual, the family, and the caregivers. Some examples of service settings are:

- Adults in residential services
- Adults receiving vocational services
- Children in residential schools
- Children in neighborhood schools
- Individuals receiving therapeutic services (Occupational Therapy, Physical Therapy, Speech and Language Therapy, Counseling, Specialized Behavior Support, and many more depending on the individual)
- Individuals receiving specific medical and health services (general medical care and monitoring, nutrition, care for health conditions associated with PWS; vision, diabetes, sleep apnea, scoliosis, dental caries, and other specific needs)
- Individuals receiving in-home supports (behavior management, respite, academic)

After the workgroup established the different family characteristics and the services that people receive from caregivers, we looked more closely at the dynamics that affect communication between families and caregivers. We built consensus on several key points. Building a good relationship with the family is the foundation for on-going effective communication. It must start early and be nurtured continually. Building that relationship requires trust and honesty on both sides. We talked about various ways that this can and should happen and included them in our best practice guidelines.

Supportive relationships and effective communication depends on clearly articulated mutual expectations between the caregiver and family. The group felt that if families and caretakers invested the time and effort into developing shared expectations and writing them down, then disagreements and confusion would be greatly reduced. Families come with their own needs and concerns for their child. Caregivers represent agencies or institutions which must adhere to certain standards of care which are often unfamiliar, even questionable, to families. Having explicit discussions about expectations, requirements, and areas of difference which result in an agreed upon, clearly written document, will clear up any ambiguity that could cause a diminishing of trust and a breakdown in communication. Still, it was noted that there was no way to prepare for every contingency. It is impossible to come up with expectations for scenarios that have not yet been encountered, but one can put in place a process whereby new situations are addressed in a specific and mutually acceptable manner.

The next part of our discussion dealt with what to do when expectations are not met by either side of the relationship. It was stressed that an even greater effort at communication must occur throughout this phase. The group recognized how disheartening this is for the individual with PWS and how this type of breakdown in communication can affect their sense of security and overall well-being.

Finally, our group was primarily made up of caregivers, specifically residential providers. Many reported that they did not receive any training in this area and that it would be important for them, and all care giving staff, to do so.

For the remainder of our work session, we discussed what Caregiver-Family Communication training should look like. The results are incorporated into our Guidelines for Best Practice. [Please note: the task force attempted to provide guidelines applicable to a variety of situations and settings, yet recognized that some may require adaptation to a particular circumstance].

## **VII. Guidelines for Best Practice**

### *A. Building the Relationship*

It is extremely important that the relationship between the family of the individual with PWS and the caregivers be one of mutual respect and trust. Here are some best practice guidelines to establish and build an effective and enduring relationship:

- Establish communication prior to the person receiving services.
- Conduct and facilitate support groups for parents.
- Spell out rules, regulations, and expectations.
- Acknowledge that mistakes will be made.
- Emphasize that the person with PWS is the main focus and is at the center of all decisions being made.
- Be proactive:
  - Initiate contact early and maintain a routine.

- Identify and provide appropriate agency contacts.
- Reinforce the development of the partnership by educating each other:
  - Family is the expert on the individual.
  - Caretaker/Provider is expert on the service being provided.
  - Collectively, they have expertise on PWS as experienced by the person with PWS.
  - Relationship will be tested for loopholes by the individual with PWS.
  - Conflicting opinions can be confusing and cause anxiety for the person with PWS. They may also use differences of opinion, or lack of consistency on either side in carrying out agreed actions, to play one party off against another.
- Appreciate the role of the family:
  - Recognize past struggles and success.
  - Respect what is important to the family.
  - Establish systematic means for input into decision making.
  - Recognize that the relationship between caregiver and family is similar to a “marriage” with the individual with PWS in the middle.
- Respond promptly to questions and concerns:
  - E-mail is quicker and very useful for brief information sharing, but these abbreviated descriptions and discussions of more complex issues can lead to misunderstandings.
  - Return phone calls within 24 hours even if you don’t have all the information, just to acknowledge and let them know you are following up on their concern.
  - Establish trust:
    - Be candid and honest.
    - Admit mistakes and seek input on how to avoid misunderstandings in the future.
- Don’t blame:
  - Identify problems and seek solutions without assigning blame.
  - Identify how to avoid similar difficulties in the future.
  - Move on...

### *B. Mutual Expectations*

In any relationship, it is important that the expectations of each member of the relationship are established from the beginning. Once expectations are established, there is less room for disagreement and a greater likelihood of a mutually satisfying relationship. Establishing expectations allows for a shared understanding of how the relationship will work and what each party can expect of the other. The following are some areas to be discussed and agreed upon for establishing mutual expectations:

- **Caretakers Expect Parents/Families to Support Policies in These Areas:**
  - Communication with individual with PWS (by phone, mail, email)
  - Diet plan for individual

- Exercise plan for individual
  - Scheduling and attendance of medical appointments
  - Communication of issues – who to talk to about specific concerns, when to contact them, how to make contact
  - Family visits to the school or residence
  - Program restrictions:
    - Access to food and kitchen
    - Caloric intake
    - Exercise plan
    - Access to money and credit cards
    - Phone usage
    - Time alone
  - Behavior support plans
  - Levels of intervention during a behavioral crisis: who intervenes under what circumstances and in what manner
  - Routine decision-making: role of the individual, the family, and the provider/caretaker
  - Discipline
  - Friendships
  - Romantic relationships
  - Sharing concerns about staff
  - Visits to family’s home for holidays, vacations, weekends, overnights:
    - Following pre-established caloric intake guidelines and exercise plan to the extent feasible
    - Scheduling and notification
    - Communication about visits
    - Asking for help and support when visits are difficult
- ***Caretakers/Providers also want families to:***
    - Understand and appreciate some of the constraints they work under imposed by funding sources, licensing or oversight agencies
    - Be open and honest about what happens when the individual is in their care
    - Work with them in developing mutual trust
    - Establish a healthy balance of interaction and involvement
    - Acknowledge and recognize the work of caregivers
    - Show interest by being involved and asking questions
    - Be open to discussion toward finding solutions when problems arise
    - Support their child’s growth toward greater independence
  - ***Parents/Families Expect Caretakers/Providers to:***
    - Provide information about institution or service; purpose and function

- Provide regular and current information about the individual’s daily life at agreed upon intervals pertaining to:
  - Health/Medical
  - Behavioral
  - Relationships
  - Activities
- Focus on improvements and positive developments in their son/daughter’s life; provide unscheduled proactive and frequent communication.
- Involve them in the planning process, decisions, and goals
  - Ensure on-going staff training and supervision
  - Ensure an adequate number of trained staff
  - Provide regularly scheduled community based recreation and social activities
  - Provide immediate notification of problems
  - Recognize their past struggles and triumphs
  - Be flexible with rules and routines when warranted
  - Look at the whole individual, past the “PWS label”
  - Be available to meet in home setting
  - Conduct social gatherings with families
  - Remain open to learning from individual and their family
  - Listen and recognize parent’s hopes and dreams for individual
- Understand their level of involvement in consideration of:
  - Needs and demands of all family members
  - Distance from program
  - Work schedules
  - Health concerns
  - Traditions and beliefs
  - Financial and other resources
- ***To Ensure That Expectations are Met, Both Parties Should:***
  - Be proactive:
    - Start prior to admission or onset of services
    - Routinely update and communicate expectations
  - Write the expectations down:
    - Contracts
    - Memorandums of Understanding
    - Agreements
    - Policies and procedures
    - Signed by individual, family, and caregiver
  - Deliver clear communication about:
    - Holiday celebrations
    - Home visits
    - Vacations
    - Absences from program

- Participate in frequent meetings to discuss expectations:
  - Are they being met?
  - Are they appropriate and attainable?
- Discuss the failures and successes

It is important to recognize that despite how the expectations are set, they should be as clear and concise as possible with little “wiggle room” allowing for misinterpretation.

- ***Providers: What to do if expectations are not met***

Despite the best efforts of all involved, there are times when expectations will not be met and there will be conflict between the parent and the caregiver, often with the individual with PWS caught in the middle. Here are some best practice guidelines for what to do if expectations are not met:

- Remember who you are here for: the individual with PWS
- Be objective – don’t blame –be understanding, yet firm
- Problem solve and build consensus:
  - Apologize when called for
  - Clarify expectations
- Move forward
- Continue to communicate
- Call meetings:
  - Utilize an objective facilitator when coming to an impasse.
  - Local government or funding source involvement as appropriate.
- Collaborate with an acknowledged PWS expert:
  - Sends a message that you are invested in the individual and their family.
  - Serves as a good resource for staff who may be caught in the middle.
  - Looks at a situation more objectively.
  - Identifies things that can be revamped.
  - Acts as a buffer between family and caregiver.
  - Provides recognition and validation of family caregiver.
  - Identifies and validates things that are working.

### ***C. Develop a training curriculum for caregivers on communication with families***

Caregivers should be provided with training on building effective relationships with families. The following content would be the core of that training:

- Learning active listening and empathy skills
- Understanding the perspective of a parent:
  - Utilize a parent whose child is not being served by the program

- Focus on appreciating the parent/child relationship
  - Recognition and understanding of the guilt, fears, doubts, and triumphs
- Involving families in special events and celebrations
- Establishing proactive and routine communication:
  - How and when does this occur
    - Phone, face-to-face, written (e-mail)
    - Daily, weekly, monthly, etc.
    - Including the individual with PWS
- Establishing and maintaining boundaries:
- Negotiating and implementing contracts, memorandums of understanding, other agreements
- Understanding the role of the family in the individualized planning process
- Contacting and communicating guidelines
  - Who does the family talk to about certain things?
  - What is the best time to contact people?
  - What is the flow of communication with the individual with PWS?
- Clarifying appropriate conduct/contact between caregivers and family:
  - How does this affect person with PWS?
  - What is balance between being too personal and too professional?
  - What are the roles of the various caregivers with the family?
- Providing practical help for parents to deal with the bureaucracy involved:
  - Eligibility for services
  - Payment for services
  - Establishing financial benefits
  - Implementing educational plans
  - Obtaining necessary auxiliary therapies
  - Residential or vocational placement process
- Understanding and appreciating the needs and concerns of the individual at different developmental stages and sharing those understandings with the family:
  - School placement
  - Adolescence
  - Vocational opportunities
  - Residential placement
  - Relationships
- Educating parents about realistic expectations regarding what caregivers can and cannot do:
  - Parent is the expert on the individual, the caregiver is the expert in providing [a particular] service to individuals with PWS
  - Individual can lead a happy and productive life with structure, safety, and continuation of traditions
- Understanding that parents cannot always provide/do as caregivers:
  - Home visits and diet maintenance
  - Community experience

- Degree of oversight and supervision
- Holidays and vacations
- Supporting the individual with PWS and parents to live separate lives:
  - Involves risk and trust
  - Person is receiving support to make good decisions
  - Person is becoming independent of them

#### *D. Unanswered Questions*

There were several questions and considerations that could not be addressed in this forum due to different rules and beliefs among the participants:

- How much information is too much information?
  - Will sharing of mistakes erode trust?
  - What parents want to know and not know is individually determined.
- What if the individual doesn't want certain information shared with the family?
  - What are the legal and ethical ramifications for the caregiver/provider?
  - Will not sharing erode trust with the family?
  - Will sharing erode trust with the individual?
- In certain countries and cultures, who parents talk to about specific topics varies greatly. All service systems are arranged and composed differently and individual communication protocols need to be developed by each service organization.
- What is the responsibility of the caregiver with regard to the family's religious beliefs and potential conflict with caretaker philosophy and legal obligations?
- The legal relationship between the individual and their parents varies from country to country and affects many of the issues identified above; therefore resolution may be country-specific.

#### *E. Future Considerations*

Moving forward, there are several themes and ideas that the group established for future consideration:

- A standardized curriculum (with appropriate cultural adaptations) should be established for communication between families and caregivers. (Separate curricula to be developed according to service: educational, residential, vocational, therapeutic, medical, and other.)
- Identification of strategies and activities organizations can employ to include all families with respect to their culture, race, religion, and socioeconomic status?
- Identify the needs of families of individuals living at home how caregivers can best support these individuals and families across their lifespan?

- Recognition and financial support for caregivers to provide necessary additional support to families in order to more effectively serve the individual in their care:
  - Training for families and all others involved in the care of the individual with PWS
  - Therapeutic services beyond the scope of funded services
  - Increased communication between parents and caregivers
  - Support groups
  - Transportation for visitation
  - At-home behavior and environmental supports
- Consideration of the need for caregivers to access additional outside assistance to effectively support the parents and the individuals they serve:
  - National and regional PWS organizations
  - Recognized professional experts

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First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workgroup B4: Environmental Structure for Work with  
People with PWS

**I. Workgroup Leadership Team**

**Presenters:**

Renate Scharfenberg – Germany

Dorthee Pederson – Denmark

Marguerite Rupnow – USA

**Facilitator:** Dr. Hubert Soyer – Germany

**Scribe:** Jackie Mallow – USA

**Translator:** Monika Fuhrmann – Germany

**II. Workgroup participants**

Parents, caregivers, and providers from Europe, Scandinavia and USA

**III. Presentation Abstracts: A, B, and C**

- A. Presenter: Renate Scharfenberg (Psychologist and Psychological Psychotherapist), Regens Wagner Absberg, Germany  
Title: Environmental Structure for Work Situation in Sheltered Workshops

Right to work for people with PWS

- Assessment by a special committee to decide the adequate work situation: free labour market or sheltered workshop
- criteria for a sheltered workshop: some social skills, not too much nursing, minimum of a commercially useful performance

Rights like an employee

- works council (aim: co-determination, play an active part, express wishes and interests)
- insurances
- workshop decree
- work contract
- monthly salary

### Phases of a sheltered workshop

- job skills training phase ( one or two years): basic skills are trained (e.g. punctuality, reliability, social-cognitive skills, solving of conflicts and problems) in addition special skills for a certain workgroup are trained (e.g. use of tools and machines like a soldering-iron); the aim is to find the adequate workgroup which meets the needs and interests of the individual person with PWS
- after the vocational training: the competences, the knowledge of skills and experiences in the work fields are further broadened; in the workgroup: interests, physical abilities, strengths and resources are considered

### Aims of sheltered workshops

- participation in social life by working ( a way of earning money and of self-realisation)
- integration and inclusion (working with other people of different kinds of disabilities in different work fields, partly in the community)
- empowerment, self-confidence, acceptance, performance
- social and vocational rehabilitation (free market, if possible)

### Staff training

- knowledge of PWS-characteristics (e.g. problems like tantrums, skin picking, difficulty in adapting to new situations, hoarding, stealing, manipulative behaviour, stubbornness, self-injurious behaviour, many food-related behaviour problems due to an increased appetite and a lack of satiety, problems with the short-term memory)
- nutrition management in cooperation with the residential services or parents
- management of physical exercises
- behaviour management
- prevention (e.g. planning small steps, adapting the workshop place)

### Broad spectrum of a variety of workgroups (at Regens Wagner Absberg)

- garden/lawn care
- cleaning and house-keeping
- packaging
- agricultural work (cattle, fields, garden-market) and gardening
- metal working
- fitting and assembling (e.g. toys)
- electric assembly
- laundry
- computerized work
- creative production (e.g. pottery)
- sewing
- printing

The range for work is from simple repetitive work to complicated work with a lot of detailed and sophisticated steps.

#### Work structure

- regular breaks (lunch time, intermediate meals) and individual breaks for recreation
- a given time structure for work and breaks
- resting rooms for recreation (in case of head-ache or tantrums...)
- nutrition management (daily food plans; information what they bring with them for the breaks, how much money and how many cigarettes; find compliance, if they eat the intermediate meals on the way to the sheltered workshop, look for loop-holes,...); controls, if necessary (e.g. the lockers where sometimes the stolen food is hoarded)
- different workgroups (see above)
- recreation groups: two hours per week: such as belly dancing, Nordic walking, computer, drums, playing parts, singing, music, Veeh-harps, fishing, games, sewing, needle-work, pottery
- one group leader plus assistant who support (by monitoring, controlling, motivating and consistent consequences, by explanations in advance, by adapting the work situations, by smaller steps, in a preventive way, by nursing, by repeating,...)
- supporters for the groups and the people with PWS: a social worker and a psychologist (e.g. help to overcome emotional instability, crisis or behaviour problems or to improve the work processes)
- individual support plans plus documentation to improve skills or self-esteem, to enhance different aspects of the personality or to manage some of the behaviour problems
- remedial gymnastics, physiotherapy, lymphatic drainage due to scoliosis or oedema
- cooperation with the residential services, parents...

B. Presenter: Dorthee Pederson – Denmark

Title: At the Job

***“Every person has dreams and wishes for their future. How do we manage so their (the persons with PWS) dreams can come true?”***

This presentation describes the follow up on a survey from 2001 (unpublished) that included 53 persons with PWS above the age of 18 and living in residential homes. There were 45 questionnaires sent out to sheltered workshops all over Denmark and to the 4 workshops in Denmark for only people with PWS. The group homes for persons with PWS received questionnaires as well; 25 came back (60%) response. The objectives of the survey in 2001 were:

- To evaluate the existing offers of employment
- How many persons with PWS was employed and in what kind of employment?

- What kind of jobs were they offered?
- How much compensation for the specific handicap did they receive?
- What kind of compensation was given?

### **The Results:**

- ***Places of employment in 2001***
  - 2 persons was in supported employment
  - 12 worked in sheltered workshops
  - 22 in training centres with varying offers. 2 of them needed to have a person to support them through the day.
  - 14 worked in workshops for only persons with PWS.
  - 1 was in a special high school for persons with learning disabilities
  - 1 was unemployed
  - 1 worked in a special offer, where the person was the only one with two persons/staff as support.
- ***Sheltered workshops 2001: The staff in the sheltered workshops were aware of – but...:***
  - Attention and supervision according to food related issues
  - Need of constant support
  - The staff had generally few resources to support the person with PWS
  - The staff had generally little knowledge of PWS and not much time or resources to require the necessary knowledge. However, they had of course lots of hands-on experience in the work with the people with PWS.
  - Generally the people with PWS used their colleagues at the sheltered workshops to get food, money and so on.
  - Not much cooperation with group home/staff at the group home
  - Production work – production of medical equipment
- ***Visit in a sheltered workshop 2008 – the workshop participated in the survey 2001.***
  - In a group of 20 persons with different disabilities – 4 had PWS.
  - 2 staff members (in 2001 there was only one to the same group)
  - Both members of the staff was educated social educationists (in 2001 the staff member didn't have an education related to the work she did, and no knowledge about PWS)
  - Generally the work in this workshop is based on neuropsychological principles and relevant knowledge about each handicap represented. The staff used an "I'm okay – you are okay, but I make the rules" pedagogical basic.
  - Strict rules about food related issues and cooperation with residential home/staff was seen as essential in 2008.
  - Working hours are planned individually

- There is a salary once a month
  - Sport games and exercise is planned in working hours
  - The staff had attention on and supervision was given in social matters –according to the need of “translation” in social matters.
  - Production of medical equipment
- ***Workshops for persons with PWS only (2001):***
    - No salary
    - Daily structure related to the persons with PWS needs
    - Different kinds of activity
    - The staff had knowledge and lots of hands-on experience according to PWS
    - The staff had attention on and gave supervision according to behavioural and food related issues
    - Social training was practised
    - There were many conflicts and tantrums. The staff had plans for how to intervene.
    - Staff was educated social educationists
    - Everything concentrated about the group
    - Close cooperation with group homes/family
- ***Visit in a workshop only for persons with PWS 2008 – the workshop participated in the survey 2001:***
    - Here worked 7 persons with PWS
    - 2 educated staff members – social educationists
    - Working hours 8,15 – 14,45
    - Everyone had a day off during the week to clean up their room, pay their bills, go to the bank or to the doctor etc.
    - No salary
    - Daily structure related to the persons with PWS needs – individually
    - Individual supervision in food and behavioural issues
    - Training and supervision in making decisions for them self, to cope with conflicts and disagreements etc.
    - Activities is among other things; physical training, making handcrafts to sell on a annual market, visits in, and cooperation with the surrounding society , cooperation and participation in activities with sheltered workshops in the neighbourhood
    - Cooperation with group home

**General conclusions:**

- ***It is obvious that there has been a lot of things happening in the field of work for persons with PWS as well as for other groups of persons with learning disabilities:***
  - From group thinking to taking care of the individual in the group
  - The staff have today all a special education

- The staff knows a lot more about the syndrome in 2008 than they did in 2001
  - The staff knows the importance of cooperation with family/group home
  - In the workshop only for persons with PWS, staff have moved from “watching over” all the time, ready to intervention in conflicts, to learning the people with PWS to be their own decision makers, solve their own conflicts (of course with help if needed) and they are much more included in society and involved with other people.
- **Suggestions to guidelines for environmental structure in the field of work for people with PWS:**
    - The Staff must have knowledge about people with PWS
    - There must be individual supervision and rules according to food related, behavioural and social matters.
    - Daily structure
    - Respect for the individual – let them have a say in decision making and have influence – where they are able to-
    - Physical activities in working hours
    - Cooperation with everyone relevant – group home, family etc.

**C. Presenter:** Marguerite Rupnow  
**Title:** Work services For People with PWS

### Paragon’s Mission

- To establish programs that maximize the capabilities and potentials of the people we serve, enhancing their physical, emotional and social experiences as a result.
- We accomplish this through support and training by a concerned and caring staff that works cooperatively with each person, as well as their family/guardian, referring agency, other service providers and our community.

### Paragon’s keys to achieving its mission

- Emphasize individualized services
- Provide a full spectrum of services
  - Center based work services
  - Community Integrated enclaves
  - Transportation
  - Community based employment services
  - Supported employment services

### Work Services Include

- Center-based, piece rate production on a variety of tasks, including: packaging, assembly, laundry, inspection, labeling, shredding and basic machine operation
- Community integrated enclaves – small group of workers from the center based program work in larger community based work environment. Supported and supervised by Paragon staff.
- Supported employment opportunities in the community
- Majority of the work done is Sub-Contracted Work – which means that product completed is for another vendor to sell to retailer or to be part of another product.

### Paragon Work Services

- Assists people with disabilities to develop skills that will benefit themselves and the community.
- Individuals gain marketable job skills and work experience while learning to seek and maintain meaningful employment.
- Individuals gain self-esteem and the community benefits by an increased labor force of contributing citizens.

### Work Services Locations

- Oconomowoc, WI: 80-100 clients.
  - Cafeteria
  - Day Program Area
- Whitewater, WI: 12 clients.
- Duct-O-Wire Enclave: 8 clients.
- Oconomowoc Mfg. Enclave: 8 clients.

### Paragon and Prader-Willi Syndrome

- Paragon is currently the largest provider of work services to clients with Prader-Willi syndrome in the USA!
- Paragon works with an average of 77 clients with Prader-Willi syndrome per day
- Clients are involved in a full spectrum of work activities, ranging from pre-vocational work, to integrated enclaves, to supported employment in the community
- Paragon's facility, procedures and highly trained staff helps provide a safe and secure atmosphere where people with Prader-Willi Syndrome can achieve their full potential!

### Paragon's Vocational Development Center (VDC)

(All Services are provided with an emphasis on the individual's needs)

- Job Placement and Development
- Vocational Evaluation
- Work Experience
- Work Adjustment
- Job Coaching

- Supported Employment

#### Transportation

- PWHO provides transportation services to our Work Center in Oconomowoc & our two enclaves at Duct-O-Wire & Oconomowoc Manufacturing.

#### Paragon Candles Fundraising

- High quality candles manufactured by our clients.
- 15 different scents in 4 different sizes.
- Organizations receive 40% net sales of fundraiser.

### **IV. Define the nature of the problem and why it is important:**

Too many individuals diagnosed with Prader-Willi syndrome are placed in work environments that are inadequate and unprepared to support their unique and challenging needs. These unprepared and undertrained work environments set the stage for failure and frustration, which in turn decreases the ability for these individuals to feel a sense of accomplishment and lessens the natural desire to be an asset or productive member of society.

### **V. Nature of the Problem**

Across all cultures individuals with PWS struggle to find meaningful employment in an environment where the employer is willing to make environmental changes.

- Security surrounding Food, Money, & Personal Belongings
- Structure & Schedules
- Behavior Management Supports
- Motivational Approach to Skill Development & Production
- Rewards & Recognition

Due to the special environmental supports required to successfully transition these individuals into the workforce, work options are limited by their employer, and often are not given the same chance to explore different work opportunities.

Employer's limited education, knowledge and training on PWS creates frustration, and increases stress which in turn limits the individual's skill development up to and including hasty termination.

## **VI. Impact of the Problem**

For the individual diagnosed with Prader-Willi Syndrome (PWS) the inability to find supportive employment within a therapeutically safe environment has an extreme negative impact on their self esteem. Most individuals with PWS like the average person want to feel a sense of accomplishment and sense of purpose. As adults they seek the normalization of finding solid work opportunities and skill development which enhances the sense of purpose and social status in their communities. Lack of employment options greatly reduces this normalization as they transition into the work force. Inability to secure substantial work also creates financial instability and decreases their ability to provide themselves with necessary personal items.

Families struggle as their loved ones are dependent on structure and routine, and without adequate environmental work supports they have difficulty in providing the necessary twenty-four hour continuum of care. The emotional and financial burden it places upon the family also creates a stressful and sometimes unhealthy strain on them as a whole. For most families they experience a true sense of loss in regards to their inability to support or recognize their child's or siblings hopes and dreams.

Caregivers working in environments without the necessary training and supports increases stress which leads to poor choices and ineffective approaches to dealing with challenging behaviors. This inability to provide quality of care increases the risk of caregiver burnout, turnover, and an unstable work environment for the individuals.

Providers who are not allowed to develop programs with adequate environmental supports due to licensing/governmental restrictions, limited funding or training have a higher risk of providing poor or ineffective work/vocational programs. The inability to meet the client's individual needs creates a negative image up to and including possible legal or licensing sanctions. Without consumer trust the negative feedback may reflect the future growth of the agency.

The system of care becomes compromised when inadequate environmental supports are not put in place surrounding the person diagnosed with PWS. Underprepared or non-supported work/day services promote unhealthy choices in the individual, and places an unnecessary burden upon the family, the caregiver and the system as a whole.

## **VII. Discussion Summary**

The workshop participants heard consistent themes within the presentations and consensus of the group surrounding the lack of structured and supportive work environments available for a person with PWS.

It was easy to agree collectively on what was necessary in order to promote a healthy work environment for the person with PWS. Most successful programs were using creative approaches to enhance their participants/employees work experience to include some recreational, artistic and therapeutic outlets as well.

Creativity, knowledge and training were also key components when addressing the environmental and behavioral supports in community job placements. The more willing the employer was to adapt to the need the more successful the outcome.

Although the workshop participants came with a wide range of personal experience and knowledge there were no issues outside general consensus.

## **VIII. Guidelines for Best Practice**

*All guidelines should take into account the assessment of the individual to assure health and safety to promote quality of life.*

The individual's opinion should be taken into account when following these guidelines whenever possible.

In order to provide a safe, therapeutic, as well as productive work environment a thorough assessment should be completed on the individual with PWS prior to employment. The assessment should include a current and previous skill assessment. If there has not been one done recently this should be an initial step. Information on performance and competency testing should also be recorded. If the individual is transitioning from school or another placement a meeting with the parents, caregivers, school and workshop representative should be held in order to gather all the necessary information. It is also extremely important that the individual has the opportunity to have their personal hopes and dreams reflected and supported to the best of ones abilities. The assessment outcome should be written in a goal driven format to ensure that all members support and understand the necessary steps to reach the long term outcome.

People with PWS thrive in environments where there is a higher level of structure and predictability. Posting schedules, routines and other visual aides allows them to know what will be expected and what will be occurring throughout their day. If change is immanent allow the person the opportunity to process the information ahead of time in order to decrease anxiety. As change is difficult on everyone try and state information in a supportive manner and by asking for their assistance in addressing the change/need empowers them to make positive choices for a positive outcome. The posting and reviewing of a clear set of rules and expectations provides care givers with a structure for consistency and provides the individual with the knowledge with what is to be expected. Unwritten rules and expectations create an atmosphere of emotional uncertainty and an almost

immediate negative reaction. It is also apparent that environments that use proactive approaches to activities and noise which cause a sensory overload for some individuals, experience less emotional outbursts.

When providing work place or workshop employment opportunities for a person with PWS it is imperative that security efforts are in place surrounding food, money, and personal belongings. Plans should be well developed to ensure their personal safety and maximum benefit from the job placement. A managed food plan and a locked food storage system will decrease the anxiety of the person with PWS and increase their work productivity. In an integrated program it is imperative for safety reason that we are even more aware of these concerns. If an individual is placed in supportive employment they will require the ongoing support of a job coach or support staff in order to ensure personal safety, food security, and appropriate social boundaries.

Thorough PWS specific education and training will need to be provided to all individuals who are involved in the care and support for the person with PWS prior to the employment opportunity and ongoing there after in order to ensure overall safety, understanding, and guidance. For direct care givers individual specific information, medical concerns, and the therapeutic approach plan to prevent and support crisis situations should also be trained and reviewed ongoing to ensure follow through and consistency.

Workshops and other employment agencies need to be educated on the multifaceted health concerns related to individuals diagnosed with PWS. They should be provided with up to date medical alerts and have a medical crisis plan in place. A binder containing all necessary medical information and contact numbers which can be used in the event of an emergency is an effective tool for communication with medical professionals if the need arises. A system to manage medication needs to be in place and careful oversight to ensure administration or supervision of doctor's order. Many individuals with PWS require special attention to skin and wound cares. It is important that the work place be knowledgeable on the area of skin and wound care prevention and suggested plan for treatment or medical supports needed.

Due to the complexity of the syndrome, a person with PWS needs ongoing behavioral and emotional supports, which should be developed collectively through the individual assessment process, and through ongoing evaluation. An effective method of achieving consistency is through a carefully designed therapeutic approach plan which would also incorporate strategies to handle conflict resolution, preventative measures, and motivational approaches to enhance overall outcome objectives. This approach consistency helps reduce stress and increases self awareness and empowerment for the individual to make health choices. If the individual requires additional emotional support outside counseling may be necessary to assist the individual and the team support them. Some issues of concern maybe better supported in a contract

involving the person with PWS to come up with a support strategy that they feel best meets their needs.

Through the use of an individual support plan personal growth and development can and should be ongoing. Empowering the individual to play an active role in decision making and by providing choices enhances their emotional stability and the work/vocational environments overall effectiveness. Continued educational opportunities and ongoing therapy services should be considered, if there are financial resources and it is determined that the individual could benefit from future participation. Social skills development should also be included in the support plan identifying both areas requiring growth and methods to increase overall self awareness. Individuals with PWS are extremely outgoing however need support and guidance surrounding personal boundaries, communication and safety with others. By improving on their social skills it also enhances the individual's ability to have some of their wants and needs met in a healthy manner.

Programs incorporating opportunities for recreational social opportunities are encouraged. These programs help assist with self expression, artistic creativity, and an improved sense of self. Incorporating exercise or some type of cardiovascular activities also reinforces and promotes a healthy lifestyle.

## **IX. Conclusion**

All areas of support such as family, medical, psychological, therapeutics, education, legal and community need to work cohesively together in order to provide a continuity of services, which promotes an environment where the individual with PWS has the opportunity to live, learn, and succeed. An important key to this is to create a PWS supported work environment This opportunity must include safety and security, skill development, choice, an ability to promote positive self image, and allow them the personal experience of being a productive member within their community.

## **X. Challenges and items for future consideration:**

The most significant challenge presented was how to increase the limited number of choices and opportunities for individuals with PWS due to the insufficient number of programs/workshops/employment options available. Most communities have limited locations, lack of funding and governmental support, and poor personal resources to accommodate for the need. In addition, with the lack of experience and personal knowledge on PWS employers are fearful to adapt the necessary environmental supports necessary to create a healthy supportive employment.

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First International PWS Caregivers Conference  
Herne, Germany  
June 3-5, 2008

Workgroup S2: Aspects of Psychological Work with  
People with PWS

**I. Workgroup Leadership Team:**

**Presenters:**

Tony Holland, MD – UK

Palma Bregani, PhD – Italy and Irune Achutegui, PhD –Spain

**Facilitator:** Leopold Curfs, PhD – Denmark

**Scribe:** Linda Gourash, MD – USA

**Translator:** Mrs. Schmidt – Germany

**II. Workgroup Participants:**

Participants were caregivers and professionals representing Germany, Italy, UK, Russia, and the USA, and parents representing New Zealand, Finland, and Germany.

**III. Abstracts A and B:**

**A. Presenter:** Tony Holland, MD

**Title:** PWS Psychological Aspects

A1. 'Behavioural Phenotype' of PWS

- Eating disorder
- Repetitive and ritualistic behaviours
- Temper outbursts
- Skin picking
- Mood disorders and psychotic illness

A2. Research questions

- Why do such problems occur in excess in PWS and at what stage in development?
- Is it the same across genetic sub-types and both genders?
- Are there behaviours/psychiatric problems that occur in association with each other, e.g., skin picking and mood disorders?
- Do they have similar or different underlying mechanisms?

- Direct
- Indirect
- How are they best managed/treated?

### A3. Understanding the eating disorder

- Eating behaviour in PWS
  - Occurs in 100% of people with PWS
  - Some variation in intensity
  - Onset 2+ years of age
  - Persists throughout life (?lessens in later life)
  - Abnormality of satiety
  - ?Hypothalamic origin
- Major concerns related to PWS, diet and independence:
  - Risk of severe obesity as increasing independence leads to access to food
  - Availability of, and access to, money that can be used to buy food
  - Vulnerability of people with PWS to exploitation and abuse due to naivety and/or wish for food

### A4. Other Behaviours

- Population-based Study of PWS Obsessive Compulsive Symptoms:

<b>Symptom</b>	<b>PWS (n=89)</b>	<b>contrast (n=36)</b>	
Ask/tell	36/78 (46%)	4/29 (14%)	**
Routines	26/80 (32%)	4/33 (12%)	*
Hoarding	19/80 (24%)	1/33 (3%)	**
Repetitive	18/80 (23%)	3/33 (9%)	NS
Ordering	11/80 (14%)	0	*
Cleaning	2/80 (2%)	0	NS
Counting	0	0	
Checking	0	0	

Clarke et al 2002 BJ Psych; 180-358

- Repetitive, ritualistic behaviours, and temper tantrums:
  - Characteristic profile of behaviours – similar to but more severe than that found in early childhood in typically developing children;
  - Onset in childhood – may persist into adult life;
  - Best understood developmentally and as a combination of an increased propensity to such behaviours – these behaviours can in turn be shaped and reinforced by support strategies.

- Repetitive behaviors and hyperphagia occur via different mechanisms.
- Repetitive and ritualistic behaviours in PWS compared to autistic children:

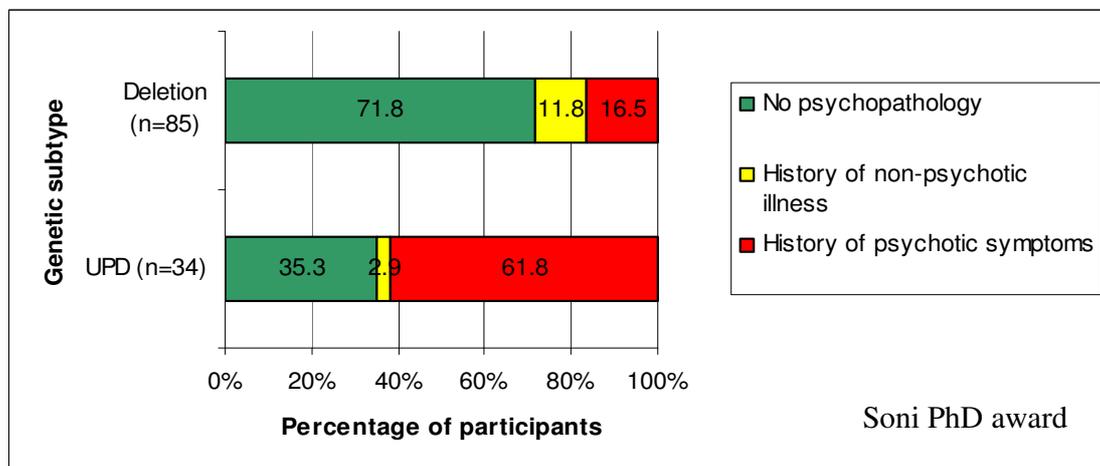
Childhood Routines Inventory			
	PWS N=80;		Autism N=89
Total score	13.1	(5.1)	14.1 (4.2)
Just right factor score	3.4	(1.6)	3.8 (1.4)
Repetitive factor score	3.6	(1.6)	3.8 (1.2)
Total freq/intensity	52.6	(16.6)	54.3 (15.6)
Just right freq/intensity	13.1	(5.2)	14.3 (5.1)
Repetitive freq/intensity	14.6	(5.8)	15.5 (4.7)

Strongly significant negative association between DQ and frequency/intensity scores in PWS less so in autism

Greaves et al, 2006 JIDR, 50, 92-100

#### A5. Affective disorder and psychotic illness in PWS

- Prevalence of psychotic symptoms:



- Psychotic illness more common in m-UPD than deletion  $p < 0.001$ , effect size 0.45
- Psychotic symptoms overall are mood and anxiety related not schizophreniform.
- Psychosis occurring among those with deletion condition appears to be the same as in UPD, although there is more likely to be a

- history of maternal depression, and non-psychotic mood abnormalities appear to be more common in the deletion condition.
  - There may be different genetic processes contributing to the etiology of these symptoms.
- Affective disorders:
  - Develop in later childhood or early adult life – unexplained increase in existing maladaptive behaviours or onset of new maladaptive behaviours;
  - Characterised by disturbance of mood – from rapid and brief fluctuations to more profound and longer mood disorder;
  - For some this may be associated with the development of hallucinations and delusional beliefs and ‘hypomanic’ mood swings;
  - Proper diagnosis – appropriate medication – environmental/psychological management

## A6. Management of behaviour

- Overview:
  - Understand the ‘mechanisms’ underpinning the behaviour.
  - Understand the different mechanisms for different behaviours or mental illnesses:
    - Eating
    - Repetitive behavior
    - Mood disorder
  - Effective intervention requires combining knowledge about *the individual and the syndrome*.
- Intellectual disabilities and challenging behaviour:
  - Etiological vulnerabilities:
    - Developmental
    - Biological
    - Psychological
    - Social
  - Factors:
    - Predispose
    - Precipitate
    - Maintain
  - Formulation: *The bringing together of information from a variety of sources and theoretical perspectives to arrive at possible explanations for the index problem that then informs the interventions.*
- Interventions:
  - Should be based on the formulation
  - May take different and varied forms including:

- Psychological and communication strategies
- Staff training
- Treatment of physical or psychiatric illness
- Should be evaluated by further observation and data collection

**B. Presenter:** Palma Bregani, PhD & Irune Achutegui, PhD:

**Title:** The role of the Psychologist

B1. Psychological evaluation is used for these purposes:

- Indications about psychological interventions
- Psychological data in medical case histories
- Requests for individual teachers or educators
- Welfare contributions, resources for the disabled

B2. Psychological approach to prevention or limiting PWS emotional disturbances

- Our program's aim:
  - To help both parents and caregivers to understand the emotional life of PWS, as well as their own motivation and feelings, upon which reactions are based.
  - To stimulate children and adolescents with PWS in their socialization skills and in the control of their emotions.
- Evaluation instruments and tools:
  - Interview(s) with parents
  - Developmental tests
  - Tests and questionnaires, for different aspects
  - Interviews with children and adolescents
  - Behavioural observations
  - Information from teachers and and/or other caregivers
- Therapeutic interventions:
  - Consultation/supervision with teachers, educators, caregivers, and volunteers
  - Psychological groups for children and adolescents
    - Goals:
      - i. To observe the children or adolescents while playing or interacting in the group to complete a psychological profile
      - ii. To identify, precociously, the symptoms of emotional disturbance in order to prevent its development

- iii. To stimulate acquisition of social skills and to help them acquire a positive self image
- Individual counseling for parents
- Psychological groups\* for parents:
  - The goals of the parent group are:
    - i. To better understand the emotional states of their children
    - ii. To overcome stereotyped perceptions of their children with PWS
    - iii. To understand their own motivations and feelings when interacting with them
    - iv. To become aware of the potential positive influence of their children's emotional well-being
    - v. To become increasingly able to accept criticism and new perspectives
    - vi. To learn new stimuli for their children

**Note:** Parents have the most important affective relationship with their children with PWS, and they are the most influential. The aim of the PWs parent group is to help parents understand their child and to help them acquire a capacity to cope with their child's difficult behavior. The group experience gives parents increased insight into themselves and into their child with beneficial consequences for their relationship with their child and each other. The child responds to changes in parents' attitudes by feeling less stressed and more secure.

- Research conducted by our program:
  - Self-injurious behavior in PWS (2001)
  - Relationship between cognitive development and dietary compliance in PWS patients (2002)
  - Trend towards autism in children with PWS (2006)
  - Sibling relationship with young PWS and their information about the syndrome (current study)

#### **IV. Nature of the problem**

- The role of the psychologist in serving PWS is largely undefined and varies from organisation to organisation.
- The workgroup participants are not aware of any literature on this topic.

#### **V. Impact on the Person**

- The psychologist provides behavioural programming.

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\* Based upon Michael Balint's model of group intervention

- The psychologist often negotiates individual contracts with their patients in the context of counseling them about their needs and opportunities.
- The psychologist is frequently in the role of individual therapist for the client with PWS providing a safe haven for discussing any topic, but in particular, issues concerning caregiving.
- As a confidante of the patient, with proper observation of confidentiality rules, the psychologist is an advocate for the patient's point of view with the treatment team and the family.
- The psychologist interprets the patient's behaviour. There was consensus that the patient's emotional experience needs to be further explored in order to fully appreciate the origin or contributing factors of mood and behaviour issues.

## VI. Impact on the Family

- Families of adult children in placement are frequently left out of the caregiving process.
- Persons with PWS cannot thrive without strong emotional support from the people most involved in their lives; caregivers and family members are the most important.
- The psychologist provides the parent with general information about the syndrome and with clinical insight into their child's thinking, feeling, and behavior.

## VII. Impact on the Caregiver

- The psychologist often assists in the education and training of caregivers.
- The psychologist is frequently the clinical supervisor of the caregiver.
- The psychologist is a consultant who may be externally employed or an in-house member of the treatment team.
- The psychologist has a role in providing supportive counseling to caregivers who are experiencing emotional challenges in caring for their residents. In particular, the psychologist provides emotional support when caregivers have experienced aggression or injury on the job.

**Note:** The relationship between the person with PWS and the caregiver has therapeutic elements. It is intense and complex. Because of the intensity of the relationship, the caregiver may see themselves in the role as advocate for the person with PWS. It is complex because the person with PWS may pull the caregiver into a more personal role and then devalue them. This process causes hurt feelings on the part of the caregiver that may take time to resolve. Although the person with PWS may hold grudges, it is more typical that their intense emotional interactions dissipate quickly; but, the caregiver may need extra time to recover.

## VIII. Impact on the System of Care

- The psychologist provides expertise in psychometrics (psychological testing) and interprets this information for a better understanding of the person by the family, school, and caregivers.
- The psychologist may be involved in educating the school personnel and workplace personnel – a role also provided by PWS associations.
- The psychologist has a role in developing and engaging in research on PWS.
- The psychologist who is involved primarily with clinical care has a role to be informed of the research and to interpret this for application for parents and caregivers.
- A psychologist with specific knowledge and experience with PWS is a valuable resource. Small provider organizations do not employ their own psychologist, and they must access a team provided by the National Health Service. Also, small organizations may be isolated from national PWS associations that could provide information and resources about the syndrome.
- The role of the psychologist may vary across different systems of care in different locales. For example, the psychologist in New Zealand interfaces with the team behavioural specialist who in turn interfaces with the caregivers of the person with PWS. In New Zealand, it is the behavioural specialist's role to talk and listen to the person with PWS.

## IX. Best Practices

- The work group reached consensus on these items regarding the role of the psychologist:
  - The psychologist plays a primary role when providing direct supportive counseling or therapy to the person with PWS.
  - The psychologist has a secondary role in providing emotional support and guidance to caregivers and family members.

*Both of these roles were highly regarded by the group.*

  - The psychologist can play a key role in facilitating communication among all stakeholders, including the person with PWS.
- The work group reached consensus on these items regarding the psychological aspects of management of the syndrome:
  - “All behaviour is a form of communication.” There was agreement that asking the person about their behaviour, depending on their communication skills, can be an effective means of understanding their behavior and helping him/her to avoid future behavioural incidents.

- There was agreement that an opportunity for apology and forgiveness is desirable after disruptive or offensive behaviours and that most people with PWS welcome and even seek out this opportunity.
- The team’s insight into behavioural incidents is the key to providing the necessary supports. Supports include the following strategies:
  - Identifying sources of stress and anxiety
  - Developing coping strategies including healthy avoidance
  - Relaxation techniques
  - Rehearsed asking for help (*please listen to me*)
  - One-on-one support and companionship in stressful situations
  - Positive verbal encouragement and praise
- ‘Time-out’ (TO) as a management strategy has the potential for abuse. There was agreement that time-out is an essential tool when used to support the person in calming down. There was similar agreement that time-out as a punitive tool in response to behaviour problems is counter-productive. There should be an effort to help the person learn to identify when they need to go into TO voluntarily.
- Safety comes first when managing disruptive behaviours – for the client, the other residents and the caregivers.
  - Replace breakable objects with safer objects (e.g., glass with plastic).
  - Distract, de-escalate and support early on
  - ***Please listen!***
  - Prevention
- As a member of the treatment team, the psychologist’s responsibilities to the patient with respect to confidentiality are at times complex. This is especially true if the psychologist functions as psychotherapist. In some countries (UK), the psychotherapist is not a member of the treatment team because of these confidentiality issues. It was suggested that the psychotherapist obtain permission from the person with PWS to share information with the treatment team.
- Parents provide both love *and* guidance to their child with PWS; they should be considered as part of the treatment team.

## X. Group Observations and Discussion

- What are the behavioural and psychological issues among persons with PWS?
  - The specific risks associated with PWS:
    - Obesity due to over-eating
    - Mental ill-health
    - Maladaptive behaviours

- The provision of support must include elements of both:
    - Choice (respect for autonomy)
    - Protection (risk for exploitation of vulnerabilities)
  - Persons with PWS have a capacity to ‘self-manage’ provided they are given good support; the amount of support may vary from person to person.
  - Blaming behaviours entirely on ‘PWS’ undermines the process of identifying and providing the level of support the person requires.
- Group process and discussion led to agreement on the following issues:
    - Understand the specific issues related to the person having both PWS and also an intellectual disability;
    - Understand the individual with PWS – his/her strengths and weaknesses;
    - Apply this knowledge to ensure the optimum support and the minimisation of potential or identified difficulties, and their effective management, if or when they occur;
    - Plan for the future – seek and engage the necessary interdisciplinary support and services to meet individual needs.

- Discussion questions:

1. Can a person with PWS reflect on their behaviour and benefit from insight?

Answer> The person with PWS has a limited ability to reflect on their behaviour to obtain real insight. They cannot internalize rules. However, learning how to behave more appropriately can be augmented with material incentives.

2. How can you give positive feedback to *caregivers* on how *they* are doing?

Answer> Have the caregiver obtain antecedent/behaviour/consequence (ABC) charts; collect and refer to this data (organizes staff). Often, food stealing is a staff mistake that should be dealt with privately.

3. Is the person with PWS responsible for their behaviour?

Answer> YES! “Give the person responsibility for their behaviour, and then give them support as needed.” However, food is a compulsion for people with PWS; they do not have the competence or capacity to control their food intake. Also, food is so highly rewarding, food stealing behaviour becomes its own reward.

4. Does the person with PWS know what the caregiver knows about their food stealing?

Answer> They lack perspective taking ability, and they need to blame others for the missing food.

5. What is the role of the psychologist?

Answer>

- To design incentive plans (token systems)
- To know how time-out or calming rooms are to be used in accordance with local regulations
- To talk to patients and develop contracts
- Team/individual coaching
- individual counselling

6. What are the human rights issues regarding the use of time-out (TO)?

Answer>

- Time-out is defined as *time out from positive reinforcement*; that implies that there is a positive reinforcement program ongoing.
- Staff cannot force a person into TO; they can encourage suggested separation or relaxation.
- It is best to teach the person with PWS when it is time necessary to use time-out or a calming room; then, they may respond to a verbal cue from staff *before* the situation becomes out of control.

QED

# INTERNATIONAL PWS CARETAKER'S CONFERENCE 2009 - HERNE

DEVELOPMENT OF GUIDELINES & STANDARDS

06. JULY 2009 – 08. JULY 2009  
HERNE, GERMANY

## Program

## 1. Conference day - 06.07.2009

<u>Time</u>	<u>Content</u>	<u>Moderator/ Speaker</u>
10.00–10.15	<b>Opening and first information about the conference</b>	Norbert Hödebeck-Stuntebeck (Germany)
10.15-10.45	<b>Greetings</b> 1. Mayor of the city of Herne 2. International Prader Willi Syndrome Organization 3. Regens Wagner Absberg 4. Diakonische Stiftung Wittekindshof 5. Prader-Willi-Syndrom Association Germany	Erika Wagner (Germany) Linda Thornton (Australia) Hubert Soyer (Germany) Irene Stenzig (Germany) Volker Holzkämper (Germany)
10.45-11.15	Lecture <b>„Self-determination and PWS”</b>	Leopold Curfs (Netherlands)
11.15-11.45	Lecture <b>“Behavior management”</b>	Janice Forster (USA)
11.45-12.15	Lecture <b>“Environmental structure of living”</b>	Hubert Soyer (Germany)
12.15-12.30	Introduction and information about the workgroups	Norbert Hödebeck-Stuntebeck (Germany)
12.30-14.00	Lunch	
14.00-16.00	<b>Start Workgroups A1 – A4</b>	Workgroup leaders
16.00-16.15	Break	
16.15-18.30	<b>Workgroups A1 – A4 (Continuing)</b>	Workgroup leaders
18.30-19.00	Break	
19.00-22.00	Leaving for the barbeque	

## 2. Conference day - 07. July 2009

<u>Time</u>	<u>Content</u>	<u>Moderator/ Speaker</u>
08.00-09.45	<b>Workgroups A1 – A4</b> (Continuing)	Workgroup leaders
09.45-10.00	Break	
10.00-11.00	<b>Workgroups A1 – A4</b> - work out the results	Workgroup leaders
11.00-12.00	Lecture: <b>PAM EISEN LECTURE</b>  Introduction by: Janalee Heinemann  Lecture given by: Tony Holland	Janalee Heinemann (USA)  Tony Holland (UK)
12.00-13.30	Lunch	
13.30-14.15	Lecture: <b>General health in PWS</b>	Susanne Blichfeldt (Denmark)
14.15-16.00	Start <b>Workgroups B1 – B4</b>	Workgroup leaders
16.00-16.15	Break	
16.15-18.30	<b>Workgroups B1 – B4</b> (Continuing)	Workgroup leaders
18.30-19.00	Break	
19.00-20.00	Dinner	
20.30–22.30	<b>Bus is leaving at 20.00</b> Cultural Program <b>Bus is leaving from Museum at 22.30</b>	

### 3. Conference day - 08. July 2009

<u>Time</u>	<u>Content</u>	<u>Moderator/ Speaker</u>
08.00-09.45	<b>Workgroups B1 – B4</b> (Continuing)	Workgroup leaders
09.45-10.00	Break	
10.00-11.00	<b>Workgroups B1 – B4</b> - work out the results	Workgroup leaders
11.00-12.00	Lecture <b>“PWS INTERNATIONAL LECTURE” PWS in India</b>	Prof. Jeyachandran (India)
12.30-13.30	Lunch	
13.30-14.00	Presentation of the results of the <b>workgroups A1 and B1 “Interpersonal relationship”</b>	Chair: Janice Forster (USA)
14.00-14.30	Presentation of the results of the <b>workgroups A2 and B2 “Self determination”</b>	Chair: Leopold Curfs (Netherlands)
14.30-15.00	Presentation of the results of the <b>workgroups A3 and B3 “Environmental structure of living”</b>	Chair: Hubert Soyer (Germany)
15.00-15.30	Presentation of the results of the <b>workgroups A4 and B4 “Behavior management”</b>	Chair: Norbert Hödebeck- Stuntebeck (Germany)
15.30-16.00	<b>Evaluation of the conference and Perspectives “Final Greetings”</b>	(Organization Committee)

# Workgroup program

## I. Block

**Workgroups: A1, A2, A3, A4**

Day one (July 06) and day two (July 07)

<b>Number</b>	<b>Title</b>	<b>Presenter, facilitator, scribe, translator</b>
A 1	<b>“Interpersonal relationship”</b>	<u>Chair:</u> Janice Forster (USA)  Jackie Waters (England) Barbara Withman (USA) Jonathan Smith (USA) Winfried Schillinger (Germany)
A 2	<b>„Self-determination and PWS”</b>	<u>Chair:</u> Leopold Curfs (Netherlands)  Margje Sinnema (Netherlands) Susanne Blichfeldt (Denmark)
A 3	<b>“Environmental structure of living”</b>	<u>Chair:</u> Hubert Soyer (Germany)  Jackie Mallow (USA) Gayle Benet (UK) Mary K. Ziccardi (USA)
A4	<b>“Behavior management”</b>	<u>Chair:</u> Norbert Hödebeck-Stuntebeck (Germany)  Irene Achutegui (Spain) Larry Genstil (Israel) Rob Pleticha (USA)

# Workgroup program

## II. Block

Workgroups: B1, B2, B3, B4

Day two (July 07) and day three (July 08)

Number	Title	Presenter, facilitator, scribe, translator
B 1	<b>“Interpersonal relationship”</b>	<u>Chair:</u> Janice Forster (USA)  Jackie Waters (England) Barbara Withman (USA) Jonathan Smith (USA) Winfried Schillinger (Germany)
B 2	<b>„Self-determination and PWS”</b>	<u>Chair:</u> Leopold Curfs (Netherlands)  Margje Sinnema (Netherlands) Susanne Blichfeldt (Denmark)
B 3	<b>“Environmental structure of living”</b>	<u>Chair:</u> Hubert Soyer (Germany)  Jackie Mallow (USA) Gayle Benet (UK) Mary K. Ziccardi (USA)
B4	<b>“Behavior management”</b>	<u>Chair:</u> Norbert Hödebeck-Stuntebeck (Germany)  Irene Achutegui (Spain) Larry Genstil (Israel) Rob Pleticha (USA)

**Second International PWS Caregivers Conference**  
**Herne, Germany**  
**July 6-8, 2009**

**Workgroup A1: Interpersonal Relationships**

**I. Workgroup Leaders**

**Presenters:**

Janice Forster (USA)

Winfried Schillinger (Germany)

Barbara Whitman (USA)

**Facilitator:** Jonathan Smith (USA)

**Scribe:** Jackie Waters (UK)

**II. Workgroup Participants:**

There were two workgroups of 13 and 11 attendees, respectively. The first group had more accumulated experience working with PWS. Both groups were comprised of parents, caregivers, providers, and professionals from Germany, New Zealand, the UK, Russia, Finland, Norway, Sweden and the USA.

**III. Introduction, Objectives and Abstracts: A, B, and C**

**Introduction:**

*Every culture has its own myth about why babies are born with Prader-Willi syndrome. In the Hindu religion in India, the syndrome is regarded as the final life stage in the progression to a God-like essence. There is also a creation myth that takes place before conception. It describes many souls who will be born with PWS arguing over who their parents will be...*

Prader-Willi syndrome is a unique developmental disability that has been characterized by hyperphagia and morbid obesity. But the mechanism that causes the hyperphagia affects other regulatory systems as well, resulting in deficits in adaptive behavior that require specialized, syndrome-specific interventions. Environmental adaptation is the only intervention that has been successful in reducing the morbid obesity associated with the syndrome. Group homes that offer a controlled environment have been life saving and cost effective. As a result, many adults with PWS reside together in group living situations. This does not occur in any other developmental disability with a specific genetic etiology. Everyone who has worked with more than one person with PWS recognizes that there are certain interpersonal characteristics that transcend temperament to make PWS unique among all of the developmental disabilities. Yet these interpersonal characteristics that are so important to shaping the nature of the relationships among individuals with PWS have never been described in the literature.

This workgroup on **Interpersonal Relationships** set out to answer the following questions:

- What are the neuropsychiatric aspects of PWS that lead to social skills deficits?
- What is the nature of the interpersonal problems in PWS due to these social skills deficits?
- How do social skills deficits affect courtship and sexual behaviors in PWS?
- What are the methods that caregivers have used to help persons with PWS acquire social skills and healthy interpersonal relationships?

The work group had two opportunities with two different sets of conference attendees to resolve these questions. These two sets differed somewhat in years of experience with the syndrome, but parents, professionals and caregivers represented both sets of groups. Janice Forster, Winfried Schillinger, and Barbara Whitman initiated the working process with a series of didactic presentations. A group discussion followed. One of the groups was given the task of solving a set of situational scenarios prepared by Jonathan Smith. Each of these scenarios pertained to a problem arising from interpersonal conflict among persons with PWS.

### **Work Group Objectives:**

- A. To describe the *capacity* for interpersonal relationships that is unique among persons with PWS
  - Strengths and challenges
- B. To explore the *nature* of interpersonal relationships among persons with PWS from the caregiver perspective
  - Itemize and prioritize problems
  - Discuss solutions
- C. To explore the *impact* of interpersonal relationships among persons with PWS
  - Person to person
  - PWS to PWS peer
  - PWS to non-PWS peer
  - PWS person to caregiver
  - PWS person to parent
  - PWS person to sibling(s)
  - PWS person to pets
  - PWS staff to PWS staff
- D. To develop best practice guidelines
  - Facilitating the development of interpersonal relationships
    - Managing problems with interpersonal relationships
    - Imposing reasonable safeguards
- E. To discuss factors limiting successful implementation

## Abstracts of Presentations

**A. Title:** The Neuropsychiatric Basis of Social Skills Deficits in PWS

**Presenter:** Janice L. Forster, MD - Developmental Neuropsychiatrist  
Pittsburgh Partnership, Pittsburgh, Pennsylvania USA

PWS is an imprinted, contiguous, multi-gene disorder that is recognized for hyperphagia and morbid obesity. Environmental management of diet, exercise, and activity schedules is life sustaining. As a result many adults with the syndrome reside in group homes where they achieve optimal function with support. Although food related issues appear to be of primary concern in the syndrome, there are cognitive and behavioral issues that challenge environmental management and complicate interpersonal interaction. The cognitive and behavioral phenotype is characterized by impulsive, oppositional and disruptive behavior; anxiety and insecurity; cognitive rigidity and inflexibility; and skin picking. The neuropsychiatric underpinnings of these behaviors are related to alterations of motivational drive, emotional regulation, cognitive processing, and communication abilities. The regulatory feedback deficits that result in hyperphagia have similar effects on neuropsychiatric processes. These alterations have a great impact on the development and maintenance of interpersonal relationships.

Motivational abnormalities apply to appetitive drives (reward based learning); any episodic behavior that is reinforced is likely to be repeated, and reverse learning (extinguishing previously rewarded behavior) is paired. In addition to food, this applies to the drive to acquire objects (collectibles as well as friends and romantic relationships); competitive drive (to be the best and have the most); and dogged persistence (to persevere when feedback becomes negative or reward disappears).

Processing problems result in sensory abnormalities such as pain and temperature perception, impaired cognitive perception of shame and disgust in association with body image, skin picking, and incontinence; and response latency manifested as noncompliance and shutdowns. Persons with PWS also display satiety deficits that apply to food as well as repetitive, perseverative behavior and never knowing when enough is enough (not knowing when to stop).

### **A. Summary: Neuropsychiatric Basis of Social Skills Deficits in PWS**

#### **Abnormal drives**

- Heightened reward mechanisms (appetitive drive)
- Learns well with reinforcement (episodic learning)
- Difficulty with "reverse learning"

#### **Impaired feedback**

- Satiety deficit: never gets enough
- Once something gets started, can't stop
- Excessive behaviors, rituals

#### **Processing problems**

- Decreased awareness of pain, incontinence, shame, disgust
- Repetitive behaviors: skin picking
- (other picking: nose, rectum, vagina)
- Perseveration, difficulty with transitions
- Apparent noncompliance, shutdowns

#### **Emotional dysregulation**

- Stress sensitivity, affective reactivity

#### **Cognitive Difficulties**

- Egocentrism, rigidity, single mindedness
- Poor judgment/problem solving
- Expects things to be his/her way; need for sameness

#### **Communication difficulties**

- Expressive, receptive and pragmatic language difficulty

Emotional dysregulation is manifest by affective instability and heightened emotional reactivity. But impaired emotional response is also noted with absent, delayed or idiosyncratic stress response, as well as apparent lack of empathy with inappropriate laughing when other's experience misfortune.

Cognitive difficulties include impaired judgment and diminished capacity, regardless of IQ. This is caused by single-mindedness and cognitive inflexibility, and egocentrism. Difficulty with perspective taking also interferes with social skills and adaptive problem solving. Finally variability in both verbal and non-verbal (pragmatic) language skills is apparent, and may relate to genetic subtype.

In summary, the cognitive and behavioral phenotype of PWS is related to underlying neuropsychiatric skill deficits that result in maladaptive behavior including social skills deficits. The need for environmental management of the syndrome results in residential placement, often with others who have the syndrome. Under these conditions the group home environment becomes an ideal situation for observing and characterizing the nature of interpersonal relationships, not only among persons with PWS, but between PWS-persons and their caregivers, family members, and other significant persons in their lives.

**B. Title:** Interpersonal Relationships in People with Prader-Willi Syndrome Based on Experiences with PWS Adults Living in Group Homes at Regens Wagner Absberg  
**Presenter:** Winfried Schillinger, Dipl.-Pädagoge (Univ.) Regens Wagner Absberg  
Germany

Deficits in social skills and behavioural problems have been reported often in people with Prader-Willi syndrome, besides the problem of hyperphagia. Yet the nature of interpersonal relationships of people with PWS has been poorly determined. The main factors influencing the interpersonal relationships of people with PWS in a negative way are egocentrism, poor social skills, difficulty

**B. Summary: Interpersonal Relationships in People with Prader-Willi Syndrome Based on Experiences with PWS Adults Living in Group Homes at Regens Wagner Absberg Germany**

**Factors influencing interpersonal relationships in people with PWS:**

- Poor social skills
- Difficulty in seeing something from a different perspective
- Language problems (reception, perception)
- Poor experiences with peers and social isolation in childhood
- Impaired judgement
- Autistic tendencies, compulsive behaviour
- Impaired self-reflection ability
- Weak control of frustration, aggression, emotion
- Mental retardation
- Egocentrism

**Categories of relationships:**

**— PWS person to PWS person**

- Poor real relationships
- Weak continuity, almost no long-term friendships
- Competition about privileges or social status, equal status only in one way
- Story telling to impress others
- Not accepting compromises, to be always interfering
- Profit seeking in relationships, often tendencies to a partnership of convenience
- Endless discussing and arguments without a real dialog
- Poor ability of conflict management and to solve problems, that often leads to escalation with temper outbursts, aggressions or self injuries

**— PWS person to other persons with disabilities**

- Profit orientated relationships, business connections
- Striving for power, try to reach a dominating position, to apply pressure e.g. with verbal aggressions
- Tendency of avoiding relationships with stronger people

*continued...*

seeing something from a different perspective, language problems, mental retardation, impaired judgment, autistic tendencies and compulsive behaviour, impaired self-reflection ability, and weak control about frustrations, aggressions and emotions. Additionally, poor experiences with peers and social isolation in childhood have a strong and lasting effect on difficulties in relationships.

There are 58 people with PWS living in group homes at Regens Wagner Absberg. Supervising this program has provided the opportunity and experience to observe interpersonal relationships among persons with PWS. In addition to the phenotypic behaviors associated with the syndrome, there seems to be a unique and recognizable characterisation of interactional behaviour affecting the relationships amongst PWS-persons, between PWS-persons and other persons with developmental disabilities, between PWS-persons and staff, and between PWS-persons and their parents.

The interpersonal relationships among PWS-persons are often characterized by poor real quality of relationship with weak continuity. Basically there are almost no long-term friendships among their peers. Also in the PWS group homes, it has been observed that there is a lot of competition about privileges or social status. Further, story telling to impress others and endless discussing and arguing are very common. The poor ability to use problem solving to manage conflict often leads to escalation of behaviour with temper outbursts, aggression or self-injury. The relationships of PWS-persons to persons with other disabilities are characterized by profit orientation, e.g., to get food or money. PWS-persons often try to reach a dominating position in these relationships, e.g., by applying pressure with verbal aggression. Also, PWS-persons tend to avoid relationships with stronger people who have other disabilities. In their relationships to staff, PWS-persons demand a lot of attention, persevere to routines, and search for limits by provoking. They show strong fixations on persons of authority, and they try to influence staff for their own interests.

In summary, the main observable deficits in social skills leading to interpersonal conflict among these 58 PWS-persons living at Regens Wagner Absberg were

**B. Summary (cont'd): Interpersonal relationships in people with Prader-Willi syndrome based on experiences with PWS adults living in group homes at Regens Wagner Absberg Germany**

**– PWS person to staff members**

- Demanding attention
- Strong fixation on certain staffs
- To persevere to routines
- Special reference to persons of authority (e. g. leading employee)
- Searching for limits, provoking and power play
- Staff as a reference-caretaker to trust
- Try to exploit staffs for own interests
- Rarely a continuity in relationship if a staff changed to another group-home

**Main deficits in social skills leading to conflicts:**

- Egocentrism
- To admit no delay of personal needs (cannot delay personal gratification)
- Poor problem solving strategies
- Poor self-reflection abilities
- Poor ability to talk about problems, emotions or needs

egocentrism, inability to delay personal needs, poor problem solving strategies, poor self-reflection abilities, and a poor ability to talk about their emotions, problems or needs. Future interventions focusing on these social skill deficits in PWS-persons might improve their capacity for making and sustaining interpersonal relationships.

**C. Title:** Sensuality and Sexuality in Persons with PWS

**Presenter:** Barbara Whitman, PhD Saint Louis University, St Louis, Missouri  
USA

Since the initial description, multiple endocrine system dysfunctions and deficiencies precluding normal growth and normal sexual maturation, along with small, immature genitalia have been cardinal features of Prader-Willi syndrome. Based on early descriptions of these abnormalities, coupled with evidence of absent or incomplete pubertal development, there has been an assumption of universal infertility. As a result, the importance of, and even more basically, an awareness of the role of sexuality and sexual behavior in the lives of people with Prader-Willi syndrome is only beginning to receive attention. The combined impact of sex hormone deficiencies and cognitive/social deficits have resulted in a long-held clinical assumption that sexual understanding, interest and activity rarely, if ever, reach that noted in non-affected pre-adolescents. Behaviorally, except for the rare female who has exchanged sex for food or the unwelcome aggressive sexual behavior among males treated with testosterone replacement, most clinicians have viewed affected individuals as essentially "asexual." Although many younger individuals frequently talk about dating, getting married and having babies, this has been seen as a learned socio-cultural behavior, not a manifestation of psychosexual maturation. Three coalescing factors demand a reassessment of previously held beliefs regarding sexual maturity, sexual behavior and fertility in this population: (1) recent reports of at least three successful pregnancies; (2) the experience of the author with three adults, two females and one male, all sexually abused as youngsters, who have demonstrated indiscriminate and predatory like sexual behavior; and (3) an apparent "cross-over" effect of growth hormone replacement therapy (GHRT) on sexual maturation with many treated adolescent girls developing menses, and treated males demonstrating virilization. This presentation will report on (1) the initial survey data from parents and community-based residential providers regarding sexual behavior in adolescents and adults with Prader-Willi syndrome; (2) if this behavior is more prevalent among those receiving supplemental hormone therapy or psychotropic medications; and (3) to investigate the policies regarding sexual behavior and sex education in group homes.

Parents and direct care providers of adolescents and adults with Prader-Willi syndrome were surveyed using two "informant observation" questionnaires: (1) the Sexual Behavior Questionnaire and (2) the Aberrant Behavior Checklist. Preliminary data document that virtually all affected individuals at some time or another engage in heterosexual handholding, hugging, kissing, and planning to get married and have babies. In an initial small sample, at least 50% of adult females are reported to have had at least one experience of intercourse and 50% of males are reported to masturbate.

Multiple factors demand a better understanding of "normal" or "baseline" sexual behavior among those with Prader-Willi syndrome including (1) extended life expectancy due to improved strategies for preventing or minimizing the complications of morbid obesity; (2) the use of growth hormone and sex hormone replacement therapy in both sexes; (3) the side effect of increased estrogenization from selective serotonin reuptake inhibitors widely used as adjuncts to behavioral management

strategies; (4) the recently reported pregnancies; (5) a shift in adult services from larger institutions to a more “normalized” community integration model, and (6) current “disabilities rights” legislation that endorses sexual activity, and advocacy groups that promote the rights of cognitively handicapped adults to engage in sexual activity in residential settings. These results suggest the need for much more attention to designing and delivering the appropriate sex education services for this population.

**C. Summary: Sensuality and Sexuality in Persons with PWS**

- Contrary to previous belief, some adults with PWS *are* interested in sexual activities.
- In a pilot study, all participants engaged in courtship behaviors.
- In a pilot study, adult females engaged in sexual activity at least once and some did with regularity.
- In PWS sexual behavior appears as a driven behavior parallel to that seen in the drive for food acquisition; it is not related to reciprocal emotional involvement or commitment.
- Males have engaged in masturbation but not interpersonal sexual activity.
- More studies are needed pertaining to physical maturity and sexual response in persons with PWS.
- There is a need for sex education and protection from sexually transmitted diseases (STD’s) and abusive relationships, BUT exposure to information can result in perseveration and acting out.
- Issues for further consideration:
  - Self-determination?
  - Decision-making about safety in relationships?
  - Who accepts responsibilities for consequences?

**IV. Workgroup Results**

**Objective A. Capacity for interpersonal relationships**

Strengths	Challenges
<ul style="list-style-type: none"> <li>• Desire for interpersonal interaction               <ul style="list-style-type: none"> <li>– Seek social proximity</li> <li>– Initiate verbal interaction</li> </ul> </li> <li>• Caring and nurturing qualities               <ul style="list-style-type: none"> <li>– Can demonstrate empathy for situations that have also happened to them</li> </ul> </li> <li>• Respond to learning and enrichment               <ul style="list-style-type: none"> <li>– Can learn from early experiences with family and friends, e.g., etiquette and social skills</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• Context-specific skill-sets               <ul style="list-style-type: none"> <li>– Can learn social skills, but may not exhibit them</li> </ul> </li> <li>• Require a safe, secure and trusting environment in which to learn</li> <li>• Need healthy, appropriate role models               <ul style="list-style-type: none"> <li>– Reactions learned from peers, TV soaps, etc</li> </ul> </li> <li>• Poor self reflection (inability to see themselves as wrong), requiring external input               <ul style="list-style-type: none"> <li>– Test limits</li> </ul> </li> </ul>
Strengths	Challenges

Strengths	Challenges
<ul style="list-style-type: none"> <li>• Social referencing <ul style="list-style-type: none"> <li>– Desire to be the same as peers or role models</li> <li>– Influenced by standards and behavior they see in others</li> </ul> </li> <li>• Memory for people and information about their life situations that can be used to initiate conversations (contextual)</li> <li>• Keen perception of interactional patterns involving others, especially dynamics of power and control</li> <li>• Persistence</li> <li>• Enjoy buying and giving gifts</li> <li>• Understand/demonstrate courtship behavior</li> </ul>	<ul style="list-style-type: none"> <li>– Hierarchical –“If I don’t get what I want, I will go to a higher authority.”</li> <li>– Difficulty internalizing standards and self esteem</li> <li>• Easily exploited</li> <li>• Egocentrism <ul style="list-style-type: none"> <li>– Difficult to teach/promote social skills in PWS-specific homes when everyone is self centered (does not play to their strengths)</li> <li>– Need to be better than others can lead to arguments, especially when unsupervised</li> </ul> </li> <li>• Shutting down/blocking out behaviour is common</li> <li>• Inability to tolerate uncertainty</li> <li>• Despite their syndromal similarities, each person with PWS has their own personality that reflects their unique background and development (may also need to be addressed).</li> </ul>

## Objective B. Nature of interpersonal relationships

General Problems	General Solutions
<ul style="list-style-type: none"> <li>• Hierarchical: learn power and control issues among people quickly</li> <li>• Provoke others: know what buttons to push</li> <li>• Manipulate</li> <li>• Shift blame</li> <li>• Seek attention</li> <li>• Reactive emotions</li> <li>• “Fickle:” relationships of convenience and context <ul style="list-style-type: none"> <li>– Drive to obtain food determines relationships and behavior in social situations</li> <li>– Relationships are not enduring</li> </ul> </li> <li>• Lack of respect for personal boundaries <ul style="list-style-type: none"> <li>– Steal from others</li> <li>– “PWS police:” intolerance; find fault with others; tattle</li> </ul> </li> <li>• Share with difficulty</li> <li>• Problem solving by proxy <ul style="list-style-type: none"> <li>– Use of parents to sort out problems</li> </ul> </li> </ul>	<p><u>Environmental interventions:</u></p> <ul style="list-style-type: none"> <li>• Co-ed residences have less same-sex squabbles</li> <li>• Use space to facilitate conflict resolution; designate a room for calming down</li> <li>• Use music to calm the environment and to relax the individuals</li> <li>• Restrict access: lock doors to bedrooms to safeguard personal belongings; staff supervision always</li> <li>• Periodic room searches; return belongings</li> </ul> <p><u>Proactive interventions:</u></p> <ul style="list-style-type: none"> <li>• Rules for everything!</li> <li>• Produce guidelines of rules for parents</li> <li>• Proactive contracting for behavioral interventions</li> </ul> <p><u>Feedback models:</u></p> <ul style="list-style-type: none"> <li>• Contain emotional outbursts; invite the person to move to another location to calm</li> </ul>

General Problems	General Solutions
<ul style="list-style-type: none"> <li>• Lie; tell stories (not universal)</li> <li>• One-way sense of fairness               <ul style="list-style-type: none"> <li>— Complain when a situation is unfair to them, but cannot perceive when they are involved in a situation unfair to others</li> </ul> </li> <li>• Pragmatic communication problems</li> <li>• Hearing acuity is keen, but selective; pick up/understand pieces of conversation and misinterpret the meaning</li> <li>• Referential thinking (listen to others' conversation and then believe that the information pertains to them)</li> <li>• Sexual or pseudo-sexual activity in inappropriate situation or place</li> </ul>	<ul style="list-style-type: none"> <li>• Encourage personal management of emotional control (See below)</li> <li>• Staff training-DO NOT REACT</li> <li>• Conflict resolution models               <ul style="list-style-type: none"> <li>— Resident counsel</li> </ul> </li> <li>• Restitution/apology</li> <li>• Explain one person's behavior to another</li> <li>• Write a "concern list" for later discussion with staff – concerns which are major in the morning may diminish by evening</li> </ul> <p><u>Analysis of behaviour</u> (functional analysis of behavior):</p> <ul style="list-style-type: none"> <li>• Identify triggers for lying and stealing</li> <li>• See the person's behavior as a form of communication of a need</li> </ul>

### ***Capacity to care for others***

A rare situation was shared with the group; the behavior of a woman with PWS who got pregnant and had the baby. The mother with PWS was delighted to tell others about her baby, and she appeared to thrive on the attention that she received. With respect to mother/infant interaction, she was not able to breast-feed, and she appeared to be more interested in her own meals than in feeding her baby. When the baby was taken away to live with relatives, she showed no emotion. (She does still visit the baby, however.)

### ***Lying, confabulation and story-telling***

When lying, the PWS person rarely sticks to the same story. The behavioural mannerisms associated with lying, e.g., eye-rolling, blinking etc, may be a manifestation of expressive language problems or a coping strategy while the person thinks of what to say next. Hence it is important to know the individual. There are two different types of lying, and both must be managed with a non-confrontational approach. Lying to avoid getting caught or to get out of trouble occurs commonly, and preplanned consequences will apply. Lying for personal gain requires a review of the person's situation and background in order to understand the cause of the behavior; then the underlying reasons can be addressed.

Confabulation is a type of "story-telling" – when the person makes things up because they cannot fill in the gaps. A non-PWS person who confabulates cannot remember what they have told to whom, so the stories are inconsistent.

The story telling in PWS is absolutely consistent in a given context; that is, the person with PWS will tell the same story to one person and may tell a different story to the next, but they always remember the stories they tell to whom and in what situation, and they don't get them mixed up. Also, they genuinely believe the story that they tell. So, it is difficult to determine if the story that they tell is true or false unless corroborative information is obtained.

## ***Stealing***

What is the basis for stealing? It appears to carry emotional significance to the individual. Often items that are stolen have no obvious material significance for the person themselves. Is stealing a manifestation of the “more is better” drive? Is the stealing opportunistic or planned in advance? What consequences are used if a person with PWS is caught stealing? Group homes had different strategies:

- If something is damaged, the person is expected to pay for it.
- If staff have set the person up to fail, e.g., by leaving a fridge open, then there are no consequences for the stealing, but staff can use this information to change their own behavior and to safeguard the environment.
- If the person has stolen from a shop, the person is escorted back to the store to take it back. Then, the PWS person must:
  - Return the item and also pay for it.
  - Explain to the shop manager why they stole the item.

Other contingencies include:

- Loss of a privilege; miss out on the activity of shopping the next time it occurs, or increased staff supervision is provided during the next similar activity.
- Natural consequences: the shop does not allow the person to come back

The problem of the *persistent* shop-lifter was described. The person was taken through the whole police and court process, only to have the case dismissed. When this occurs the person with PWS feels like they can do whatever they want, and they feel as if societal rules do not apply to them. This is not an adequate solution. The person with PWS must be held accountable for their behavior, even if the consequence does not match the severity of the incident. Many judges will indicate that the person is guilty of a crime and they are to be on probation for an extended period of time with the expectation for scheduled reports to the court regarding the person’s progress. When a person with PWS displays persistent shoplifting behavior, it is an indication that the person’s access is too great. Staff can help the person by limiting access to shops or by providing more supervision on outings.

## ***Access to money – solutions***

- Supervised access
- Money in own tin locked in the office
- Sign for money; require receipts to document expense always.
- Institute pre-entry contracts and agreements between the group home and the parents regarding money access and spending.
- Is important to recognize that each person’s capacity for money management may differ.
- If parents want the PWS person to have extra money, then the terms and conditions around amount, access, spending and accountability must be established.

## ***Hierarchical issues – solutions***

- Have a rule: “Staff on duty is staff in charge” – discuss complex issues later

- Involve a third person to diffuse conflict– but this can become a rewarding mechanism and inadvertently perpetuate the behavior
- Some staff *say* they have called senior management, *even when they have not*. This assurance may calm down the situation.
- Have the PWS person make an appointment to see a senior staff person or administrator on a regular basis; develop a concern list in advance to be reviewed at these meetings.
- Make sure the staff is trained on how to deal with hierarchical situations, such as when the PWS person says that they are going to call their parents or another authority figure.
- In general, authoritarian relationships poison everything.

### ***Helping people with PWS to manage their own emotional response***

- Use role-play sessions or social stories with person-specific problems and appropriate solutions.
- Pre-determine a point system or token economy in which the person is rewarded if they have coped in a situation where they would normally struggle (use points or tokens towards a larger reward)
- Use relaxation techniques such as progressive muscle relaxation (PMR)
- Use sensory techniques to help calm the person; other residents can use a hand roller massage to help the person calm down.
- Communicate clearly and directly with the person and use low expressed emotion

#### **– Using visual cues:**

- Use a series of coloured cards (visual cues) to hold up: Cope – Get a grip – Chill out
- Use visual cues, and use *low expressed emotion* (Staff must manage their own emotional response.)
- Empower the person to recognize when they should have “time out” and cue this with the rehearsed use of an “escape card”

#### **– Using environmental factors:**

- Change staff or have staff leave the situation
- Many group homes have a room designated as a calming room. One group home has several rooms painted in different colors to correspond with management of feelings; there are red yellow and green areas. The green room facilitates the expression of feelings.
- One group home residence uses exercise and change of venue as an intervention. On the day following an outburst, the person may accompany staff on walks, but their usual activity schedule is suspended to give them an opportunity to *reflect* on their feelings and their behavior.

### ***Provoking others – “pushing buttons” – solutions***

Sometimes this behavior is related to the person’s need for control; sometimes they start trouble to focus the attention away from themselves.

- Train staff so that they know not to take things personally

- Give the persecuted person coping skills e.g., “putting on their body armor”
- Teach persecuted person visual imaging, like putting hurtful comments into a trashcan.
- Divert, redirect attention to something else
- Low expressed emotion (staff must use/model/teach emotional *continence*)

### ***Suggested guidelines for sharing and socialising***

Most people with PWS enjoy board games; this can be a useful way to teach turn-taking, social skills, and tolerance of losing. Groups of 2-4 players can play together, but don't expect that they will be able to play with partners. Games of chance and those requiring the roll of the dice permit staff to say “the dice will decide.” The electronic Wii game can be fun, competitive, and the Wii Fit offers exercise activities. Other games that have been used successfully include cards, board games, Bingo, and dice games.

Some persons with PWS enjoy sharing social company while they are involved in a solitary activity such as fishing. Other persons have enough motor strength and coordination to enjoy sports competitions. A group member described their experience with a PWS soccer team where all of the PWS players wanted to play the same position! Above all, people with PWS enjoy sharing humor!

Music therapy has been explored. Some sing very well and display both pitch and rhythm. Others do not, but they have demonstrated that they can learn. Dancing can be enjoyable as most people with PWS have a sense of rhythm. Dancing promotes socialisation, motor skill development and sensory motor integration.

In the UK some group home residents enjoy going to the pub together. They save-up their money and talk about looking forward to their scheduled weekly outing to the pub. When they arrive at the pub, they order their drinks, they drink them down quickly, and then they are ready to go home; the social aspects of the encounter do not occur. The calories for the alcoholic drink are factored into the daily calorie allowance. The number of drinks is limited and known by the person and their staff beforehand. No undue effects of moderate alcohol intake by PWS-persons were observed by the workgroup. However, two anecdotes were shared. One workgroup member told the story of a PWS-person who drank an entire bottle of Bailey's Irish Cream and demonstrated no signs of a hangover the next day. Another workgroup member said that a PWS-person nearly died of alcohol poisoning. Vomiting did not occur in either case.

Recommendations for sharing and socializing include:

- Have a rule of no lending or borrowing, as this inevitably results in disappointment.
- Staff will always be required to teach and monitor the rules of the game.
- Social skills do not come naturally to persons with PWS, and they may require instruction (scripting), modeling, cueing and supervision by staff.

### ***Rigid “black and white” thinking – solutions***

- Don't commit to anything you can't carry out.

- Don't say something definite unless you are absolutely certain that it is!

**Tattling – “the PWS police” – solutions**

Tattling refers to telling staff about other residents' behaviours. The group considered that this behavior may result from a PWS-persons feeling of uncertainty or insecurity. The PWS-person may feel that a behaviour must be reported in order to maintain their own security. This behaviour may also reflect the PWS-person's absolute need for fairness, and their inability to understand that some rules are subject to interpretation. When tattling occurs:

- Reassure the PWS-person that the issue will be addressed.
- Reaffirm that it is staff's job to deal with the problem.
- Depending on the circumstances of the situation, you might thank them for bringing the problem to your attention, or you might ignore them.
  - Sometimes the person just needs to know that they are right, and staff will say, “You are correct.”
  - Sometimes the person should be told to “Mind your own business!”

**The need to be correct (having to be right) –solutions**

- Staff need to realize that quite often the person with PWS is correct! They may know the rules better than anyone else. But they are not always able to understand the exception to the rule.
- Difficulties emerging in peer-to-peer conflict – “You are both correct.” Both are right because they are looking at an issue from different perspectives.

**Manipulative behaviour - solution**

- Staff tend to see all behaviour as manipulative. But for the person with PWS, it is a survival skill. Staff may feel manipulated; hence it is staff's problem.
- What is the issue behind the behaviour? Is it an issue that the staff has created? If so, staff training is needed.

**Objective C. Impact of personal relationships**

**PWS Person to PWS Person**

Problem	Solution
<ul style="list-style-type: none"> <li>• Conflict: “picking” at each other; getting into each other's business; (toxic effect on the group)               <ul style="list-style-type: none"> <li>– Poor ability to manage conflict or resolve problems, leading to escalation with temper outbursts, aggression or self injury Tension/hyper vigilance around persons with known tendencies to be aggressive and challenging</li> <li>– Competition</li> <li>– Not accepting compromises</li> </ul> </li> <li>• Status-seeking relationships select for:</li> </ul>	<ul style="list-style-type: none"> <li>• Discuss the problem and the solution in the group context</li> <li>• Send conflicted peers on a preferred activity together</li> <li>• Change the environmental context for a day</li> <li>• Channel interpersonal competition and status-seeking into games</li> <li>• Use peer pressure to maintain conformity</li> <li>• Some people just need to have conflict in their lives; find out why.</li> <li>• If one resident is abusive to another, move</li> </ul>

<ul style="list-style-type: none"> <li>– Intellectually able peers</li> <li>– Humorous peers</li> <li>– Story telling peers</li> <li>– Dominant peers; sometimes the most aggressive one is the dominant one</li> <li>• Status-avoiding relationships: <ul style="list-style-type: none"> <li>– Person with behavior problems is excluded; especially if the group has received a consequence as a result of that person's behavior</li> </ul> </li> <li>• Poor <i>real</i> relationships <ul style="list-style-type: none"> <li>– Weak continuity; almost no long-term friendships</li> <li>– Profit seeking in relationships; “partnerships of convenience”</li> </ul> </li> <li>• Poor social communication skills (poor pragmatics) <ul style="list-style-type: none"> <li>– Endless discussing and arguing without a real dialog</li> <li>– “Talk at, not talk with”</li> <li>– Residents who have known each other a long time have ritualised or “scripted” dialogues</li> <li>– Volume of speech gets loud, shouting over one another</li> <li>– Interrupt each other</li> <li>– Laugh inappropriately at discrepant events</li> <li>– Need to have the last word, not knowing when enough is enough</li> <li>– Competition: “My day has been better; I did something more important than you did; “Listen to what I did.”</li> </ul> </li> </ul> <p>Tension and anxiety they feel when they interact is a function of the uncertainty they feel about the interaction, and it results in being over-focused on details.</p>	<p>one of them to another house</p> <ul style="list-style-type: none"> <li>• Script and model social skills</li> </ul>
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**Stalking**

Stalking is the obsessional pursuit of another person. It can happen in the group home, in the work place, or in the community. The object of the stalker can be a staff person, a teacher, a peer or a community person. It can also happen at a distance through excessive calls, texts, e-mails and letters.

- Look for the trigger in the perpetrator – is there stress that needs to be addressed?

- Allow natural consequences such as report to the police, but talk to the police ahead of time to determine a plan of action to deter the behavior
- Move one person to another group home as a last resort – could be either victim or perpetrator
- “Couples therapy” with psychiatric or psychological help using problem-solving/meditation
- Empower the victim to say no
- Set limits and provide distractions for the stalker

**PWS Person to non-PWS person (who has learning disabilities or intellectual deficiency)**

Problem	Solution
<ul style="list-style-type: none"> <li>• Dealing/bargaining/brokering of valuable items (mobile phones, play station games) for food or tobacco products (inequality of value of exchange)               <ul style="list-style-type: none"> <li>– Profit-orientated relationships, “business connections”</li> <li>– Striving for power; trying to reach a dominating position; applying pressure with verbal aggression</li> <li>– Tendency to avoid relationships with stronger people</li> </ul> </li> <li>• Romantic relationships can be a problem               <ul style="list-style-type: none"> <li>– Exploitation of the person <i>with</i> PWS</li> <li>– Stealing from the person <i>without</i> PWS</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• If brokering is discovered, discuss with the person</li> <li>• Weigh regularly to ensure extra food is not being obtained. If weight is increasing, find/secure the source of extra calories. As last resort, restrict diet and/or activities.</li> <li>• Risk assessment &amp; safeguards (see below)</li> </ul>

**Risk Assessment and Safeguards**

A risk assessment regarding sexual/romantic relationships determines what is the risk for abuse and/or exploitation, what is the risk for emotional injury or disappointment, and what are the potential outcomes of the relationship. Some individuals are not able to handle the intensity of a romantic relationship because of the uncertainty inherent in all interpersonal relationships. Sexual activity is an index of intensity. Sexual interest/activity can be seen as an example of the drive abnormalities in PWS; it may be latent, but once it begins, especially if it is reinforced, the capacity to know when enough is enough is impaired, and the context of mutual satisfaction in the relationship is lost. There are no studies examining capacity for sexual response in PWS, but it is likely that sex hormone replacement will increase sexual drive.

- Establish an action plan for when things go wrong and involve the person in the process
- Agree with the person in advance about desired outcome as well as potential negative consequences
- Be honest about outcomes for not keeping to the contract and carry them out (loss of privilege)

- Staff remain in earshot and monitor the situation (chaperone)

### PWS Person to Caregiver

Problem	Solution
<ul style="list-style-type: none"> <li>• Manipulation:               <ul style="list-style-type: none"> <li>– “Pushing people’s buttons”</li> <li>– Tells a new staff member a lie, e.g., he has not received his food/cigarette allowance</li> </ul> </li> <li>• PWS person appeals to a higher authority               <ul style="list-style-type: none"> <li>– Almost savant ability to understand the power and control issues in relationships (like an organizational puzzle)</li> <li>– Parallels behavior with parents</li> </ul> </li> <li>• Always demanding attention               <ul style="list-style-type: none"> <li>– Competition among persons for staff time</li> </ul> </li> <li>• “False social contract:” Caregivers have social attachment skills and naturally assume a trusting relationship. “They wouldn’t do that to me...”</li> <li>• Selecting favorite/non-favorite staff, but no depth to the relationship</li> <li>• If staff leaves, there is fleeting emotional response; there is no sadness, no depth to the</li> </ul>	<ul style="list-style-type: none"> <li>• Dealing with inconsistency between shifts:               <ul style="list-style-type: none"> <li>– PWS person: I was allowed to do that last time</li> <li>– Staff says, “ We are going to do it my way this time. Then, we will go back and check; if I am wrong, I will apologise.”</li> </ul> </li> <li>• Shift manager is the boss: “Staff on duty is staff in charge.”</li> <li>• Lie! (God will forgive you.) Say that you spoke with the authority figure, and they said staff was correct.</li> <li>• Regular staff meetings to review client behavior to assure that staff are not being manipulated and staff respond consistently</li> <li>• Phone parents when things are going right as well as wrong – schedule regular calls</li> <li>• Don’t tell the person with PWS if a staff member is a volunteer; otherwise, they may not be respected.</li> <li>• Avoid triangulation to limit the potential for manipulation</li> <li>• Consequences should be predetermined, and if there is any question, the manager should be the person who determines them; this maintains good working relationships with the key workers, who can still be supportive to the individual.</li> <li>• Interference from a higher authority (parent/manager) will only reinforce the behaviour</li> <li>• Structure activities</li> <li>• Limit duration of 1:1 time together with staff, or limit the nature of the interaction, e.g., 5 questions</li> <li>• Assign a primary staff and establish a regular 1:1 meeting time</li> <li>• Have the person write down concerns and schedule a weekly meeting to review them</li> <li>• Establish a resident counsel</li> <li>• Have a plan for dealing with triangulation</li> </ul>

Problem	Solution
<p>attachment.</p> <ul style="list-style-type: none"> <li>• Acute hearing is common, but... <ul style="list-style-type: none"> <li>– At times they know things they shouldn't</li> <li>– They pick up/understand pieces of conversation without context, misinterpret, and cause distress or trouble.</li> </ul> </li> </ul>	<ul style="list-style-type: none"> <li>• Schedule a preferred activity with non-preferred staff</li> <li>• If the client shuts out a key staff worker, another colleague should step in.</li> <li>• If the client divulges information about staff that they should not have been privileged to, ask "What makes you think that?"</li> </ul>

### **PWS person to parents –**

The relationship of the person with PWS and their parents is a long-lasting, constant relationship characterized by attachment and love. Because of this lifetime of experience, the parent knows their child better than anyone else. (For example, they may know how to calm violent behaviour.) As advocates and guardians, parents have an active role to play throughout their child's life, even though their child resides in a group home. Sometimes it is difficult to negotiate the transition of moving from home into a group residence. Parents may have mixed feelings about giving up their caretaking role to another, and they may be confused about what their new role in their child's life will be. The clarification of expectations by all persons involved helps make this transition and emotional adjustment more successful. Ongoing communication between the parents and the group home is essential for a smooth working relationship, and this facilitates the adjustment of the person with PWS. In the sad event of the death of a parent, the magnitude of the person's grief response may be a function of loss of activity shared with parent and not a reaction to the loss of the relationship with the parent.

Parents complain of the loss of their child's social etiquette when they leave the parental home. An example of this is polite eating behaviour, i.e., not eating with fingers. Caregivers should eat together with PWS persons to serve as role models and to provide reminders to eat appropriately. Eating with caregivers also provides an opportunity for modelling social interaction during a meal. Setting the table for a formal dining experience (like one finds in a restaurant) will provide the *context* to encourage appropriate mealtime behaviour in the group home, during home visits and during community dining experiences. Staff who have a low tolerance for negative eating behaviour should be trained to use low emotional response when prompting with verbal cues. Staff should always set a good example.

Caregivers described several examples of how adult persons with PWS display the internalization of parent's expectations and ideals. One person was not able to find the words to explain his sadness. He displaced these feelings onto the birds outside the group home; he said that the birds sing when they are sad. Then he told his staff that his father didn't like it when the birds sing. He said, "My father doesn't want the birds to sing." The story of the birds describes how the PWS person used psychological defense mechanisms to address and discuss his feelings of sadness.

He knew that his father did not want for him to be sad, so he displaced these sad feelings onto the birds.

Problem	Solution
<ul style="list-style-type: none"> <li>• Parents have the higher (highest) authority for establishing expectations for their children, but their children can use this for manipulation against staff.</li> <li>• Contact with parents is usually initiated by their child, and may be characterized as excessive or over involved by using a mobile phone to call home several times a day.</li> <li>• Failure to display a typical grief response after the death of a parent; grief can present as disappointment, in terms of what activities they will not be able to do with the parent any more.</li> </ul>	<ul style="list-style-type: none"> <li>• Staff must form a relationship with parents               <ul style="list-style-type: none"> <li>– to clarify expectations proactively;</li> <li>– to communicate regularly to review concerns; and</li> <li>– to notify about accomplishments as well as problems.</li> </ul> </li> <li>• Parents must be given guidelines about how to resist manipulation by their adult child               <ul style="list-style-type: none"> <li>– Advise parents not to continue talking about an issue of concern; it reinforces potentially false information.</li> <li>– How to deal with over involvement</li> </ul> </li> <li>• Staff help to maintain a relationship between the adult PWS person and parent(s) by prompts               <ul style="list-style-type: none"> <li>– To buy gifts for birthdays, holidays</li> <li>– To remember scheduled telephone calls</li> </ul> </li> <li>• Staff may ascertain parent's wishes regarding etiquette and polite social behaviors</li> <li>• One group home has a rule that there are no parental home visits during the first 3 months following admission</li> </ul>

### **PWS person to sibling –**

The relationship of the PWS person with a sibling is the longest lifetime relationship. However, siblings have not had the care giving experience of their parents, and sadly, many become involved *after* the death of a parent.

Problem	Solution
<ul style="list-style-type: none"> <li>• Distant relationship; usually only important if their parents die</li> <li>• The relationship with a sibling can be nurtured to be important and relevant</li> <li>• Siblings may wish to take the person out for visit, but things go badly; this affects future visits.</li> </ul>	<ul style="list-style-type: none"> <li>• Structure activities with siblings</li> <li>• Inform siblings about management strategies because they may not know what to do!</li> </ul>

### **PWS person to pet –**

Many group homes have animals as pets. One home has a scheduled rotation for the care of their dogs. Another group home has an arrangement for residents to walk dogs from the local animal rescue centre with staff supervision. A rural group home has a variety of animals. A farm animal was killed deliberately because the person with PWS thought that it was being mean to another, favoured animal. In another situation, a person with PWS put another's animal in the clothes drier.

One PWS person had a parakeet, and she learned to feed the bird and clean the cage appropriately. But when she got angry, she threw the birdcage (with the bird inside) onto the floor. To safeguard the bird, it was taken to the resident manager's house. The PWS person can visit the bird there; she knows that the bird belongs to her, but she also knows that she may injure it, if it remains in her possession.

Because persons with PWS can be violent towards animals, a risk assessment needs to be done to determine what safeguards need to be in place when the person with PWS is around pets. Ongoing supervision of the PWS person may be required to prevent them from eating the pet's food (even birdseed) or the overfeeding the pet.

In summary, although nurturing and caretaking behaviors can be displayed by many individuals with PWS, many persons with PWS lack the attributes to successfully to care for animals as pets because of the:

- Inability to recognize the needs of the pet as different from their own (overfeeding the pet or eating the pets food)
- Tendency to treat the pet as a possession, not a living creature;
- Use of the pet as a proxy for anger or retaliation; and
- Risk of mistreatment or harm to the animal.

Therefore, each person's capacity must be assessed, and a contract with guidelines and contingency plans must be established. The PWS person must know in advance that they will lose the privilege if undesirable things happen:

- Be honest about outcomes for not keeping to the contract (and carry them out)
- Even with a contract and an agreement, staff must always supervise and monitor the situation

### **PWS staff to staff interactions –**

<b>Problem</b>	<b>Solution</b>
<ul style="list-style-type: none"><li>• Persons with PWS "read" others emotions, even though they are not insightful into their own; staff "issues" become stressful to them.</li><li>• One staff talking about another in front of a person with PWS sets a bad example; it will polarize the entire group and result in increased manipulation</li><li>• Staff –staff disagreements are toxic to the living environment.</li></ul>	<ul style="list-style-type: none"><li>• Make a rule: "Talk <i>to</i> staff not <i>about</i> staff"</li><li>• Staff must resolve their differences; if they cannot do this together, then the issue must be brought up at a staff meeting for everyone to discuss and resolve.</li></ul>

## Objective D. Best Practice Guidelines

**Assumption:** Among persons with PWS, relationships of any sort are constrained by the “genetically and neurologically” determined incapacity for reciprocal relationships. The capacity for emotional understanding is further limited by a lack of opportunity and experience. Thus, relationships in PWS tend to be fleeting and built on shallow emotional connections; they reflect a primitive and tenuous capacity for object relationships. Thus people can be easily substituted in order to attain basic needs and satisfy basic drives. Unless managed, relationships are stressful. So relationships on all levels need to be externally managed and supported to reduce stress.

### Overall guiding principles:

- A structured environment with appropriate boundaries and limits is essential.
- Well-functioning teams who provide structure and consistency must be maintained.
- Social environments must be managed just like food and behavior.

### General guidelines:

- **For the individual:**
  - Know each individual well; they are a PERSON who happens to have PWS.
  - Try to make the person a part of the decision making process with mutual goal setting (does not work for all individuals).
  - Conduct a skills assessment to identify the PWS person’s strengths and weaknesses.
  - Identify social and interpersonal goals in the individual’s service plan:
    - Determine a way to measure progress.
    - Review and revise goals at least every year.
  - Plan on how to address skill deficits and needs:
    - Inform the PWS person what the goals are and how they will be reached.
    - Schedule interventions and rehearse skills.
  - Build on strengths such as contextual learning and rote memory ability, and use those strengths to teach better interpersonal and social skills.
  - Provide visual supports for all areas of communication (e.g., feeling charts, activity schedules, and to-do lists using language that the person can easily understand).
  - Articulate appropriate rules around communication (talk to them, not about them).
  - Conduct a risk assessment for some interpersonal goals.
  - When a person perseverates on a non-reciprocal relationship (or stalking), limit access to the other person.
  - For “complainers,” have them make a concern list with scheduled times for discussing items, with a rule that issues cannot be discussed at other times.

- Facilitate the expression of problems:
  - Repeat back what they say back to them to ascertain if that is what they meant.
  - Write down the problems so the person can see them in words.
  - Use “feeling charts” to identify emotional states.
  - Use expressive arts to facilitate emotional expression.
- When asking a person with PWS a question, be specific rather than open-ended to get the most accurate information.
- Specify rules and behavioral expectations; write them down for the person to see.
- Broaden social boundaries:
  - Provide measured, enriched experiences based on interactions with other group homes and resident peers.
  - Base social interactions on common interests.
  - Explore personal interests through variety of activities.
- People with PWS don’t have the internal skills to manage conflict, so it must be managed for them.
- **For the residence:**
  - Have house meetings once per week to discuss problems, make/review rules, and plan activities. Over a period of years, one group home has managed to get residents together without staff, and one person is elected to represent the group at the staff meeting. This is an opportunity to coach the individual on how to register a complaint appropriately.
  - Once a cohesive group is established, use peer pressure to modify behavior.
  - If available, use an external advocate for an apparently irresolvable conflict.
  - Build a community identity for the residence.
  - Teach skills on how to manage conflict with appropriate feedback.
  - Make the person with PWS part of decision making process, both at the individual level and residence level (e.g., resident councils) – phase out staff participation, but always maintain monitoring.
  - Individuals under stress also stress the group – staff must attend to both.
    - Help the person on an individual level.
    - Help the group recognize and understand the person’s problem.
    - Maintain the consistency, structure and predictability of the living environment.
  - When interpersonal conflict arises, plan pleasurable activities together so the interpersonal context is different, and each person gains a different experience of the other.
  - Expand opportunities for social experiences with regularly scheduled, structured activities with people from other settings and group homes through birthday parties (without food), social visits, “dates” and even sleep-overs.
  - For “shy” individuals or “outsiders”, address skill deficits or provide alternative pleasurable activities.

- Sometimes people don't want to go to social events because people don't talk to them – role play in advance how to help them interact.
- Schedule 1:1 social activities with staff, but give added reinforcement if a friend is invited.
- Schedule preferred activities with less preferred staff to facilitate relationship-building.
- Ultimately, conflict must be discussed and resolved between the two people in question.
- **For the parents:**
  - Develop a contract of expectations between residence staff and parents.
  - Phone parents when things go well just as do when things problematic.
  - There is always a kernel of truth in story telling – the trick is to get at that truth.

## **Objective E. Factors limiting successful implementation**

- Lack of social or communication skills
- Psychotic illness or psychiatric illness such as depression
- Poor training, lack of knowledge on part of staff
- Dominant person in the group who abuses power
- Fear
- Fractured team and dysfunctional staff relations e.g., inadequate leadership, inadequate support, inadequate resources
- Unresolved previous history (bad previous experiences)– trying to unlearn behaviours
- Lack of respect for the individual

The example was given from the teachings of Christian Aashamar, a Norwegian pedagogue and PWS expert. When a person with PWS asked Christian if he would ever live independently, he replied, “Yes, but you will always need help with four (4) things: food, money, grooming and work.” This statement is reassuring but it also accurately describes the nature of assistance required for maximum function with support.

## **Common Scenarios**

### **Scenario #1**

In a unit of 8 females, age 18 to 21, one 20-year-old young lady is approaching her 21st birthday and she will legally become an adult. Recently she has begun targeting an 18-year-old girl; she has been stealing her belongings. She has also become aggressive, slapping the younger girl in the face on two occasions. She has also been stealing indiscriminately from other persons, staff, and her family.

#### **Problems:**

- Slapping
- Stealing from peers
- Stealing from staff and family

#### **Solutions:**

- Talk to each girl separately and investigate reasons, get more details
- Older girl is stressed – challenging behaviour can be expected. Find out what's worrying her.
- Make the offender do something for the victim: restorative justice
- Stealing from staff – only have themselves to blame – items should be locked away
- Use previously agreed consequence for stealing and then have a fresh start
- Create programme to learn alternative ways to deal with frustration. "You are having a hard time – how can we help?"
- Work on issue of older girl using situation to gain power and young girl possibly envious of older girl.

## **Scenario #2**

In a unit of 7 ladies, a new lady has recently been added. Up until this point, the ladies were getting along quite well with one another thanks to a vibrant, sensory rich, activity program. However, since the new arrival there has been an increase in squabbling and in fighting among the ladies. There has also been an increase in picking behavior among the ladies, at levels not seen in months. The new arrival's behavior has been characterized by episodes of sudden dyscontrol leading to tantrums and attempts to run away by climbing out her window. Her behavior otherwise is typically submissive and quiet.

### **Problems:**

- Disturbing the group
- Running away and climbing out of the window
- One lady is "new"

### **Solutions:**

- Going on a day trip as a group and getting away from problems
- Doing activities together
- Do group introductions and welcome new member
- Divide into smaller groups
- The ideal number for a group was thought to be 3 – it is easier to find solutions, and can be used for therapy groups and activity groups
- Check lady's history – is this new behaviour? If so, address stress. If not, investigate triggers.
- Check for medical basis (depression etc) or stress of moving
- Are the 7 others instigating the behaviour?
- Think about the problem – don't jump too quickly to concrete solutions. Are behaviours inherent or precipitated?
- She may be trying to get thrown out in order to go back to family home
- Talk to psychologist/therapist

## **Scenario #3**

In a group home for 4 young men, one is talking on the phone with his mother about weekend plans. Another one has been struggling with his weight, gaining 5 to 8 kilos at home over a two- day stay at home. He is in one of his aggressive moods. He "hangs up" the phone call on his housemate, exciting him. This provocative behavior continues for hours.

### **Problems:**

- Psychological problem about gaining weight – compensates by using aggressive behaviour/acting out
- Group disharmony

**Solutions:**

- Walk away from the conflict and calm down with staff member
- Talk about the weight problem and call family for further details
- Letter to parents about liability of severe health consequences of stomach dilation after excessive eating

**Scenario #4**

Three youths have stopped at a fast food restaurant after shopping for clothes. One youth mixes a little sweetened soft drink (Dr. Pepper) into her diet soda at the self-serve counter. Staff notices this and plans to address it with her in a less public forum. Another youth notices it and confronts the girl in the store leading to a public row. “Why don’t you mind your own business!”

**Problems:**

- Helping herself from the self serve
- Conflict between peers
- In a public place

**Solutions:**

- Risk assessment. Staff need to be clear in advance about circumstances in which she can self-serve (if ever.)
- Closer supervision
- Predetermined consequence of stealing food and drink
- Staff to address this later.
- Praise the person who noticed for keeping to the rules and telling staff
- Ensure consequence for transgressor is enforced.
- In advance, tell group they will all go home if they do not conduct themselves appropriately. (And reward when they get home if they comply)
- If there is resistance to leave the store, staff member starts a discussion with one member of the group and backs towards the door – others will follow
- Get back-up staff from the group home

**Scenario #5**

All of the ladies at one group home have pets. Three of them have cats, and one has a guinea pig. When they first got their pets, they wanted to keep their pets, their pet’s food and other necessities in their own rooms. They argued constantly over whose cat went in whose litter box, and whose cat ate who’s food.

**Problems:**

- No initial guidelines
- Using cats as symbols of anger – vehicle for anger/conflict expression
- See cat as a possession

**Solutions:**

- Contracts and develop guidelines
- If contract broken, cat must go
- Let parents know what is happening
- Schedule what will happen to the group, not as individuals.

QED

**Second International Caregivers Conference  
Herne, Germany  
July 6-8, 2009**

**Workgroup: Behavior Management in PWS  
- Guidelines and Standards -**

**A Synthesis of Workgroups A4 and B4 from the Herne I  
Conference\***

## **I. Introduction**

This paper is the result of three workgroups which were made during the caregivers' conferences in 2008 and 2009 in Herne, Germany. The first workgroup in 2008 was called "**Crisis<sup>†</sup> Management**" and were one of ten workgroups held during this conference. The second and the third workgroup which were held during the conference in 2009 were called "**Behavior Management.**" These two workgroups were conducted separately during this conference with different participants and were one of four workgroups held. The results of these two workgroups were combined at the end of the 2009 conference and presented during a plenary session.

The main goal of the workgroup in 2008 was to discuss the themes of crisis management and behavior management and to clarify what we mean with these words in the field of PWS. The participants had the chance to find the same "language" for this field.

- Definition of a crisis
- Nature of the crisis
- Which types of crises exist
- How long does a crisis typically last
- The impact of a crisis

The main goal of the workgroups in 2009 was to discuss specific crisis and behaviour management methods when daily working with people with PWS. The basis of the 2009 workgroups was the results from the 2008 workgroups. The main 2009 themes were:

- The feeling of the caregiver in a crisis situation
- How to prevent a crisis/ problematic behavior
- How to handle a crisis/ problematic behavior
- How to reflect a crisis/ problematic behavior

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\* The format for this write-up is different to accommodate the material presented.

<sup>†</sup> When we talk about crisis in this paper, we mean behavior crisis.

## II. Workgroup Participants:

Over all, there were **57 participants** in the three workgroups (30 in 2008; 12 and 15 in 2009). They came from **15 different countries**: (7 in 2008; 8 and 10 in 2009): Australia – Austria – Bulgaria – Denmark – Finland – Germany – India – Israel - New Zealand – Romania – Spain – Sweden – Switzerland – UK – USA. The participants came from **various professional backgrounds**: caregivers in group homes (this was the main group of the participants) - psychologists – pediatricians – nursing assistants – special educators – managers of sheltered workshops – group home managers – parents of people with PWS – genetic researchers – psychiatrists – special education counsellors – teachers. The participant's range of **work experience with people with PWS** differed from a few weeks to more than 15 years. Most had experience of more than 5 years.

### **The workgroup leaders and presenters in 2008 were:**

Janice Forster (USA)  
Mary K. Ziccardi (USA)  
Gregory Cherpes (USA)  
Evan Ferrar (USA)  
Norbert Hödebeck-Stuntebeck (Germany)

### **The workgroup leaders and presenters in 2009 were:**

Irene Achutegui (Italy)  
Larry Genstil (Israel)  
Robert Pleticha (USA)  
Norbert Hödebeck-Stuntebeck (Germany)

## III. Herne I, 2008

**A. Stimulus Lectures:** There were three presentations given as an introduction of the theme in the first workgroup in 2008:

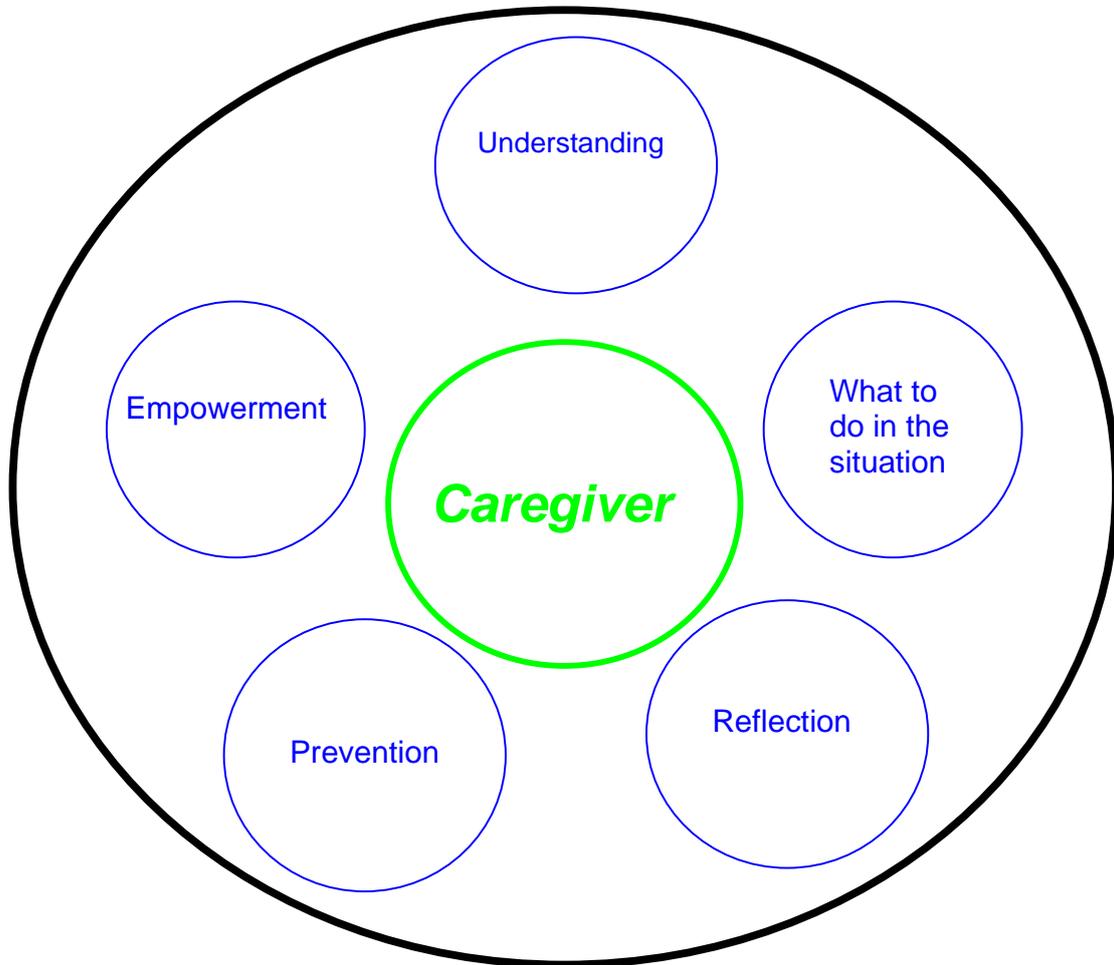
### **1. Presenter: Norbert Hödebeck-Stuntebeck**

**Title:** Crisis management in support of people with Prader-Willi syndrome: Understanding, acting, reflecting and preventing a crisis

Every person who is involved with the care and management of a person with Prader-Willi syndrome will eventually experience a crisis situation. And each crisis experience offers an opportunity to learn how to better manage a crisis in the future. Crisis management is best understood as a circular rather than linear process as demonstrated by the diagram below. Note that the caregiver is at the **center** of what is called ***the circle of support***.

## THE CIRCLE OF SUPPORT

### People with Disabilities (PWS)



### Behavior problems

This model suggests that successful management and prevention of crisis situations depends on the completion of several steps.

**First, caregivers must understand the crisis.**

Some strategies for understanding include:

- Asking searching questions to gather information about what the person in crisis is thinking and experiencing.
- Observing the person's physical reactions, expressions, and gestures.

- Making eye contact with the person so that communication remains open throughout.
- Examining any environmental factors that contributed to the crisis.
- Noting who is present and involved in the crisis.
- Establishing what happened before the crisis erupted.
- Determining if this is a new behavior.
- Exploring what the client hoped to gain from this behavior.
- Recording any consequences (negative or positive) that were applied and to what effect.

Caregivers can then explore what meaning can be derived from the information gathered, while completing additional steps in the process.

**Second, caregivers must know what to do when a crisis occurs.**

It is important that caregivers understand their own goal in a crisis situation, and how they will act as a leader in the situation to restore control. This foreknowledge will allow caregivers to present a clear and stable personal plan for the person with PWS to follow out of the crisis. Specific strategies can be utilized such as:

- Allowing space to the person in crisis
- Use clear and calm speech
- Delay of consequences
- Avoid power struggles
- Help client to get “unstuck” from behavior or thought contributing to crisis
- Know limits

**Third, after the crisis is resolved caregivers should take time to reflect with the client involved.**

Some issues for client reflection include:

- What did the client do well during the crisis or in resolving it?
- Encourage the client, as able, to explore the perspectives of others involved in the crisis.
- Develop, if possible together, a new behavioral plan to encourage improved behavior.
- Build bridges with the client through the reflection process. Even if the client is resistant to reflecting allow their participation.

This step should also include reflection of the caregivers to identify what they learned and experienced. A significant portion of time should be allowed for this step and no restrictions should be placed on what is said or shared. The goal is to see the crisis honestly, accurately and with some distance and perspective.

**Fourth, caregivers should use information and insight gathered in the three previous steps to create prevention strategies.**

Prevention strategies can include making changes to:

- The daily schedule

- Opportunities for people with PWS to expend energy throughout the day
- The number of available caregivers
- Qualification of caregivers (i.e. would additional crisis response training help?)
- Crisis response protocol
- Level of support the institution provides for crisis intervention
- Utilization of external support (clinics, psychologists, etc.) to help in crisis situations or prevention planning.

**Fifth, caregivers must feel empowered.**

This model is demanding of caregivers and requires strength to complete. However, if implemented properly and continuously, these steps allow caregivers to draw strength from their own experiences and the support of those around them. Only empowered caregivers can create a process of crisis management and prevention that is reliable.

**2. Presenter: Mary K. Ziccardi**

**Title:** Management of crises in community settings: Educational and legal

The ongoing support required to manage an individual with Prader-Willi syndrome must occur in every environment and in all situations. A caretaker's failure to anticipate and respond to potentially difficult situations will either immediately, or eventually, result in a crisis requiring more significant intervention, along with the potential for an undesirable or even irreversible outcome.

For children and young adults, the challenges present in an educational setting may be significant, and certainly problematic, if ignored or incorrectly addressed. Delayed intellectual functioning, coupled with behavior patterns that draw negative social attention, can lead to a difficult, and often socially isolating educational experience. These factors, coupled with the anxiety of a food-laden environment, require both the competency and commitment of all school personnel to assure a successful educational experience.

Intervention by law enforcement presents special concerns. When summoned, law enforcement personnel are typically trained to "respond"; many times, a wait and see approach, along with verbal reassurance, will result in a favorable outcome and avoidance of an escalated crisis situation. Educating law enforcement prior to needing their assistance is recommended. Further, law enforcement personnel must understand that, contrary to their training, the implementation of a consequence may have absolutely no effect on whether the same "bad act" will be repeated. Lastly, use of physical intervention must be a last resort, and only if imminent danger is perceived or present. Law enforcement personnel would benefit from advanced information about the risks associated with PWS and restraint, especially positional asphyxia and increased risk of fractures. When necessary, a cooperative partnership between the caretaker and local law enforcement authorities will increase understanding and reduce risks.

**3. Presenter:** Gregory L. Cherpes MD

**Title:** Prader-Willi syndrome and crisis management: Inpatient treatment setting

Individuals with Prader-Willi syndrome are at risk to experience physical and behavioral health difficulties that in the absence of adequate environmental management may reach critical dimensions. Certainly not all difficult behaviors are considered crises. A crisis could be defined as a situation where the supports available to an individual are less than the individual's needs at that time. Crises may be short lived or chronic in duration. They may be brought about by factors unique to the individual, factors associated with the PWS phenotype, or factors associated with the environment's response (or lack of) to the individual. When the individual's current environment cannot adequately: promote necessary steps toward health, sustain physical or emotional well-being, or prevent the recurrence of issues leading physical or behavioral health emergencies, consideration can be given to treatment in an inpatient setting. (Note: in the United States, "inpatient treatment" refers to care provided in a hospital where the patient stays both day and night until he is discharged to home).

The Prader-Willi Syndrome/Behavioral Disorders Unit of The Children's Institute is one such program that offers a model of inpatient treatment for the management of crises related to physical or behavioral health, including crises related to obesity, life-threatening conditions associated with morbid obesity, intense food seeking behavior, skin picking which reaches the level of self-mutilation, person-directed aggression, destructive behaviors, and identifiable psychiatric disorders. Whether "medical" or "behavioral," the presenting difficulties that warrant the use of an inpatient management setting typically share the qualities of imminent threat to safety (self or others) and/or the presence of severe impairment in the quality of life. Alternatively, a specialized treatment unit may be considered when other treatment settings cannot meet the needs of the individual with PWS.

Over the course of the past 25 years, a specialized milieu has been developed to meet the needs of individual with PWS in crisis. Key components of the milieu include: 1) A structured, predictable environment, 2) Absolute food security, 3) A restricted calorie diet based on the individual's current need, 4) A daily exercise program supervised by a physical therapist, 5) An individualized behavioral management plan, 6) Accommodations to meet the needs of individuals with severe morbid obesity.

While alleviating the immediate crisis is of great importance, an effective treatment program must also address the factors present or absent in the individual's home environment which may have directly or indirectly caused or failed to prevent the crisis. Family or other caregiver involvement in education and training throughout the course of the individual's inpatient treatment episode is a necessary, capacity-building component of effective treatment. Likewise, capacity-building in the form of

education and support among the individual's health care providers is essential to increasing the individual's likelihood of a successful return to their home setting.

## **B. Definition of Crisis**

### **1. Definition:**

The group worked out the following definition:

*A crisis is a negative situation that is out of control with risk to health and safety.*

### **2. Categories**

There were collected many examples for crisis situations by the participants and they were ordered in categories of crisis of high, intermediate and low severity:

**IMMEDIATE RISK** (High severity): A crisis that must be dealt with in order to prevent immediate danger to self or others.

- Self-injury
- Aggression
- Medical illness (acute onset or exacerbation requiring medical attention of admission to treatment facility)
- Psychiatric illness (psychotic or acute episode of new illness)
- Running away
- Property destruction involving damage that can cause injury to self or others, (i.e. breaking a window)
- Tantrum resulting in aggression towards others or self

**POTENTIAL RISK** (Intermediate severity): A situation that places the caretaker "on alert" because it could escalate to a crisis.

- Tantrum
- Threats of aggression/property destruction/running away
- Food acquisition
- Psychiatric illness
- Property destruction
- INVOLVEMENT OF LAW ENFORCEMENT – either by the person calling emergency numbers or others calling law enforcement due to the severity of the incident –added by the committee

**MANAGEABLE RISK** (Low severity): Situations where a crisis may be emerging, but it is not as yet at the point to require full crisis intervention. In these situations, implementation of lower level preventative measures might stop a crisis from developing.

- Shut down from programming
- Tantrum
- Food acquisition
- Breaking the rules
- Psychiatric illness
- Splitting system of care

- Impaired judgment reflecting inappropriate behavior
- Bereavement
- Property destruction
- Uncertainty or changes in programming

### **3. Why is crisis management important for PWS?**

- Proper crisis management provides security for a person with PWS.
  - Security is knowing they will be taken care of no matter what. This builds a relationship of trust.
- Crisis management provides a “skin/buffer” to protect a person with PWS from the stresses and pressures of the community.
  - They are not doing it on purpose; they cannot express themselves appropriately (i.e., anger, sadness).
- A crisis is a learning opportunity for prevention.
  - It provides an educational opportunity to clarify the underlying issues.
  - It gives perspective; a reminder that the little things can become big things very quickly.
  - It provides the tools to be prepared.
  - It reaffirms that it is important to be one step ahead.
  - It provides an opportunity to prevent frustration before escalation.
- Crises are important in order to get to know each other (a process of relationship development).
- Crises cause stress and interfere with personal development and competence.
- Crises are exhausting, demoralizing and interfere with ongoing relationships.
- Crisis management keeps everyone safe.
- Crises are universal; everyone who works with persons who have PWS has experienced a crisis.

### **4. What makes a crisis more likely to occur when working with persons who have PWS?**

Persons with PWS are highly dependent upon environmental parameters for their adaptive functioning, regardless of their level of intellectual ability. Further they require a high degree of consistency and predictability in their daily plan.

FOOD SECURITY is the hallmark of the environmental infrastructure. This provides *no doubt* about their diet plan; *no hope* of deviating from this plan (and because of secured food access, there is no chance of getting any more than predicted); and

having fulfilled both of these requirements, there is *no disappointment*, and hence, no emotional distress and no acting out behavior. This *no doubt, no hope, no disappointment* paradigm also applies to the daily schedule. The person knows the schedule of activities and tasks throughout the day. *Preferred* activities follow less preferred ones to assure smooth transitions. All activities are time-limited (even leisure activities) so individuals are less likely to *get stuck*. By providing structure and consistency in the environment, the stress sensitivity that is characteristic of the syndrome is managed, and behavior problems are diminished.

There are several personality traits (the behavioral phenotype) typical of persons with PWS that inevitably place them into conflict with the care-giving individuals who are responsible for securing their environment. Persons with PWS display cognitive rigidity, inflexibility and perseveration that interfere with changes or transitions. As a result of their single mindedness, they have difficulty taking another person's point of view; their capacity to understand the impact or the consequence of their behavior on others is diminished. They can be noncompliant, argumentative and oppositional. Their judgment is impaired regardless of their intellectual ability. They have selective interests and excessive/repetitive behaviors that often require limit setting. Their emotional response can be intense and reactive or unexpectedly minimal. They can display a high degree of persistence and refuse to end an activity, or become easily frustrated and withdraw from an activity. Some of the most challenging behaviors include: aggression, self-injury, property damage, running away, shutdowns, temper tantrums, withdrawals or refusals.

These behaviors elicit a response from the caregiver. The nature of the caregiver's response can determine the outcome of these behaviors. Appropriate management maintains harmony with the environment; mismanagement results in greater conflict between the person with PWS and their environment. This conflict increases the person's stress, and increases their acting out behavior. Thus, a crisis can be precipitated when a system/provider/care-giver is not aware of the unique needs and personality traits of the person with PWS or when typical PWS behaviors are mismanaged.

### **C. When/where do crisis situations happen?**

Consensus existed among the workgroup that a *risk for crisis* was high among individuals with Prader-Willi syndrome. This consensus may of course reflect a bias among the panel presenters and group members who elected to participate in this topic.

A change of any kind in the life of a person with PWS can cause a crisis. Some examples include:

- Change of placement
- Illness
- Bereavement
- Life changes

- Uncertainty – especially with food

In addressing the question “When/Where do crisis situations happen?” an extensive list was generated and appears immediately below in the order presented by the group to the facilitator:

1. Parent visits (before or after)	16. Too much or insufficient information
2. Special events	17. Food situations
3. Communication issues	18. Social context (including intimate friendships)
4. Changes within care team, rules, or staff	19. Being rushed or hurried (time management)
5. Before breakfast	20. ‘Planned’ crisis
6. When there is too much free time	21. Unknown situations
7. Meals with other residents	22. Fear of evaluation
8. When children come home from school	23. Unmet expectations
9. Workplace	24. Arguments in social context
10. Seasonal	25. Crisis never occurs when sleeping
11. Full moon	26. Low structured weekends-more free time
12. Menstrual cycles (of staff!)	27. After lunch
13. Task specific	28. Time before going home
14. Function of expectation	29. For some, when left alone
15. Anticipation/expectation (planning)	

Ultimately, the above lists reflect the fact that a crisis may arise at any point in a person’s day or life when the available resources, both intrinsic and extrinsic, are inadequate to maintain an individual’s health and safety. Vulnerability to the wide variety of external stimuli noted above underscores the importance of establishing and maintaining predictable routines, including all aspects of meals. Items listed above that reflect an internally mediated locus of control serve to underscore the importance of developing an appreciation for the individual. While certain characteristics are attributed to persons with PWS, an in-depth knowledge of the particular individual being served is of equal or greater importance in identifying specific triggers to potential crisis-inducing situations. Personality style, coping skills, temperament, self-esteem, and past experiences vary from person to person and all contribute toward a person’s tendency to experience a crisis. The presence of identifiable affective, anxious, or cognitive disturbance will serve to further influence an individual’s response to the above situations.

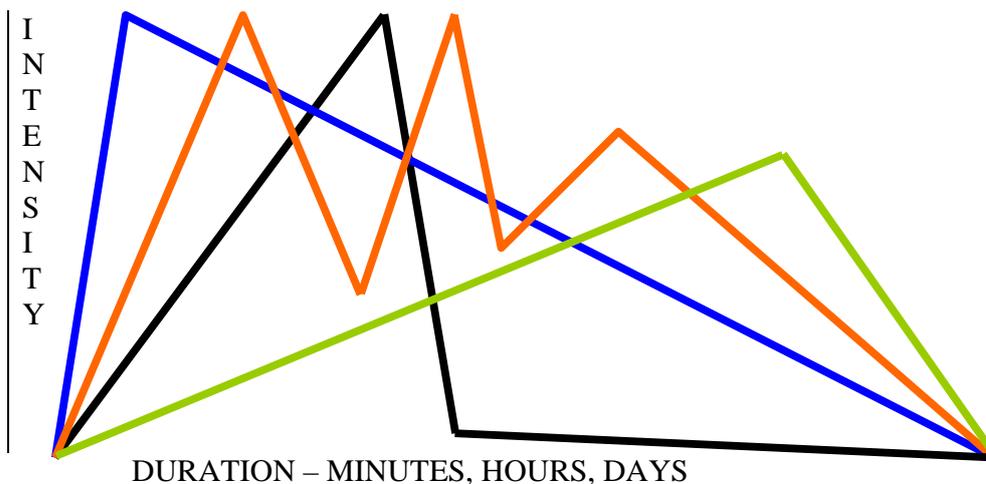
The members of the workgroup recognized the potential of caregivers to contribute to the emergence of crisis situations. A group member mentioned “menstrual cycles (of staff)” somewhat humorously. The spirit in which this comment was made was to emphasize the role of the caregiver’s outlook, mood, and their own well-being. Poor communication among caregivers was also identified as an important contributor to potential crises.

While the broad list of potential triggers for a crisis may seem daunting, clearly individuals with PWS do not spend most of their lives in crisis. The list generated

does reflect the powerful need for vigilance and mindfulness on the part of caregivers to help alleviate the risk for crisis. There is always reason to be hopeful that attention to, and anticipation of, internally and externally mediated triggers, which will certainly vary among individuals with PWS, will help to enhance the individual's well being.

## D. How long does a crisis last?

Anatomy of a crisis:



There is a high degree of variability in the intensity and duration of any given crisis.

- Some present unexpectedly,
- Others build slowly in evolution.
- Some appear to be resolving when they suddenly peak again.
- Some resolve quickly.

Each pattern is unique to the person and to the context of the situation.

Due to time constraints, the workgroup was not able to fully explore this topic. The two comments generated were: tantrums – 1 hour, or until the next meal. Certainly, a great variability exists in terms of the duration of a crisis. Duration is influenced by:

- the cause of the crisis,
- the presence of potential perpetuating factors,
- the resources of the caregivers to respond to the crisis.

While “until the next meal” may reflect the experience of the workgroup member who offered the comment, the leadership team would like to emphasize the importance of caregivers not attempting to use food as a means of resolving a crisis situation!

## **E. Impact of the Problem**

### **1. Person with PWS**

A crisis impacts the life of a person with PWS in significant ways. To begin with, a crisis calls into question whether or not their world is secure and their caregivers are trustworthy. If those questions are not answered affirmatively through the effective resolution of a crisis, then the increased anxiety of the person with PWS will likely lead to even more crisis situations. Conversely, if the crisis situation is handled appropriately and effectively, then proper crisis management can strengthen the bond between the person with PWS and their caregivers, leading to a more crisis-free environment in the future.

### **2. Family**

Family members can also be dramatically affected by crisis because a crisis can trigger concerns that their loved one is in a living environment that does not safe guard their basic human need for safety and security. These family concerns can result in increased friction with caregivers and possibly lead to removal of their loved one from the program. Also, friction between family and caregivers can slowly erode the partnership between the caregiver and the person with PWS that is so essential to maintaining a healthy living environment.

### **3. Caregivers**

The impact of a crisis can have a profound affect on caregivers. Exhaustion, increased excitement, physical tension and a feeling of helplessness can accompany a caregiver's crisis response. Effective crisis management requires a high level of professionalism in the face of escalating stress; it demands clarity of focus and a neutral emotional approach. Caregivers rely on their supervisors to empower their response through the availability of necessary resources, such as adequate staffing patterns, training, and appropriate policies and procedures to support them before, during and after a crisis situation.

Crisis situations are particularly difficult because, by definition, they are emotional events – both for the person in crisis and the caregivers who are responding. When asked how crises made them feel, work group participants shared 10 responses; only one of these was a positive response: focused and calm. The rest of the responses captured a range of negative reactions including feeling helpless, tense, a lack of emotional control, a sense of failure, indecisive, overwhelmed, and a need to lower expectations. Work group participants also indicated that in some cases their flight/fight impulse was activated during a crisis.

Interestingly, frustration was identified as an issue for both caregivers and the person in crisis. For the caregiver, frustration builds because of the ability of people with PWS in crisis to push the caregiver's emotional buttons to cause hurt feelings. These hurt feelings potentially lead to anger for the person with PWS which can extend long after the crisis if not resolved in a healthy and constructive way. For the person with PWS, frustration can grow with the caregiver's response to the crisis. In

time, if that frustration is unresolved, it can turn into a long lasting anger towards the caregiver. This simmering anger can lead to behavioral problems – and possibly another crisis – in the future.

#### **4. Providers**

Crisis management challenges providers on both a person-centered and an agency-centered level. On the person-centered level poor crisis management can result in staff burnout; physical and emotional injury to both staff and people with PWS; serious disruption in daily activities and services; and a loss of the sense of community within the living environment. On the agency-centered level a provider can experience potentially expensive property damage, litigation, and staff turnover by not managing crisis effectively. Significant problems with crisis management can attract the attention of monitoring agencies resulting in additional oversight - which can be time consuming and demanding - and in some cases it can result in a decrease or elimination of funding. Frequent crisis situations also diminish a provider's reputation in the provider community and among potential consumers.

#### **5. Supervisors and program administrators**

Participants acknowledged there could be a difference between the perspective and understanding of Prader-Willi syndrome among frontline staff, their supervisors, and program administrators. This difference can be harmful to the effective functioning of an agency, as conflicting opinions and views can lower staff morale, decrease the value of programming, and prohibit the kind of consistency and structure necessary for effective PWS care giving. Therefore, it is very important for an agency to have a shared organization-wide commitment to working with people with PWS, based on available best practices. This might mean, for example, adjusting caseloads for people working with PWS clients who may require more time. In addition, most successful organizations have people in management with direct service experience working with people with PWS so they have an appreciation for and understanding of the challenges staff face. This understanding will contribute to better communication between staff and management and reduce frequency of conflict and burnout.

#### **6. System of care in society**

Crisis response has an impact on the system of care in society. Escalating crisis cases can exhaust local supports when community resources (i.e. medical, educational, and legal) cannot handle the level of care needed to respond effectively and appropriately to the person in crisis. When this happens, a person with PWS will often be placed in a situation where the primary responders (for example, the police) are not trained to react in a constructive manner. This can lead to a worsening of the crisis situation and an even longer disruption of the daily therapeutic programming so vital to the stability and health of a person with PWS. As a result of these factors, crisis interventions beyond the immediate caregiver level are costly and frequently ineffective. Thus, it is a priority to give primary caregivers the crisis management tools, training, and resources needed to respond immediately

and effectively to a crisis situation so that it does not escalate to involve a local system of care that is often ill equipped to manage a PWS specific crisis situation.

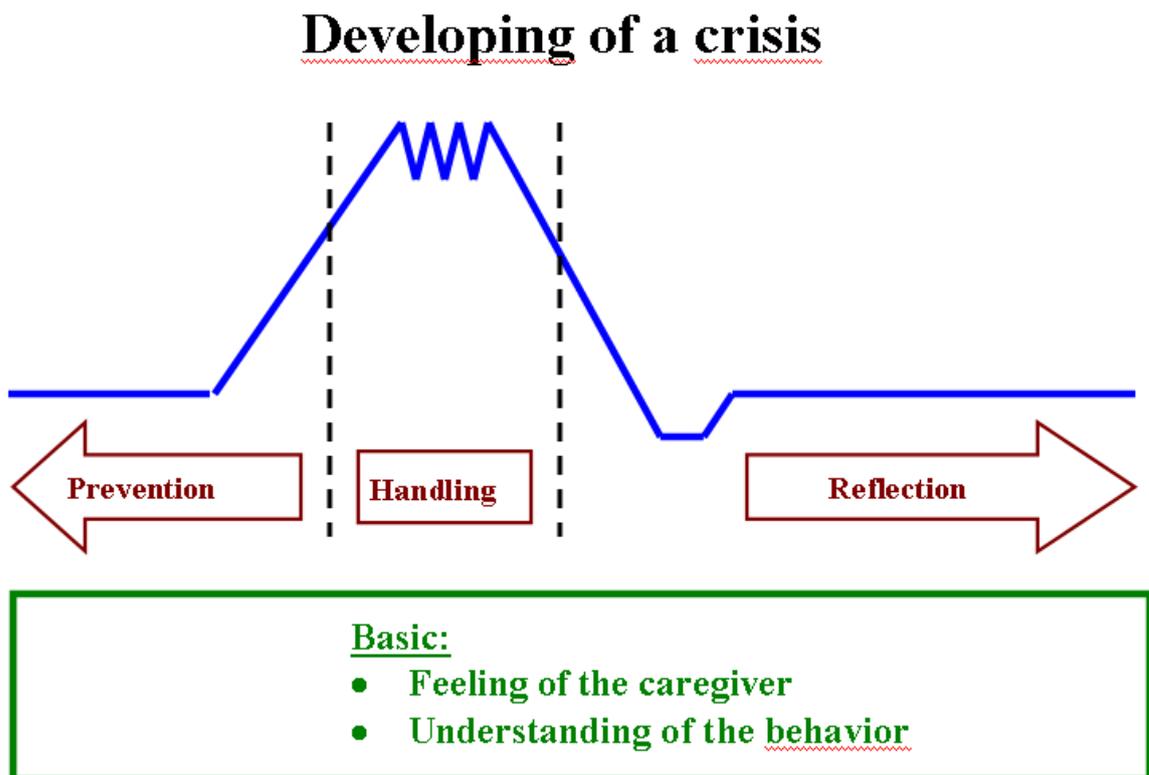
## IV. Herne II, 2009

### A. Standards and Guidelines for prevention, handling, reflecting in PWS and the role of the caregiver

In the 2009 workshops, the following four themes were focused on and discussed on the basis of the results of 2008:

1. The role and the feeling of caregivers
2. Prevention
3. Handling
4. Reflection

In the following picture you will find all the parts integrated:



## 1. Role and feelings of the caregivers

During the 2009 workshop we choose to begin with a description of a crisis situation, with immediate risk, in order to start an in-depth discussion involving issues beyond that of typical behavior of individuals with Prader-Willi syndrome (PWS). The caregivers' feelings, attitudes and points of view influence their intervention and their relationships with the individual with PWS, families, colleagues, institutions, etc.

The feelings of the Caregiver are an additional component that is extremely important and relevant in a good behavioral plan in order for the staff to be able to feel effective and consistent. It is also important for the Caregiver to understand that there are emotional and attitude factors affecting his job performance that may be difficult for him to come to know. For example, if staff feels negatively about an intervention, the probability of the intervention being implemented in a consistent manner is low. The hypothesis is that everybody, especially in stressful situations, behaves according to inner motivations of which each one is not often aware.

Understanding and reflecting sometime after the crisis, helps caregivers to know more about themselves (their own motivation and feelings, behavior/response, expectations, goals), to know more about the individual with PWS they work with beyond mere syndrome characteristics (also his/her emotional state, sentiment and feelings) and to know more about the relationship, and the communication between the caregiver and the client.

Individuals in the work groups offered emotional support to each other by sharing similar problems, worries and experiences. In large part due to this support, caregivers are more able to accept new perspectives and criticisms. They can recognize, for instance, that they had misinterpreted their own attitudes or the feelings of the individuals with PWS in a crucial moment of their relationship. The group discussions gave caregivers more insight into themselves and into their relationship with an individual with PWS with beneficial consequences to their relationship.

The 2009 workshop gave caregivers the opportunity to reflect on: how they feel and act in stress situations, how/where they look for support, how they can re-establish control, what to do after, what to do for prevention, how they can handle similar situations in the future. Even with so many different countries and cultures represented in the workgroups, **feelings and needs of caregivers are universal**. Each participant agreed that we all have all the same feelings and we all need the opportunity to express these feelings and get support.

Behavioral crisis situations can take many forms for people with PWS. It might be an increase in tantrums. It might be a new habit of hoarding or perseverating. It might be an increase in irritability or food seeking.

Just as crises can take many forms, so can the reactions and feelings of caregivers following a behavioral crisis.

### **A. Feelings of caregivers**

These emotions may include:

- feeling of powerlessness
- helplessness
- deep concern for the individual and self
- controlled by negative, fearful emotions that are present when individual is in crisis
- nervous
- scared
- angry
- frustrated
- anxious
- shocked
- disappointed in individual and self
- under pressure from the environment and people around witnessing the crisis
- guilty and responsible for not being able to manage situation
- as if the individual in crisis was in control of the situation and not the caregiver
- betrayed: feeling that a good relationship exists with the individual and then frustrated when attack or problems occur, loss of trust in that individual
- responsible for individual's well-being
- fearful of the reactions of the work supervisor, parents of individual to caregiver's management of situation
- sense of failure with self-esteem and self-efficacy diminished

Chances are that most caregivers have felt a mix of these emotions at some point in their lives in living and working with individuals with PWS. These emotions can cause a caregiver distress and may result in burn out if solutions are not found.

### **B. Support for caregivers**

Some suggestions for caregivers to deal with these feelings include:

- Consult with a psychologist after crisis
- Collaborate with a psychologist to develop a behavior plan to gain a sense of empowerment if crisis situation resurfaces
- Create an environment of mutual staff support through weekly staff meetings, team building activities
- Make sure to have family and social activities outside of work, try not to take feelings home
- Inter staff shifts so that one staff person does not have to work with resident with whom they have just had a crisis
- Families need to be informed and be actively included and involved with caregiving institution
- Open communication with family,
- Regular meeting with family, client and caregivers

## **2. Prevention**

### **A. Individualized Behavior Plans**

It is critical to focus efforts on preventing a behavioral crisis. Focusing on prevention means that behavioral crises may be entirely avoided.

An interdisciplinary team including the individual, medical doctor, psychologist, social worker, case manager, parent, direct care staff members (group home, work supervisor) should create an individualized behavior plan including what interventions are necessary to avoid escalation to a full blown crisis situation. The individual, and all people in contact with individual, should be trained on how to implement behavioral plan and have opportunities to offer feedback and suggestions for modification to the plan. Behavioral plans are most successful when they are implemented consistently in all environments the individual interacts with.

A critical component of this plan is keeping in mind person centered planning techniques. The behavior management plan should reflect the preferences, opinions and emotions of the individual. The individual should help create the plan, accept the final version of the plan, and perhaps sign the plan to demonstrate their agreement. The plan should be tailored to each individual's mental level, and have clear rules and limits for appropriate behaviors while offering structure in the form of limited choices. While focusing on prevention of behavioral crises, the appropriate consequences of maladaptive behaviors should be explained. It is important to take into account that not all individuals with PWS will be able to comprehend the consequences of their behaviors.

Many behavioral crises may be preceded by a situation that causes anxiety in the individual. Through behavioral observation and monitoring, the plan will identify situations and triggers that create anxiety and possible precursors to crisis situations. As a way of defusing these situations, the plan can offer the individual limited choices on where to go and what activities to partake in to reduce anxiety. It is important that very few options are offered because too many choices can be overwhelming and result in further escalation of maladaptive behaviors. For example, if the individual is beginning to show signs of increasing anxiety and agitation, the plan can include options to redirect to a different environment such as a bedroom or designated quiet area to go to in order to calm down. Additionally, a different activity options can be suggested to the individual such as listening to relaxation music or any other option the individual takes pleasure in like working on a puzzle or playing a computer game. Rewards or reinforcers for good behavior may also be built into the plan for individual's that need extra incentive to choose positive behaviors.

Since PWS is characterized by strong visual perception and possible other sensory deficits, an effective plan may include a visual prompt card so that the individual can understand expectations of appropriate behavior, while avoiding verbal instructions

that the individual may find overwhelming. For example, if a crisis seems to be building, a caretaker can give a pre-made card (preferably in a calming blue color) to the individual with the option to redirect them to a calming, quiet area like the individual's bedroom to take some time to relax. It is important that the same visual prompt is used every time with the individual as cognition at a time of high stress or agitation may be difficult.

### **B. Important considerations of prevention especially for: direct care staff, group home staff, work supervisors, teachers**

When working with individuals with PWS, it may help to have information about client's family's background in order to fully appreciate the efforts of parents in the past and to work in accordance with the individual's goals, family history, and educational experiences. It is also important to understand the history of the client's behaviour in the family context.

After gaining the necessary case history about an individual with PWS, it is also important to have an open, communicative relationship with the client. This relationship involves:

- respecting the individual's feelings,
- accepting their suggestions,
- and working to improve self-efficacy, and
- self-esteem through interactions.

This "healthy" relationship can act as a guide for client's to work towards constructive relationships with caregivers. With regular consultations with a psychologist, the caregiver can learn how to related more effectively with the client to express emotions in constructive way, define feelings, and share frustrations in order to prevent crises.

Transition times to and from home visits in a group home can be especially challenging for individuals with PWS. The individual needs to be given time to adapt back to the rules of the group home and order to create a positive and secure atmosphere. When the individual feels informed and prepared for changing expectations they can help to problem solve through potential problems.

Staff working with individuals with PWS, should be comfortable in their fellow colleague's abilities to be consistent, and share information and approaches amongst each other. For example, if an individual attends work at a sheltered workshop during the day and then lives at a group home; then group home to workshop communication about the individual's diet, behavior, and mood(s) is vital to all parts of the care giving system to work in harmony. It should be mentioned, that the two worlds "living" and "working" should not be too close, they must be separate worlds. Maintaining the divisions between the two environments is important so that if the individuals with PWS have a conflict, for example, in the group home, and than to go to work, that in the workshop nobody knows about this

conflict. This will allow the individual with PWS to have a "time out" for themselves. Then, when they come back in the evening in the group home they can work on this conflict.

It is the duty of staff members working with individuals with PWS to inform and prepare people in the community that the individual may encounter: What are the client needs and how to handle them in order to help the individual behave properly. This includes such community members as: spiritual advisors, hairdressers, bus or taxi drivers, cashiers, health care workers, etc. For example, if staff members know that the hairdresser usually has a dish of candy out for clients in the shop, it would help the individual's behavior if the hairdresser was called and asked to remove that access to candy for the duration of the time that the individual is receiving services.

### **C. Prevention Suggestions/Techniques:**

#### **C.1. Prevention Suggestions in a Pending Crisis Situation:**

- Assist, facilitate and encourage communication between two clients when they are in disagreement or upset at each other so that situation does not get out of control. Act as a mediator in resolving the conflict.
- Be aware that people with PWS can "share" crises. One individual's emotional outburst may trigger other's outbursts for no apparent reason.
- Reduce tension and stop escalation of situation by using humor, tickling, jokes to distract and redirect from maladaptive behaviors.
- Assign authoritative responsibility to person in escalating crisis so that they feel in control. For example, say "Go help John (staff member) with unpacking the groceries."
- If individual is prone to behavioral crises, have a card with a visual prompt which redirects the individual to a time out or cool down area.
- Offer an early time out opportunity to cool off or relax. Whenever possible, offer the individual to exit the pending crisis gracefully and with dignity intact.

#### **C.2. General Prevention Suggestions:**

- Respect what is important to client and aspects of client's lives that they have control over. Diet, food and drinks are some very important aspects of the lives of some individuals with PWS. If they are given choices of what to eat, make sure the opportunity for making those choices remains possible.
- Caregivers must be alert and anticipate personal and environmental factors that may result in crises. When the client feels good inside they may be able to tolerate frustrations or changes more easily, but in other moments those frustrations can result in a crisis. It is imperative to double check the details of an individual's schedule, diet, exercise plan, etc., when the client seems to be more sensitive to changes that are important to the environmental structure and expectations of the client. For example, if a client has just returned from a weekend at home and is feeling homesick, communicate this fact to other staff members that the individual comes into contact with and

- check to make sure that transportation details are worked out, if the client is sometimes anxious during transportation times.
- Relaxation techniques and sensory stimulation options offered and available. Examples of relaxation activities include: music, aromatherapy, breathing exercises, painting, meditation, etc. Examples of sensory stimulation activities include: exercise, stationary bike pedals, exercise resistance bands, stress balls, etc.
  - Control surprises in schedules through structure to minimize doubt and anxiety.
  - Ask individual to paint their feelings in order to become more in touch with their emotions. Include this activity in the individual's behavior plan.
  - Avoid leaving temptations in environment (unlocked refrigerator, open doors, etc.)
  - Use clear answers like yes or no, do not use the word "maybe." Maybe can be understood as yes to a person with PWS.
  - Make small steps towards behavioral change and practice challenging situations through role play with individuals with PWS.
  - Avoid challenging situations that could get out of control, especially when individual is in a bad mood. If you know a resident is upset and does not want to make their bed, perhaps wait for a later time to ask the resident to make their bed and do not insist that it is done now, if the resident seems very upset.
  - Know the individual behaviour history of problematic behaviour. When did it start (first time), how was the development (continual, not continual)

### **C.3. Prevention Suggestions Especially for Staff:**

- Guidelines in place so that each staff member knows limitations on decision making responsibilities. If a client has a question about their activities or schedule, make sure the proper staff member is answering the question. Include these guidelines in staff position descriptions so that inaccurate promises or statements are not made to individuals with PWS.
- Staff can not criticize another staff member in front of individual with PWS, it will undermine credibility, show vulnerability and cause a loss of basic structure. Staff must stay unified.
- Staff cannot go around the rules or shift responsibilities to other staff members in order to make their jobs easier. For example, if one staff member is feeling overwhelmed and gives into a resident's demands for food, the next staff member will have trouble enforcing the normal rules, once they have been broken.

## **3. Handling**

Handling a behavioral crisis can be defined as when the individual is in crisis mode (in a tantrum, physically aggressive toward self or others, throwing objects, uncontrollable crying, etc., see "Definition"). Crisis times can be emotionally exhausting and trying times for all parties involved, including the individual, peers,

and staff. Whenever possible and depending on each individual, hopefully proper prevention techniques and staff responses to possible issues, can help the individual to avoid entering into a full blown crisis situation.

If a crisis is building, but not to a point where the individual is a danger to themselves or others, ask and offer possibility for the individual to go to their own room or another safe place. Many times leaving the situation that is causing distress can create an opportunity to contain the crisis and give individual time to calm down. This should be a place where staff is not intruding on the individual, as long as they do not have a history of self harming behavior. Staff should then wait for the individual to be ready to re join the group or re enter the common areas.

People with PWS can “share” crises, or in other words, one person’s crisis can cause other people around them to become agitated or in a crisis without any good reason. Behavioral crises can be contagious; an individual with PWS may become upset when they see a peer or roommate upset. Therefore, caregivers must attempt to limit the crisis to one individual by removing the person in crisis and/or audience to safe place.

## **A. Handling Recommendations**

### **A.1. Considerations for Caregivers:**

- Care giving institution must share information on which clients have been aggressive in the past so caregiver is informed and feels comfortable managing an individual with aggressive tendencies.
- Caregivers should know the legal policy and agency stance regarding physical restraint or “holding”, any instance of restraint needs to be part of written behavioral plan developed and approved by a professional, all staff in contact with the individual should be trained in these techniques to ensure safety for everyone.
- Caregivers need to listen to the individual in a crisis and stay calm, in order to avoid escalation of crisis.
- Once in full blown crisis, the less talking, the better, individuals who are agitated and in crisis mode may not understand long and complicated sentences and it may only further confuse and agitate them.
- Caregiver uses startle response to try to shock client (if you know the client very well) to regain self control.
- Caregivers should always call for back up staff support early; don’t get trapped without back up. For example, if you know an individual has a history of violent behavior, do not get trapped in a room with that individual without an escape route.
- Do not hesitate to ask for help from outside sources: parents, other caregivers, on staff nurse or doctor. Caregivers always have the right to ask for external aid when they see no other peaceful resolution possible or are frightened for their safety or the safety of others.

- Make sure caregivers and individuals with PWS know rules in a crisis situation. For example, each resident living in a group home moves into their rooms when a fellow resident has a crisis.
- A caregiver must never forget that documenting the situation after the use of physical restraint is required by law in most areas.
- Caregivers should also consider documenting crisis situations for their own use in order to analyze and discuss the situation with everyone that encounters the individual who was in crisis.

#### **A.2. For individuals with PWS:**

- Make sure all dangerous objects are far away or removed from area. A kitchen might not be a good place for a person to calm down in due to the proximity of potentially dangerous items like knives, glass, gas stoves, etc.
- If possible ignore the behavior; the individual may want the staff attention that being in a crisis gets them.
- If it is part of a written behavior plan: promise a reinforcer or reward for regaining control. For example, "If you can calm down and stop crying, you can have twenty minutes of computer time to play your favorite game."
- If an individual that the person in crisis finds calming is available, involve that individual to help the person calm down.
- Threats are not a conducive means of reaching an end goal with any individuals with developmental disabilities. Threats like, "You won't get your dessert tonight if you throw that book!" especially do not work when an individual is in a behavioral crisis.

Some techniques would be useful depending on the situation and how well the caregiver knows each individual:

- Use the technique of overcorrecting the maladaptive behavior, have the person repeat bad behavior until it seems ridiculous. If individual slammed door, ask individual to slam the door twenty more times. (When you use this, you need to know the client very well.)
- Play calming music, display calming materials (blue), use calming scents, or other sensory items. Make sure to use that technique in every crisis situation to strengthen the individual's relaxation response to that stimulus.
- Have a picture behavior plan (blue background) to show the client so that they can see what is expected of them when they are in a crisis.

## **4. Reflection**

Following a behavioral crisis, it remains critical to discuss, reflect, and plan for the future, both with the individual who had the crisis, and amongst caregivers. The optimal time to discuss the crisis event depends on the individual, their state of agitation, and cognitive level. It is helpful when the individual is calm and ready for such a discussion, should a reflection conversation take place.

Such a conversation should take place between a caregiver or caregivers and the individual who had the crisis and be conducted in a calm, compassionate manner. The individual should not be blamed or shamed regarding their behaviors while in crisis. Rather, the discussion should focus on positive behaviors that the individual could have done to avoid the crisis and how those behaviors would have resulted in a different outcome.

For example, if an individual threw their plate at the wall and began crying when receiving the incorrect food at a mealtime, the caregiver could discuss positive ways of pointing out a problem, and perhaps help develop a picture communication card for the individual to use and show to staff when they disagree with what has been put on their plate.

Reinforcing this discussion would be the mention of the different outcomes that could have come if the individual had chosen a different behavior. For example, instead of being asked to go to a quiet area, and clean up the wall where they had made a mess, all resulting in eating at a later time, the individual could have pointed out the problem, whereby the staff would have fixed the issue.

## **A. Reflection Recommendations**

### **A.1. Considerations for Caregivers:**

- “All” crises should be reflected with the people with PWS, so that they have the chance to learn
- If a crisis is focused on one caregiver, or directly involves one caregiver, it may be necessary to distance that staff member and client until the appropriate reflection period. Additionally, it may help to have a neutral staff person, who was not involved in the crisis, lead the reflective talk.
- If the individual does not want to speak about the crisis, allow them and wait for a later time. Forcing the individual into a discussion they are not ready to have can cause another tantrum.
- Make a behavioral contract with the individual regarding the antecedent of the tantrum. Encourage the individual to suggest a possible staff response to the antecedent. Role play with a video camera, so that client can watch at later time to reinforce a more appropriate response.
- Caregivers must be careful to choose an appropriate environment and time to hold the reflection discussion. It must be mentioned that the reflection discussion is personal between caregivers and the individual and should not take place in front of the group.
- Following a crisis, group music therapy can allow individuals to express emotions, calm down, remember the strength of the group and importance of relationships within the group, help people to apologize, and express emotions through music when they cannot find words. Additionally, group music sessions can offer clients an opportunity to internalize rules and offer each other emotional support. Caregivers should always be willing to try new ideas and involve new experts such as music therapists when possible.

- It is important not to minimize the effect of crises both on individuals and caregivers. One should seek appropriate external, professional help with a serious crisis to heal wounds.

#### **A.2. During a Reflection Conversation:**

- It may be useful to use redirection from serious situation to reflection conversation. For example, a caregiver might say to someone who has just had a behavioral crisis, "Let us go in the kitchen and share a glass of cold water."
- Individuals with PWS have strong visual skills. Therefore, when holding a reflection discussion with an individual, caregivers must establish eye contact, and use visual cueing, whenever possible.
- During a reflection discussion, focus on how great it is when person is calm and in control.
- Rather than giving commands to the individual regarding the next time, ask the client what they would want to happen. Include these thoughts in a behavior plan aimed at eliminating crisis situations, and maladaptive behaviors.
- Work on creating new cognitive behavior patterns regarding potential crisis situations through cognitive behavioral therapy to choose different cognitive response, then practice new thought, write it, draw it, role play it again and again. Thereby, the individual is helped to develop new behavior set.
- Reflection conversations do not just have to take place while sitting at a table or across a desk. Consider taking a walk in nature while discussing the crisis.

## **V. Guidelines for Best Practice**

(The following list represents only the results of the workgroup in 2008)

1. The top priority in crisis management is to protect the person, staff and others from harm.
2. There must be acceptance that crises will arise when working with a person with Prader-Willi syndrome, and the overall goal is to reduce the frequency and intensity of the incidents.
3. In response to a crisis, effective staff will be able to stay calm, think before acting, know what to do and display a capacity for self-reflection.
4. Staff orientation and training should provide ongoing PWS-specific information including crisis management.
5. Effective staff should be able to offer security and "unconditional love" despite the person's crisis.
6. Effective staff will utilize a team concept with communication about advanced planning, structure, consistency.
7. Following a crisis a debriefing process should occur to find reasons for the crisis without blaming anyone.
8. Advanced planning is a strategy for prevention with two components:
  - Person-centered planning includes understanding the history of behaviors, what motivates them, and how to communicate best with them.

- Caregiver-centered planning includes adequate training and appropriate staffing patterns.
9. Administrators must appreciate the seriousness and the challenges of working with crises occurring in persons with PWS. Care-givers would feel more supported from their administrators and others by:
- Providing opportunities for relationship building with the person through 1:1 staffing.
  - Well-structured teams.
  - Advanced training on PWS and crisis management.
  - Not working alone (no one-caregiver shifts).
  - Time for staff meetings and communication.
  - Professional mentors and support networks.
  - An informed organizational structure regarding PWS.
  - Online support group (international).

QED

**Second International PWS Caregivers Conference  
Herne, Germany  
July 6-8, 2009**

**Workgroup: Environmental Structure for PWS  
Living - Best Practice Guidelines**

**Chair: Hubert Soyer (Germany)**

**Gayle Benet (UK)  
Jackie Mallow – (USA)  
Renate Scharfenberg – (Germany)  
Mary K. Ziccardi – (USA)**

Note: The reader is directed to the proceedings of the Herne I workgroup on **Environmental Structure of Living** for a review of presentations, group discussion, and standards of care. The Best Practice Guidelines derived during the Herne I conference were reviewed and refined during Herne II. The consensus of the two workgroup discussions from Herne II is presented here.

***All guidelines should take into account the assessment of the individual to assure health and safety and to promote quality of life.***

- 1. The individual's opinion should be taken into account when following these guidelines whenever possible.**
- 2. The preferred form of living arrangement is in a PWS-specific environment.** This allows for consistency in treatment and a sense of fairness to the individual. All attempts to create a family like environment including their own space regardless of group size is important for the person with PWS to have a sense of belonging as well as a place to disengage from group living whenever necessary. In addition it is also vital for the individual to have a choice of vocational opportunities, as enjoyment and fulfillment of one's own day is crucial for anyone to feel productive and an important member of their community.
- 3. Optimal success for the person with PWS will be based on the type of supports that are in place.** Those supports require an understanding of the unique needs associated with PWS. All areas of a person with PWS' day should have proper supervision to assist with those unique needs. There

needs to be a 24 hour type of supervision in place in either the form of direct caregiver support, alarms or security systems that alert caregiver or monitoring agency that the individual has either left the designated area(s) or that they are in need of assistance. A combination of the above may also be used.

4. **At times additional support may be needed due behavioral issues or need for assistance from another caregiver.** Each supportive environment, both home and vocational, should have a system in place that allows for quick response from another caregiver to assist in the need at hand.
5. **Routine and consistency is essential to the successful living for a person with PWS.** Inclusion of their ideas and preferences should be part of the planning of the structure of their regular routine. Meals should be managed and structured in a formal way to ensure that everyone knows the plan of how meals are to be planned, monitored, and served. It is necessary for every person with PWS to have a diet plan that can be followed by all. Exercise should also be a part of their daily routine including some levels of movement or fitness every day with the amount of time to be determined on individual basis. Healthy weight management is also an important part of monitoring the progress of supporting a person with PWS. Weights should be taken at a minimum of once per week based on history.
6. **Structure of the home for a person with PWS should have some form of house rules for living with others.** This allows for everyone to understand and remain consistent with those guidelines, expectations, and boundaries that are needed when in a group living environment. Another area is the complete security of food, money, and medication which should be locked and managed by caregivers.
7. **To assist in overall self esteem it is important for everyone to feel a part of their community.** Inclusion of those activities within the community should be individually assessed to determine environmental supports that may be needed. Understanding the person's own level of independence will assist in maintaining required needs for safety. Living in a community should include good neighbor relationships, it is important to create systems explaining about PWS.
8. **It is essential to have individual behavior management plans ranging from positive motivation to crisis management.** This once again allows for consistency and proper understanding of that specific individual's needs when that person may be in a crisis or to simply avoid or redirect the situation.
9. **Training of caregivers is key to the success of the individual with PWS.** Caregivers need a good understanding of the complexity of the needs of the

syndrome as well as the compassion that is needed when supporting a person with PWS.

10. **Maintaining healthy relationships once the person with PWS is outside their family's home is important for the person with PWS.** Contact and involvement with family and friends may need additional support and guidance from caregivers. That support may consist of teaching relationship building and in some incidences may require additional counseling. Human sexuality may also require support and education.
11. **A clear structure is required to develop and maintain a healthy and stable relationship between the parents, the individual, and the caregiver.** These communications are most effective if they are team-based and have pre-determined professional boundaries established.
12. **Due to the potential for life-threatening situations, it is imperative to create an environmental structure that allows access to comprehensive medical services.**
13. **To support self determination as it relates to environmental structures, choices should be offered to assist the person to individualize their room while promoting personal safety.**
14. **In order to provide effective support services there needs to be an established form of communication and information systems in place for everyone involved in the care and treatment of the individual.**

## **Conclusions:**

Across cultural lines, all participants agreed that in order for an individual with PWS to succeed and live a healthy and productive quality of life that a basic level of support, conducive to promoting safety and personal growth, must be in place prior to providing care. It was determined that **environmental supports were non-negotiable**, including but not limited to;

- food security,
- daily schedules,
- personal growth and development plans,
- trained caregivers, and a
- continuum of care throughout their day.

It was also agreed upon, that it is imperative that as care providers, we continue to explore this cohesive collaboration of efforts through on-going education, training, and awareness.

QED

# PWS MEDICAL CHECK LIST

It is recommended for all persons with PWS to have a **medical examination at least once a year**. A pediatric specialist should see children with PWS at least every 6 months. Many medical conditions can be detected through these examinations, and treatment can be initiated as soon as possible. For persons being overweight or suffering from a previously diagnosed medical condition, the frequency of medical examinations might be more often.

Below is a list of what is proposed to be checked by the doctor. Plan the visit and write down your questions before you go.

## TO BE CHECKED OR DISCUSSED WITH THE DOCTOR:

- Height and weight
- Blood pressure
- Lungs, heart and circulation
- Extremities: oedema ?
- Mouth: cavities or teeth grinding (bruxism)?
- GI: constipation or other abdominal problems, reflux? rumination? rectal picking?
- Skin: skin picking, ulcers?
- Joints: hips, knees and feet; the shoes are ok?
- Back: scolioses ( S form of spine) and/or kyphoses ( over bent back)  
Need of physiotherapy?
- Vision: need of glasses?
- Sleep (including daytime sleepiness, sleep attacks and cataplexy), breathing abnormalities during sleep (sleep apnea), or behavioural abnormalities during sleep?
- Genital: pubertal development in adolescents? sexuality in adults? need of information and protection from abuse?
- Urinary: daytime or night time urinary incontinence (enuresis)?

### Referral to specialists:

Teeth: Visit to the dentist every 6 months or more often as recommended

Food/diet: Consult with the dietician every 6 months -1 year to secure an individualised diet; more frequent checks for ongoing weight loss or weight gain.

Vision: Check by ophthalmologist every 3-5 years, or as recommended

### Blood tests:

Thyroid function and Hb-a-1-c (blood glucose profile)

Calcium and vitamin –D

Adolescents and adults: sex steroids

If growth hormone treatment: IGF-1 and liver function

### Other diagnostic tests:

Sleep studies (overnight) and possibly multiple sleep latency test (daytime sleep study) if narcolepsy is suspected

DEXA scan for detection of osteoporosis

## WHAT PARENTS, CAREGIVERS AND YOUR DOCTOR NEED TO KNOW ABOUT PWS:

Many persons with PWS will not show the same symptoms as do most other people when suffering from various medical conditions. This is ascribed to an abnormal hypothalamic function. A very high pain threshold, an abnormal temperature regulation and inability to vomit can delay the diagnoses of severe medical conditions; the person does not show the signs that the doctors usually see and ask for when examining the person for a disease. A good resource is the Medical Alert booklet: [www.pwsausa.org/support/medalert.htm](http://www.pwsausa.org/support/medalert.htm), or contact IPWSO for a copy in your own language ([www.IPWSO.org](http://www.IPWSO.org)).

### Below is given some examples of medical problems associated with PWS:

#### Obesity:

Obesity is one condition that demands medical attention. Per definition, an adult is obese (severe overweight) if the body mass index (BMI) is above 30 kg/m<sup>2</sup>; the BMI values for children are lower and age dependent. Obesity predisposes to other medical problems, especially in PWS. These include diabetes, sleep apnea, lymphoedema, metabolic syndrome and obesity hypoventilation. Early detection and prompt treatment can prevent these problems from occurring.

#### High pain threshold:

1. Fractures can be overlooked: Complaints can be few and normal pain reactions, even when examined by the doctor, can be missing. X-rays can be needed more often in PWS than for other persons after a trauma.
2. Abdominal diseases can be overlooked: Complaints can be few, presenting only as decreased appetite, which is very unusual in PWS. Be aware of signs of a distended stomach and the possibility of a recent big meal (holidays, parties, etc. ) There is risk of gastric distension and rupture in PWS, especially after uncontrolled eating. But this can occur anytime or during infection with an intestinal virus. Vomiting, if it occurs at all, is a sign that requires immediate attention. ***If a person with PWS who usually likes to eat suddenly does not want to eat, a medical examination can be life saving, if the reason is due to severe abdominal disease.***
3. Rectal picking can be overlooked: Rectal bleeding can be a symptom of serious gastrointestinal problems. Obtaining an accurate history of rectal picking can eliminate the need for some medical diagnostic tests. Vaginal picking can also occur, and this is frequently overlooked as a cause of vaginal bleeding.

#### Temperature instability:

Many persons with PWS do not have a fever when having an infection. The person's body temperature does not say anything about how serious the condition is. Elevated body temperature can also be seen without infections in PWS. People with PWS do not adjust to environmental temperature, so they may not dress appropriately when going outside. They are more likely to get frostbite or overheated. Some individuals develop very low body temperature for unknown reasons, and this requires medical attention.

#### Different reactions to some medications:

Because of a small lean body mass, there is a great risk that a person with PWS is given a too high dose of a medication, which again can give unwanted side effects. Often it can be

recommended to start with half of the typical dose especially concerning drugs with psychoactive effects, such as medication for epilepsy and allergy (anti histamines). Blood tests for some of these drugs can also tell if dose is correct. Because of the higher fat mass, some medications may take longer to get to a therapeutic level or take longer to get out of the system when they are discontinued. People with PWS are more sensitive to psychotropic medications, and doses should start low and go up slowly while mood and behavior are carefully monitored. Mood activation can occur with medications affecting serotonin, and this can look like severe mood dysregulation or cyclic psychosis, and it may go away when the drug is discontinued. See [www.pwsausa.org](http://www.pwsausa.org) for the **Psychiatric Alert**.

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