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IPOSO

WAVELENGTH

Newsletter for Parents and Professionals

...Making a World of a Difference

IPOSO INTERNATIONAL PRADER-WILLI SYNDROME ORGANISATION

IPWSO

Our Mission

IPWSO is politically neutral, with no discrimination as to race, sex or religion. It supports member associations in their efforts to:

IMPROVE the quality of life for all people with Prader-Willi syndrome and their families.

IMPROVE the physical and mental well-being, socially as well as occupationally, of all people with Prader-Willi syndrome, so that they may, according to their wishes, lead a life as normal as possible and be in a position to achieve their full potential as allowed by the constitution of their country, and what is set out in the Declaration of the United Nations on the Rights of Man and of Handicapped Persons.

IPWSO shall endeavor to co-operate and/or affiliate itself with other organizations with similar objectives.

IPWSO shall also act as a liaison to collate and disseminate PWS material for its members by organizing international conferences and by publishing newsletters. By so doing it aims to:

STIMULATE international co-operation on PWS research projects on the origins, management and prevention of the Prader-Willi syndrome.

ENCOURAGE national associations to exchange and share their PWS projects and experiences.

FOSTER the foundation and development of new national PWS Associations.

ENCOURAGE the international exchange of people with PWS and of those involved with their care.

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From the Editor:

Greetings from the South Pacific to all our members throughout the world. I am delighted to pick up the reins as Editor of Wavelength once more. Wavelength has been an important mouthpiece for IPWSO since its inception back in 1997 and now, some 10 years later, it is even more important as we need to keep up with all new developments in the PW world. So much has happened in the last 10 years, not only the increasing development of our international PWS organisation as it has grown and expanded, but also in the scientific and medical world as more and more becomes known about that tiny little deletion on the 15th chromosome which causes us all so much bother.

My daughter, Francie, is now 23 years old and was diagnosed at age 3 (the earliest diagnosis in New Zealand at the time) by me from an article in the Australian Woman's Weekly which I took along to a very disbelieving professor of medicine at Wellington Hospital who immediately diagnosed me as an "over-anxious mother". I am delighted that things have progressed a long way since then. Everything that I know about PWS – in fact nearly everything that I have learned about life - I have learned from my daughter who, although has taken me on a very rocky journey at times, has led me to the conclusion that scientific discoveries have just about caught up with her.

It is important that both scientific and medical advances are covered in this magazine, and it is equally important that parents' voices are heard too. We will try to publish them side by side so that, in true IPWSO style, both professionals and parents can advance together with one voice.

LINDA THORNTON
National Director, PWSA (NZ)

All contributions to Wavelength are welcome. Publication dates: January, May, Septemper. *Views expressed in this newsletter are those of the contributors and not necessarily IPWSO.*

C O N T E N T S

PRESIDENT'S PREAMBLE

IPWSO...Making a World of a Difference: Working together for a Brighter Future

Welcome to the new Wavelength, a newsletter designed at your request. Within these pages, you will discover a whole world of PWS and the power of people from many nations sharing information, joining forces to bring the dream of a better life for people with PWS to fruition.

Our new IPWSO Board of Directors theme, IPWSO ...Making a World of a Difference; Working together for a Brighter Future is evident in the accomplishment of the last 5 months. With the successful PWS International Workshop and Conference in Cluj Romania as our springboard, we are off to a great start. The many components of this inspirational event, including the Scientific, Parent and Professional Conferences, have already enhanced interdisciplinary global collaboration on many aspects of research, management, and daily care. One of our parent delegates wrote that attending the conference was like being "on a highway of information and trying to catch each car speeding more than 100 km/h by bicycle!"

In addition to distributing our IPWSO educational packets, we recently sent copies of our new portable life-saving Medical Alerts Booklets to all members with the offer to format translations in their native languages. Our PWS educational booth spread awareness at the prominent European (ESPE and Latin American (SLEP) Pediatric Endocrinology meetings in Finland and in Argentina.

Our first diagnostic Certification Course successfully launched the development of diagnostic labs in Pakistan and in India. While we are helping nations learn how to make diagnoses we are continuing to provide free services in countries where diagnosis of PWS is not possible. We met families in Finland and spoke at the First Estonian Conference.

Our future projects will include expansion of our "speedy information highway," developing more educational opportunities and services. Highlights of our spring and early summer calendar include The First Asia Pacific Prader-Willi Conference hosted by the New Zealand and Australian Prader-Willi Syndrome Associations in Wellington, New Zealand, March 1-2, 2008 and the First PWS Caregivers Conference in Herne, Germany, June 3-5, 2008.

I hope you enjoy reading about our activities in this issue and welcome you to join us in Making a World of a Difference!

Pam Eisen

PAM EISEN
IPWSO President

IPWSO International Conference in Cluj Napoca, Romania June 21-24, 2007

Three Unforgettable Days

by CELESTE MARTINS DA SILVA
Vice-Presidente da APPW Associação Portuguesa de Prader-Willi
Lisbon, November 2007

It was a great honour to participate in the 6th IPWSO Conference, representing our PWS Portuguese families. Our Association was created in May 2007 by a group of parents, so it was a great pleasure to be invited.

It was three unforgettable days especially because we could exchange practical experiences with parents from the entire world. Besides that, those 3 days were days of intensive learning about the scientific and medical knowledge of PWS. Every talk was important, and it's impossible not to draw strength from the messages of hope and confidence that Giorgio Fornasier, Jean Phillips-Martinsson and Pam Eisen gave us. Their smiles and good humour were also very important!

The medical talks related to PWS as a genetic dysfunction and also focused on the good medical care and vigilance needed, as well as the basic daily care to protect and assure the health and well-being of people with PWS during their life. The presentations were accessible and clear, and I felt that there was a great intimacy between parents and scientists. They demonstrated an understanding of the anguish and doubts that we feel so many times when we don't know what to do.

Until the IPSWO Conference my experience of PWS was only as a mother of a teenager. The conference helped me a lot in that area and I learned much from parents all over the world who enthusiastically related their experiences and taught us how to deal better with our beloved children. I learned more about the PWS in those 3 days than in the last 16 years. Putting into practise what I have learned will help me to help other Portuguese families.

The APPW is a very new association, is still growing, and we need to involve more families and health professionals. We know that we have a long way to go, especially in relation to public health institutions, school education, as well as the Portuguese government where we need to argue for citizen rights for our Portuguese families. For example Growth Hormone Treatment is not yet available for our children and there are schools that have difficulties in under-standing the necessity of social integration of children with PWS - they tend to choose the easiest way and send those children to special schools without a proper evaluation. Our fight would be much more difficult if we had no contact with the PWS associations in other countries who are also trying to obtain legislatives changes, medication and equipment specifically for PWS. We are very proud to belong to this great world family and we will do all to honour it!

JEAN PHILLIPS-MARTINSSON, the Honorary President of IPWSO and the founder of IPWSO.

IPWSO International Conference in Cluj Napoca, Romania June 21-24, 2007

Improving the Lives of Those with PWS

by SUSAN HENOCH
USA Parent Delegate

One in 15,000 often feels like a lonely statistic, but when parents, professional care providers and scientists met this summer for the Sixth International Prader-Willi Syndrome Organization (IPWSO) Conference, we immediately coalesced into an extended family. Representatives of Prader-Willi Syndrome Associations from 44 countries came to Cluj, Romania for three days to attend sessions, search for information and meet informally for support and encouragement. We focused on one issue: improving the lives of people living with PWS.

I had the honor of attending the Conference as the new Parent Delegate from the United States, which allowed me to engage in lively conversation with scientists about their latest findings, with providers about new program components and with parents about everyday life. All of us connected through our experiences living, working, struggling and loving our children and clients. It was especially heartening to exchange information with those from countries just beginning to recognize PWS.

In between sessions and during meals, we were able to meet informally, one of the most valuable aspects of the Conference. Since this was my first IPWSO Conference, I attended as many sessions as I could to learn about the latest medical research and creative ideas for caregiving. The primary message from the scientific sessions was that although there is no cure yet for PWS, effective treatments are gaining widespread use, such as early and long-term treatment of growth hormone and new, more targeted generations of psychotropic drugs. More subtle avenues of research, particularly in the field of genetics, are under investigation, now that there's a fuller understanding of the basic outlines of PWS.

Searching questions are being explored: What are the connections between genetic typology and behavior and what is their significance? Do these behaviors change over time and if so, why? What is the mechanism mediating between chromosome 15 deficits and behavioral issues in the first place? What role do environmental interventions that decrease stress play in mitigating or delaying onset of more severe emotional problems?

Hopefully, the answers to these questions will translate to real life strategies contributing to improving the quality of life for those with PWS. Scientists are already recommending that home and group programs implement changes that contribute to less stressful day-to-day environments.

Hopefully, the answers to these questions will translate to real One way to do so is to take what one scientist refers to as the "whole person" approach, promoting strengths and positive aspects of the individual, rather than dwelling on maladaptive behaviors. Certain interventions contribute to enhancing a person's wellbeing, such as recognition of many PWS individuals' nurturing streak by making sure they have pets with whom to bond and care for, friendships and meaningful work. Engaging in activities that increase a sense of "flow", during which a person's sense of time and self disappear into the focus and enjoyment of the activity itself lessens depression and increases the overall sense of health. These are some of the findings I found most hopeful.

During the Professional Care Provider sessions, residential directors described their programs, details of which differed surprisingly from country to country, depending on philosophy, cultural norms and values, moral beliefs, and most importantly, funding. All agreed, however, that to be successful, a program must provide ongoing staff training and development. Several countries offered inspiring examples of good care. Sweden - committed to the belief that people with disabilities share the same rights and privileges as all other citizens - provides supervised spacious individual apartments with common areas for dining and recreation and meaningful work, such as weaving and printing workshops.

DORICA DAN, the president of PWSA Romania and the hostess of the Conference.

Improving the Lives...

Residents of a group home in Denmark collect and repair toys to give away to children. The first PWS group home opened in Switzerland in 2003, with residents living in their own rooms and working in textile and other workshops in a nearby institution. A program in Israel meets the needs of from Orthodox to Jewish citizens with PWS by providing separate housing for men and women, as well as a strictly kosher diet. This program emphasizes independent living and social skills training, swimming, computer education, arts and crafts and annual camping trips.

The United Kingdom has increasingly moved away from large institutions to smaller homes for the people with disabilities; residents in one PWS home work in an onsite print shop, textile printing shop and gardens. German PWS programs are based on a core belief in integrating and balancing the body and soul, improving behavior by structuring the environment to expand individuals' social competence and self-control which contributes to their overall self-confidence and body consciousness. The government fully funds a variety of living arrangements, including group homes, apartments with staff living across the street and support for people living at home. Some residents work in sheltered workshops packing, welding or drilling.

Everyone took home ideas to discuss and perhaps incorporate into their own programs. Parent delegates met for an afternoon session to share more intimate stories. The room filled with tenderness as mothers described their children's artistic talent, their generosity with the people they love, their enjoyment taking care of babies and pets. We unconsciously nodded our heads as others talked of the need for people with PWS to be intellectually challenged in school to the best of their ability, to be involved in meaningful work even in sheltered workshop settings, to have opportunities to express themselves artistically, to be included in their communities. Parents also voiced some of the ongoing frustrations their children contend with, such as not being in charge of their own money, the lack of opportunities to participate in work in the community, their difficulties fitting into society and developing and maintaining friendships. During this vital session, we as parents actually participated in and helped create for one another the environment, values and experiences we want for our children. It was exciting to be part of the worldwide effort to confront PWS.

Everything is Possible!

Dear friends,

I live in Slovakia, a country of the former block of socialist countries, which Romania was also a part of. Therefore I was very surprised the first time I read that an international conference is to be held in a former socialist country. I thought, this was a kind of a printing error. But when I was in Cluj and saw how everything was organized and managed, I realized that the slogan "everything is possible" is correct. Because if we have hope, strength, motivation and desire to achieve targets in our lives, we can turn our effort to reality. Therefore I thank to all the people who prepared this conference for giving us the hope of a brighter future for both the parents and our children.

Two representatives of the Slovak national association of PWS (www.pwsyndrom.sk) – myself and my friend Ludmila (both mothers of children with PWS) attended the international conference of IPWSO in Cluj in June 2007. We met plenty of parents there with similar stories who have to face an everyday struggle with the syndrome. Since our children are aged 5 and 7, we found it very interesting, especially the sessions for parents who have children of the same age, to be able to shared our experience and suggestions related to the right school choice, behavioral problems, weight control, suitable exercise program and suggestions how to deal with unforeseeable situations. During these and many other special sessions I found answers to many of my questions and realized that there will always be someone who can help me or advise me if there are issues related to PWS in the future. This gives one a feeling of security and hope and also the strength and love for our special children.

Maria from Slovakia

PWS Diagnostic Certification Course

Conducted at Baschirotto Institute for Rare Diseases (B.I.R.D.)
September 10 through September 25, 2007
Made possible by an IPWSO Grant

GIORGIO FORNASIER, IPWSO (left),
PAM EISEN, IPWSO,
UROS HLADNIK, B.I.R.D.,
ANNA AND GIUSEPPE BASCHIROTTO,
B.I.R.D.

by PAM EISEN, President IPWSO, November 24, 2007

As IPWSO members have increased, we also have discovered a need for more member services. In 2002 we began to offer free diagnostic services in cooperation with the Baschirotto Institute for Rare Disease (BIRD) to members who did not have this capacity in their own country. Using a simple blood sample protocol, doctors and families from emerging countries around the globe have expressed their gratitude to IPWSO for "making the world of a difference" to these families.

As this program has expanded, we realized that the next step was to develop these services at a reasonable cost and at a high quality level in countries who were ready for this next step. India and Pakistan were eager to build labs and had doctors and biologists ready to learn the techniques. To meet this need I searched for funds and with the assistance of Bob Brown, our business and technical advisor, submitted and won a Pfizer Medical Educational Grant to provide 60 free diagnosis and to provide our first IPWSO PWS Diagnostic Certification Course. Our time frame was short and we had many aspects to develop. Thanks to the help of Giorgio Fornasier, our Director of Program Development, Anna and Giuseppe Baschirotto and Dr. Uros Hladnik Director of the diagnostic lab at BIRD , working as a team, we were able to develop a stellar program which already has already facilitated the building of a foundation of services for two of our member countries, India and Pakistan. Considering this large population where there are only a few cases of PWS known, we can only imagine the general awareness of genetic disorders and the positive impact this course will bring to families who are now in the dark.

PWS Diagnostic...

by GIORGIO FORNASIER Director of Programme Development, IPWSO

Since the year 2000 IPWSO has its office located at the Mauro Baschirotto Institute for Rare Diseases (BIRD) near Vicenza in Italy, where we can also have access to their Genetic Molecular Laboratory services.

Since 2005 we are offering free diagnosis to countries where this service is not available to achieve the vital objective of early diagnosis. We made several methylation tests already for patients suspected to have PWS from all over the world. The other dream we had, was to organise diagnostic courses to teach geneticists and biologists how to make PWS diagnosis in their own countries.

In 2007 our President Pam Eisen thought it was time to initiate a new grant for a Diagnosis and Diagnostic Certification Course. She signed and submitted the contract to Pfizer. while together with Bob Brown and Giorgio Fornasier she studied the contract to be sure that we were following all stipulations.

It was a pleasure to tell Anna and Giuseppe Baschirotto and Dr. Uros Hladnik (Director of the Diagnostic Lab at BIRD) that our application had been accepted and the grant paid. While we were pursuing candidates, the Baschirotto's have been working on the details of the curriculum and on setting the date. BIRD was planning a 4 weeks rehabilitation period for adults with PWS the last three weeks of September and the first week of October. We thought this was an ideal period to plan the diagnostic course, so the attending doctors could see real patients and have fresh blood available.

For the candidates, we chose two doctors Pam had been corresponding with for a long time, one from Pakistan, and the other from India. In light of the huge underserved population and from our experience, we agreed that these countries would benefit the most.

The doctor from India was Anil Jalan. He is Chief Scientific Research Officer, Navi Mumbai Institute of Research in Mental And Neurological Handicap, (trained at University of Vienna and at Graz in Austria & at Nijmegen University at Holland) and has been sending samples to BIRD in the past. With approval of his institution, he said to be prepared to purchase all necessary equipment for diagnosis and to be committed to helping our families get reasonably priced diagnosis and treatment.

Our Pakistan candidate was initially Dr. Tariq Moatter, Aga Khan University hospital (AKUH), who has been working on the development of a cytogenetic lab at his hospital. Unfortunately he could not attend for family problems, so he sent his assistant Dr. Farzana Murad in his place.

The Course was very successful and both candidates returned home very enthusiastic. As an immediate follow up, Dr. Anil Jalan bought a new building and ordered all equipment to start methylation tests for all India patients as soon as possible. Initially he will send the same samples to BIRD for a double check and will continue his close co-operation with all the staff at the laboratory in Italy, not only for PWS, but also for other rare diseases. We must be very proud of that.

Dr. ANIL JALAN, Chief Scientific Research Officer, Navi Mumbai Institute of Research in Mental And Neurological Handicap and Dr. FARZANA MURAD, Aga Khan University hospital (AKUH) together with IPWSO President PAM EISEN at B.I.R.D. Institue Italy.

PWS Diagnostic...

Friends,

I am Anil from India. Let me introduce myself. I am Dr. Anil B. Jalan, a Paediatrician from India, who completed MD in Paediatrics from Bombay University in 1988 and perused academic interest in genetics and metabolics after that. I was fortunate to receive training at various European Universities including Vienna. During this academic adventure, I came across many PWS patients and developed interest in the management of such genetic and metabolic disorders. Back home, we established an institution " Navi Mumbai Institute of Research In Mental and Neurological Handicap" and we deal with at least 500 new patients every year. More than 95 % cases are Paediatric but we also see some of the adult and adolescent patients as well. In India, our guess is we must have 15,000 PWS children born every year. Sadly, the diagnostic and management services are very poor in this particular aspect.

Fortunately one day like a ministering angel, Pam called me up and informed me that I have been selected to receive further training in diagnostic and management aspects of PWS at BIRD in Italy. I had known of BIRD and had interacted with Dr. Uros Haldnik and knew about their great work. I was bubbling with joy and consented to this training immediately. When I reached BIRD, I was astonished to see many children with PWS. Then the actual training session began which was quite exhausting. The team at BIRD is headed by Dr. Baschirotto and Dr. Uros. Both of them took a keen interest in my hands-on training. The junior staff, especially Gessica, Veronica, Paula, Eleonora, Isabella, Erika, and Chiara, all took special interest in my well-being and training. Dr. Uros was especially motivated and devoted, and has expertise in English.

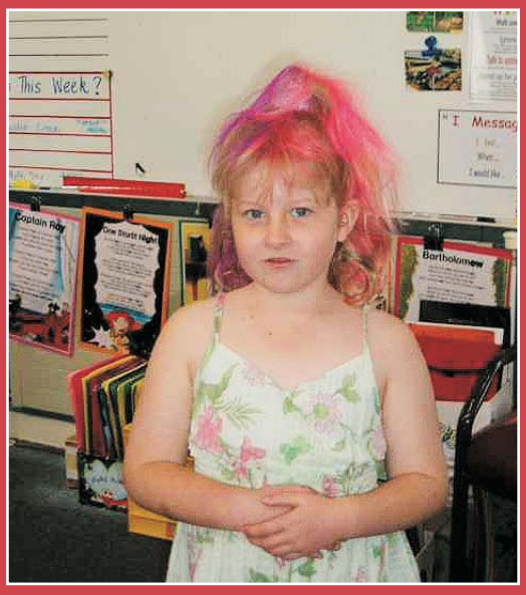
For two weeks I was given excellent hands-on training in Laboratory aspects of PWS diagnostics. Simultaneously I was also involved with meetings and discussions with the children there who had PWS. It was beyond my imagination to see the actual multifaceted management of this condition. Though I had worked in Labs of Vienna, Nijmegen etc, this was for the first time I had had an opportunity to see the actual play therapy, music therapy, their dietary management, and loads of activities which these children were involved in during their 3 weeks stay at BIRD. This opened my eyes and now I could envision the actual needs of children with genetic disorders. After 2 weeks of tender loving care I had to return back home with heavy heart, but I still wish to go back and learn many more things. I sincerely thank Pam, Giorgio and entire IPWSO team for making my dream come true.

Now that I am back home, we have started implementing all that I learned. We have established a separate unit of Molecular diagnostic lab along with other services provided by our institute. Soon we will be doing fully-fledged methylation diagnosis of PWS and management.

As we always say in Genetics
Cure - Rarely,
Comfort - Most of the times
Care - Always

Thanks a lot for this wonderful opportunity.

Anil



Wacky Hair Day

by Emma Auckland, New Zealand

On Friday 23 November it was wacky hair day. My Mum sprayed my hair with purple and pink. Some people stuck gel and hair ties in their hair. We sponsor two children who live in Africa. We give money to World Vision.

Research View Updates on PWSA (USA) Research

by JANALEE HEINEMANN, Director of Research & Medical Affairs (PWSA, USA)

We have recently received updates on some of the research PWSA (USA) is sponsoring, so I want to share this information with you.

Sex Hormones/Sexuality

Drs. Varda Gross-Tsur and Harry Hirsch of Israel are studying gonadal function in PWS and sexual behavior. To date, they have obtained blood samples from 79 children and adults with PWS, and are doing ultrasounds of the ovaries and uterus, etc. It is already clear to them that our PWS population represents a wide spectrum of reproductive hormone function and that sexuality is a very important issue. What this means to us: We now know of at least three documented cases worldwide of women with PWS giving birth (two mothers with deletion had children with Angelman syndrome. One mother with UPD has a child who appears to be typically developing.) We can no longer say all are sterile, but this study will help us understand the range of reproductive hormones. It will also define further (as we have written in the PWS management book) the sexual and relationship needs of those with PWS.

Psychotropic Medications

Although there have been significant advances in — and application of — psychotropic medication with PWS, reactions to these medications are dramatically variable. Dr. Elisabeth Dykens, Elizabeth Roof and colleagues are working to understand reasons for such variability in the response to psychotropic medications. They predict it may be related to genes involved in drug metabolism. For example, poor metabolizers may have negative side effects if given too high dosages. Their study identifies the CYP450 enzyme status in those with PWS. What this means to us: With this study, we should be able to identify more accurately what drugs work better and in what dosage for PWS.

Genetics

Dr. Merlin Butler is working on the expression of four genes between chromosome 15 breakpoints BP1 and BP2 in PWS and the impact on cognition and behavior. Preliminary analysis of the data indicates more expression of each gene when two alleles (alternative forms of a gene) are present, but a significant amount of variation in expression from individual to individual regardless of the copy number. New technology allows identification of different size deletions within the type I and type II deletion subgroups, which may help explain further the differences in expression. What this means to us: More sophisticated and accurate testing is becoming available which will help define the genetic reasons for the differences in behavior and cognition within PWS.

To view our latest PWS research news go to www.pwsausa.org

Gastric Rupture and Necrosis Deaths in PWS

PWSA (USA) recently funded a study of causes of death in people with PWS. Over several years, we recognized that 3% of reported individuals had died due to rupture and necrosis of the stomach. In addition, four other people were suspected to have gastric rupture. After reviewing the history surrounding those with gastric rupture and necrosis, vomiting and abdominal pain were often reported, which was rarely reported in other PWS deaths. The reports prompted the committee working on this project to suggest that physicians consider an emergent evaluation for gastric rupture and necrosis in people with PWS who present with vomiting and abdominal pain. Those with binging and recent weight loss may be at more risk for gastric rupture and necrosis, but further investigations are needed. Still, close supervision is recommended when people with PWS are in situations with large quantities of food in new environments (holiday events or birthday parties) so as to avoid binging episodes.

Details of this study are in the Journal of Pediatric Gastroenterology and Nutrition, "Gastric rupture and necrosis in Prader-Willi syndrome" (Stevenson DA, Heinemann J, Angulo M, Butler MG, Loker J, Rupe N, Kendell P, Cassidy SB, Scheimann A. : J Pediatr Gastroenterol Nutr 2007; 45:272-4).

Looking for the RIGHT SCHOOL

by LINDA THORNTON

There are some very comprehensive sites on the internet which set out more fully than this article the specialised care needed around educating the child with PWS. Here are two:
http://www.pwsa-uk.demon.co.uk/educatn.htm (overview for parents and educators)
http://www.pwsausa.org/Educator/index.htm (for educators)

For parents setting out along the education pathway, there are many questions to ask: Do I want my child to be mainstreamed, in a special needs classroom, or home-schooled? How successful is the school going to be at integrating my child into the mainstream classroom? Will they understand that my child with PWS will be different from any other child with Special Needs?

There is no 'short answer' to this as each child is individual even within the parameters of the syndrome. Some may show characteristics easily recognised as belonging to Prader-Willi Syndrome, and yet may show quite different abilities. Some children may show autistic characteristics as well. Most will have a combination of all. We know that having PWS means an inability to make good choices around food, poor muscle tone and growth (unless the child is on Growth Hormone Treatment), and a borderline IQ often with an inability to communicate their needs clearly.

This will pose some unique difficulties to teachers and will require good, structured support in the classroom.

Is the school your child is at, equipped to handle this?

Primary education

People with PWS are quite able to be educated. If the right support systems are there, theoretically there should be no problem successfully integrating a child with PWS into the mainstream schooling system. Knowing whether the school you choose can provide this support is the first question you need to ask as your child starts school.

Normally happy-go-lucky, friendly, willing, and helpful at primary age, children with PWS don't cause too many problems that aren't able to be overcome with careful planning. It is absolutely essential that at every step of the way, the parents are included in decision-making, programme planning, and Individual Education Programmes. Parents should be encouraged to bring along an advocate to support them during this time. Most PWS Associations will provide help with this.

Structures and Ground Rules

Ground rules established at the beginning of a child's schooling will make it easier throughout the years to continue to provide support. Care must be taken not to 'fail the child' around food. It is an absolute given that if food is easily accessible (whether in the locker rooms, staff rooms, rubbish bins, classrooms, cooking rooms etc) it will be a temptation to the child with PWS and will be eaten.

Is the school willing to provide these structures?

Structures around food must start from day one. Whether the teacher decides to make the locker room food-safe, or take all school lunches into the staff room for re-distribution, it is something that has to be put into place immediately. Children with PWS are generally rule-orientated. They like structure and respond to systems. They dislike changes in routine (unless told about it beforehand) and not knowing where they stand. They don't mind rules around food so long as they are fair, and so long as the child is not seen to be deliberately marginalised.

Individual Education Programmes

IEPs may differ from school to school, and country to country, but they need to include the support of all those concerned with the child's development - including the child him/herself. Programmes need to be fully developed, catering for every possible need, culturally sensitive, structurally set out with specific goals. If this is not done early on, the incompleteness of service delivery will continue to be flawed throughout the student's entire school life. IEPs need to include such things as:

- description of the student's conditions and needs*;
- specification of short-and long-term goals;
- statement of expected timetable for goal attainment;
- listing of all services to be provided
- identification of all service delivery agencies and personnel involved;
- specification of how services are to be delivered in the normal, or least restrictive, environment;
- statement of objective criteria and schedules for evaluation;
- provision for periodic review

**This is an area where the school's knowledge may be inconsistent.*

Looking for the...

To Label or Not to Label?

With the current dislike of 'labeling' some parents choose not to identify Prader-Willi Syndrome, but rather aim for the specific needs of the child to be identified when needed and support then put around that need. Although respect for the privacy of the child and family is absolutely necessary, open communication between school and parents is essential. The more the school understands what it means to have PWS, the more likely they are able to help.

There are arguments for and against this (privacy) situation; one could argue firmly for the rights of the teachers to know the characteristics of the syndrome so that wise and careful support can be put in place. On the other hand, if teachers are to receive information which merely states "Prader-Willi syndrome means overeating and intellectual disability", it won't help anyone. The object of your PWS Association is to educate parents just as much as providers.

Use of Teacher Aide Hours

All children with PWS should be assessed to decide the teacher-aide hours required. These may need to be around the classroom setting, but they will also need to include supervision at lunchtimes and breaks when accessibility to food may become an issue.

It is not unknown for children to rummage through rubbish bins, pick food up off the ground, trade lunches, take lunches from classrooms, go into staff rooms looking for food, etc. All teachers need to know the situation around food.

As the student progresses towards the end of primary education, a Needs Assessment for secondary education must continue to take into account the issues around food. This is where (certainly in our country, New Zealand) the teacher aide hours become stretched - the school can no longer 'share' students and teacher aides; supervision can become quite slack and the student will undoubtedly find access to food sources.

Secondary Education

Decision time again - just as you thought you'd solved the problems at primary school, it is time for your child to move on to his/her secondary education and it is here that knowledge gap between the student with PWS and his/her peers will become very apparent.

What will your choice be?

- To continue with a main-stream approach with teacher aide hours, or go into a special education classroom?
- Which school offers the better choice?
- What will suit your son or daughter?
- Where do they want to go to school?

Generally speaking, the person with PWS is said to have a 'below average IQ'. However, we also need to recognise that the person often has an IQ level well above average in some skill areas, particularly when it comes to ways and means of accessing food, or asserting their rights. As the student grows older there is a strong need for life-skills education, and as the student gets to senior level, for transition and job experience.

- Will the school you choose offer the best possible choices to your son/daughter?
- Do they have a transition programme with work experience?
- Will this have good support systems?

Educational skills can taper off, or plateau, but given the right sort of stimulation and interest, the student does not need to stagnate, but can continue to learn, albeit at a slightly different tangent.

Challenges

There will invariably be challenges - both to the student and to the teachers. Behavioural challenges can often be frightening to the unsuspecting teacher.

- How well will they cope?
- Will your PWS Association offer guidance, training courses for teachers, and advice?

Good procedures must be in place both for the student and the staff. A "safe place" for the student whether it is a room for time-out or the school counsellor's room, or sitting under a tree for a short while - all quietly monitored by staff - will be necessary. Swearing, rebellious behaviour, extreme anger, trashing, smashing, running away, physical abuse, are often the outcome of a student who has been pushed too far.

The ability of a teacher to recognise the precursors, the 'triggers' that lead to this type of behaviour, is paramount. Anxiety, stress, change of routine, pressure at home, incompatible respite care, bullying and teasing by other students resulting loss of self-respect, all these can trigger an escalation in anxiety and result in challenging behaviours. To avoid the outbursts, time needs to be taken with the student (outside the classroom) to alleviate their worry, to calm them down, and to redirect their attention.

The concept of punishment will only add to the misunderstanding, fear and anxiety of the student.

- Does the school you have chosen, understand this?

It is important for the teaching staff not to build up resentment in the student - and to be able to 'forget' their extreme behaviour so that each day is started afresh.

When the bell goes...

At the end of the day, meeting the unique needs of a student with Prader-Willi syndrome may be challenging, but it can also be a successful encounter. There is a wide spectrum of characteristics in PWS, not all students display the same needs and not all have the same challenges. Some of these can be met medically (with specialised care), but all will require patience, understanding, and a desire to help the student. A wide and supportive network must be cultivated around the student with procedures that everyone follows.

Emma Goes to School

Emma is 7 years old. She goes to Three Kings School, in Auckland New Zealand, where her older brother, Tom, also goes. She attended the kindergarten that was on the same site as the school, which made it easy for her to transition. Her first teacher had already taught her brother, so there was a great relationship already established. Emma loves school and is working well within her age group. This is Emma's second year at school and her second year of growth hormone treatment.

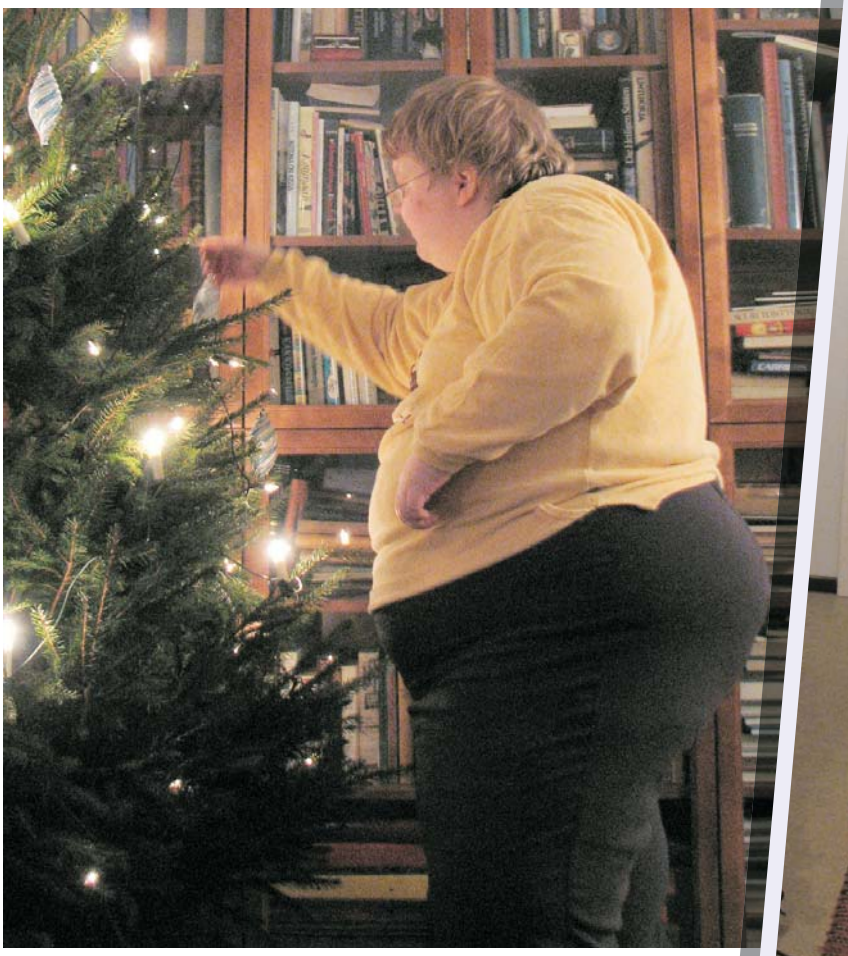
SCHOOL REPORT CARD

Reading: Emma is reading at a level above average for her age group. She continues to advance reading groups and currently is in the second top reading group in her class. Emma is self-motivated when it comes to reading and loves to read to herself at home and at school. Her comprehension has improved throughout this year. She will still read over words that she does not know, but now will attempt to sound out the word, or have a go in the context of the sentence.

Writing: Emma is working well with her writing. This year has been a particularly good growth year for her. Emma would often get stuck with her writing and move from idea to idea in the same story or repeat herself, but she is now able to complete a whole story without getting muddled up. This has been helped along by Speech Language Therapy. As Emma's verbal speech has improved and the order of forming both words and ideas in her head has become more clear, so has her written work.

Maths: Although Maths is a struggle, Emma is within the range for her year and age group. She has been working hard with mathematical ideas and concepts. She can now count to 100 in tens and fives.

Socialisation: Emma used to spend a lot of time with older children, or the new entrants because of her difficulty with verbal language and her frustration of not being able to make herself understood. This year, with the help of Speech Language Therapy and excellent staff, Emma's language has improved to the extent that she is establishing a group of friends from her class and maintaining her own group of friends.



Being Overweight is not a Death Sentence,

and it doesn't have to be a life sentence.

by TIINA SILVAST, Finland

But how to get them out of that prison? Everyone who knows anyone with Prader-Willi Syndrome knows how extremely difficult it is to get them to agree to do something about losing weight. They will tell you they are already on diet, in fact, they have been that all their lives.

- "And how about a little walk?"
- "Can't do it , it's much too hard work and I'll get sweaty!"

My husband and I have been fighting this struggle for years with our daughter who is 26 and very obese and has never been on GH treatment. We have been watching her get heavier and heavier despite the diet we were trying so hard to make her follow. She was even trying to follow it herself from time to time - she didn't want to be obese - but on the other hand, if we mentioned the idea of doing something more about it, she usually got upset , started crying and...well, you all know how it is with PWS.

It was so sad, and little by little I started to prepare myself to accept the fact that she may not live to be very old. But I didn't believe she would die in the near future, definitely not today or tomorrow.

Then came the big day when I heard Linda Gourash's alarming lecture about oedema in Prader-Willi, the day that completely changed our lives. Linda gave this important lecture in IPWSO Conference in Cluj, Romania last June. First I was shocked to hear that those who are obese have an enormous amount of fluid in their body, not only in their legs, but it can also be in rest of their body. The level of fluids tend to gradually rise up the body and it is critical that it doesn't reach the level of the heart. That, combined with hypoventilation, or apnoea, is a life threatening condition in PWS terms at any age!

I knew immediately it was my daughter that Linda Gourash was describing as seriously ill. Her tissues were, as Linda said, hard, and

Being Overweight is not...

full of fluid up to her neck and she most certainly had hypoventilation: her weight was 145 kg and she was 155 cm tall.

After this shocking news my mind was at home with my daughter and the rest of the conference seemed to last forever – I was so worried about her that I wanted to hurry home.

It was all clear to me, I knew what I had to do, Linda Gourash had opened my eyes but hadn't left me in despair: there is a way out of this prison, there is a cure, a very simple one, to START WALKING and to GET RID OF THE WEIGHT, I added the word NOW in my mind.

So all I could think of was to get home and start working with my daughter immediately. Her constant fighting back was not relevant anymore. I was going to drag her up from her chair and out of the house for an hour's walk every day, and I swore I would stay next to her watching every bite she eats during the day. I was ready to bend backwards to save her life. After saying that I started to make a plan of how to make her accept this new life style of hers. The only thing I could think of was to tell her exactly what Linda Gourash had told the audience in Cluj. I knew that it would shock her, but at the same time it would hopefully make her understand the seriousness of her condition.

I arrived home from Romania at night and the very next day I told her everything. I tried to be as honest as I could, as calm as I could and as encouraging as I could. Naturally she was shocked. We both cried when she asked me: "Mom, am I going to die now?", but as soon as I told her that she will not die as long as we start walking and having a strict diet, she calmed down and said very seriously "Yes, I will do what ever is needed." So we went for a walk. We walked around the small lake and it took her an hour. She was very tired, very, very sweaty and her legs were hurting (actually they were hurting for next three months) and when she finally sat down at home she was trembling. I assured her that eventually her legs will stop hurting and the trembling will stop.

Now we have been walking ever since, six days a week, as Linda Gourash recommended. Today it takes her half an hour to walk around the lake so we've doubled the trip. Now she loves walking, rain or shine, warm or cold, she is on the track. She says she loves the way her body feels after a brisk walk - she even likes sweating, she says it's a sign of a good exercise.

I found a good calorie chart on the Internet and printed it out for her. Together we made a daily 900-1000 calorie plan for her which she follows punctually - she does have PWS after all! We also got an appointment in the Sleep Clinic of Tampere University Hospital, Finland, and, as I thought, she had hyperventilation; now she has CPAP *(a breathing aparatus to help her breathe comfortably at night)*.

As I write this it has been five months and one week since the conference in Romania and since she started this new life of hers. After one week of walking her legs, which were thick as tree trunks and hard as stones, were noticeably slimmer and softer, her 145 kg is now 118 kg and her hard tissues are now soft. We know she still has a long way to go, both in losing weight and walking in miles (in fact the miles never end, she has to walk all her life), but she is determined to reach her goal: to be and stay a slim healthy girl. She says "This time I have only one option if I wish to live...and I do wish that".

We were fortunate, my daughter is extremely motivated. But can all people with PW be motivated by telling them the whole truth? Is telling the truth the best way to motivate them?
Is it perhaps the only way, or is it the worst way? Our children are very clever and understand many things, so can they understand how crucial diet and exercise are when we tell them it doesn't have to be a life sentence?

This big portion of delicious and healthy fish soup is part of the well planned diet.

Oedema (Fluid retention) in Prader-Willi Syndrome

by LINDA M. GOURASH

Dr. Gourash is a Pediatrician who has worked extensively rehabilitating hospitalized patients with PWS

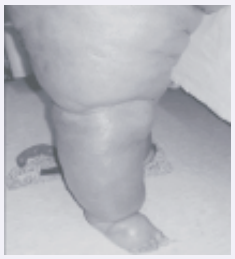
Fluid retention is usually noted first as swelling of the lower legs. In PWS this is a valuable warning sign that poor weight control is affecting the person's health. Fluid retention in persons with PWS is usually a sign of a decreased ability to breath adequately due to excessive weight ("obesity hypoventilation"). With excessive weight, breathing abnormalities first develop during sleep and can be present silently for years without any other signs that something is amiss.

Although oedema often is the earliest clinical sign of obesity-hypo-ventilation; it is frequently missed. The reason for this appears to be the visual subtlety of œdema in both the obese child and obese adult. One useful way to describe this type of oedema is by feel: the fat "gets hard" as the turgor (firmness) of the lower legs or (usually later) the abdomen increases. Tactile comparison of tissue in the lower part of the body with the upper extremities will help to demonstrate an increased density of the tissue in the lower part of the body. The level of the firmer tissue is usually well demarcated to the knees, thighs, hips, waist, or higher. This finding is less appreciable in children who can nevertheless be quite compromised by fluid retention and poor ventilation. In the absence of diuretic use, the level of oedema correlates fairly well with the severity of the nocturnal hypoxia (low oxygen). Therefore detection of a lesser degree of oedema to the knees or thighs is especially valuable as an early sign of cardiopulmonary compromise. These patients typically have normal resting oxygen saturations during the day but pulse oximetry testing during exercise will sometimes demonstrate desaturation. In the presence of any recognizable oedema, nocturnal oxygen abnormalities (levels below 92%) are usually quite extensive and may be present throughout the night without the arousal from sleep usually noted with sleep apnea.

Decreased exercise tolerance can also be a sign of obesity hypoventilation.

However, decreased tolerance is difficult to differentiate from the noncompliance with exercise often displayed by persons with the syndrome. Families do not always perceive the symptom because young children are adept at appearing to carry out their usual activities while conserving their energy. Similarly, orthopnea (sleeping with extra pillows or sitting up) and symptoms of OSA (obstructive sleep apnea) are only sometimes present.

Rapid weight gain in an individual with PWS that is not explained by increased access to food may also be the first sign of fluid retention.

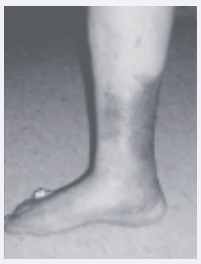


Massive oedema in 18 year old girl with PWS.

Complications:

Longstanding oedema results in chronic tissue changes of the lower body including legs and lower abdomen. The resulting venous stasis and lymphatic damage predispose tissue to ulcers, thrombosis, and cellulitis (tissue infection.)

Intervention to ameliorate the underlying condition of obesity hypoventilation is essential to avoid irreversible damage to the lymphatic and venous systems of the lower legs. There is no question that skin-picking behavior in those with PWS, while not usually resulting in infection in other parts of the body, is a major contributor to some episodes of leg cellulitis.



Two older individuals with chronic changes from long standing oedema.

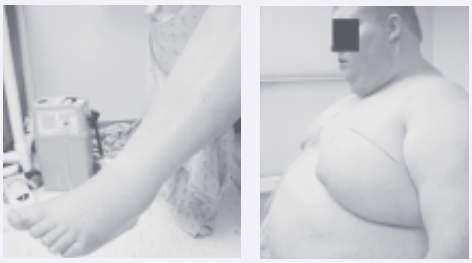
Infection (Cellulitis) of the oedematous tissue

Signs of cellulitis can be difficult to ascertain in the very obese individual since the legs are often already chronically swollen, indurated (skin thickened and hard) and discolored even without infection. A high index of suspicion and close daily examination of the legs by caretakers seeking changes in feel or appearance is essential.

Oedema...

Patients do not always exhibit fever or pain. Limited cases of cellulitis, diagnosed early, can be managed with oral antibiotics. Preventative use of antibiotics is discouraged to prevent development of resistant strains of bacteria. However, intravenous antibiotic may be necessary in severe cases, especially if there is evidence of systemic infection (fever or malaise). In all cases, an attempt should be made to identify the causative organism but this is not always possible.

Maintaining and increasing physical activity and leg elevation when the patient is sitting have proven useful adjuncts in the management of these difficult conditions. Cellulitis and superficial venous thrombosis are not reasons to limit activity; rather, the reverse is true. Patients who have ceased to walk for any reason are at high risk for thromboembolic events (blood clots) and prophylactic anticoagulation should be considered.



Despite only subtle changes in legs, this 19 year old man had oedema to the chest.

Management of Oedema:

Direct pressure techniques to the legs or use of support hose appear to be of very limited usefulness unless they are part of a full medical protocol for treatment of lymphoedema. In some cases, support stockings may be counterproductive, causing tissue breakdown from pressure or constriction of fluid outflow.

Rehabilitation to some level of ambulation is the highest priority. Ambulation without the use of diuretics is very effective in mobilizing fluid and causing a natural diuresis. This means that fluid previously contained in the tissue is moved back into the veins (by the action of the muscles contracting), returned to the heart and eliminated by the kidneys. Sometimes within days of increased activity there is evidence of increased urination and a rapid loss of weight can be documented by daily weight checks. This weight loss can be impressive (up to 4 pounds (2 kg) per day) in massively oedematous persons. Losses of .5 kg/day are more typical; this effect can be delayed for weeks in patients who are more ill.

Besides mobilizing the fluid, increased physical activity alone can greatly reduce and even eliminate the low oxygen and even the obstructive sleep apnea causing the oedema. At times it may be prudent to treat these conditions independently with CPAP or BiPAP but this is frequently not necessary if the individual can embark successfully on a program of increased daily activity.

Oxygen use especially *without* the assisted ventilation of CPAP or BiPAP is risky as persons with PWS may lose their drive to breath when they are given oxygen and die of CO2 retention. This is usually a subtle change of worsening hypoventilation that develops over hours or days. *Overuse of oxygen and of diuretics are the 2 most dangerous and common errors of rehabilitating persons with obesity hypoventilation and oedema.*

CPAP

Even persons who are very compromised from their obesity and low oxygen levels must begin to move even if it is only a few more steps per day than previously attempted. Using a walker is very helpful for the sick patient who has been very sedentary.

Using lots of encouragement and incentives, persons with PWS can be induced to walk more each day with a goal of 1 hour per day (in 2 shorter sessions). They will virtually always experience a great improvement in their oedema. If the leg swelling has been present for years some swelling of the lower legs will often remain due to the damage to the veins and lymphatics. Therefore early intervention is ideal.

Notes for Physicians

1. Pitting is usually absent even in the presence of massive oedema
2. Abnormalities on chest radiograph (X-ray picture) and even echocardiography are a very late finding long after the individual is quite compromised.
3. End stage obesity hypoventilation includes right heart failure from pulmonary hypertension and, far less often, some left ventricular dysfunction. For this reason, despite massive tissue oedema, pulmonary oedema is not typically part of the clinical picture. Mobilization is more effective and safer than diuretics.
4. Cellulitis: Abnormalities in white blood cell count, fever, pain or measurements of inflammatory markers can be delayed or minimal even with serious infections Oral antibiotics, sometimes in combination with an antifungal agent (such as fluconazole) are usually effective.

BARIATRIC (gastric bypass) SURGERY IN PWS

by LINDA M. GOURASH, MD

My husband William Gourash, CRNP forwarded to me a question about doing bariatric (gastric bypass) surgery in Prader-Willi syndrome because I am a physician with extensive clinical experience with PWS and a member of the clinical advisory board for the PWSA-USA (the National Association for Prader-Willi Syndrome.) I am a Developmental Pediatrician.

Among those physicians and others who are familiar with the syndrome I know of no one who recommends surgery. The following are my own thoughts on the matter and do not represent any formal consensus opinion:

1. PWS is one of the most complicated behavioral syndromes you will ever encounter, if not the most complex. Persons with PWS are often very noncompliant individuals; they have little insight and experience shows that they can not be trusted to follow through on any declarations of cooperation.
2. We do not know the mechanism of the satiety defect in PWS nor do we fully know the mechanism for the efficacy of obesity surgery. It would be a stab in the dark to assume that the PWS patient will benefit.
3. Anecdotally, the surgery has had disastrous results in a number of patients but these have not, to my knowledge, been documented and published; perhaps surgeons have overlooked reporting these isolated events.
4. I (and others) have reviewed the published series of PWS who have undergone bariatric procedures. I was not convinced that the patients obtained any long term success with the surgery for the following reasons:

Missing from the reports was enough information about living circumstances, family satisfaction or whether patients were able to have a less supervised, less restrictive lifestyle.

Most of the authors appeared to be unaware of the nonsurgical management approaches to PWS now implemented in many parts of the world and so evaluation of whether these approaches were being implemented or were being withdrawn after surgery were not made.

The natural course of the condition was not taken into account, that is, that weight gain is worst in the late teens and early 20s while patients are "emancipating " from parental supervision. They will go on to die of their obesity unless someone realizes that environmental restrictions (locks, no access to food or money to buy food, psychological food security with behavioral incentives and a large volume, low calorie diet) are put into place (which is often what happens even without surgery.)

Persons with PWS have overeaten to the point of gastric ischemia (perforated stomach) and death. A restrictive procedure would have to be followed by strict environmental controls on food, thus advancing the quality of life not at all since these patients are already need to be managed by strict environmental controls.

Bypass procedures can be assumed to be accompanied by the consequences of dietary non compliance.

Persons with PWS are notoriously poor at reporting or registering pain and conditions that one would expect to produce severe pain including severe abdominal pain have presented with minimal or no complaints. Post operative risk would appear to be high for undetected complications.

Persons with PWS often fail to show much of an inflammatory response even in the face of significant infection; they may not show fever or leukocytosis (increase in number of white blood cells caused by infection or inflammation etc) in situations when these would be expected.

Persons with PWS are prone to osteopenia (reduction of bone mineral density) etc.

The ventilatory abnormalities of PWS are often not taken into account when obese persons with PWS are evaluated and managed. Respiratory failure from overuse of O2 is a common iatrogenic complication in the severely obese, hypoventilating chronically hypoxic patient. Any surgeon or anesthesiologist treating these patients should take the time to familiarize himself or herself with the syndrome.

More information on PWS is available at www.pwsausa.org/At Johns Hopkins University Dr. Ann Scheimann (pediatric gastroenterologis/IPWSO Scientific Advisory Board) is identifying and studying persons with PWS who have undergone any type of bariatric surgical procedure. She may be contacted at ascheima@jhmi.edu

LINDA M. GOURASH, MD

Understanding behaviour in PWS means we need to understand, *really* understand what is going on inside the person's head. When you think about it, this applies to anyone, not just a person with PWS, not just a person with a disability.

Behaviour is a Form of Communication

by LINDA THORNTON
National Director, PWSA (NZ)

Behaviour is a form of communication and to understand it, we have to understand exactly what the person is trying to communicate. What are they feeling, seeing, hearing, reading? Together we (you and I) can look at exactly the same picture, yet see different things. We can hear the same music, yet feel different emotions, we can listen to a person talk, and yet draw different conclusions in spite of the fact we are being told exactly the same thing.

Why would this be anything different for a person with PWS?

Our reactions to what we see, hear, feel, or read, are also quite different. Just how we react can be seen in our behaviour. If we hate something, we say so, we do something about it, remove it, ignore it, or move away. If we love something, we demonstrate that in many different ways. From our reaction comes our behaviour. If we continue to hate – or love - something, our behaviour becomes more intense.

Why would this be anything different for a person with PWS?

In order to get on in the world, we tend to mix with, work with, socialise with, and marry people who think along the same wavelength as ourselves. We do this so that together we understand each other. We also do this because we have a choice in our decision-making. A lot of the time, and for a variety of reasons, the person with PWS does not have this choice. Not having a choice means their behaviour – their communication – is going to be different.

Understanding their communication is the key to understanding behaviour in PWS: we must learn to communicate on the same level. It's like learning a new language. We need to understand the person, their needs, and their feelings by listening, seeing, feeling and interpreting in a way that they know we both understand.

Sometimes behaviours erupt from out of nowhere; sometimes we can see it coming but can do nothing to prevent it. Sometimes behaviours seem random, sometimes they're premeditated so that you'd swear you've been 'had', or manipulated. Each time it happens, it is a form of communication that we need to figure out.

The most effective way to deal with challenging behaviours is to prevent them from happening in the first place. The most common identifiable thing that leads to challenging behaviour is anxiety. For any person with an intellectual disability, anxiety is just about the biggest threat to their well-being.

Behaviour is ...

What makes you anxious?

Some of the things that make me anxious will probably be the same things that make you anxious:

• Not being able to make myself understood
• Not being able to understand another person
• Having to change my plans suddenly
• Someone not being on time to meet me (and vice versa)
• Being tired, depressed, angry
• Missing out on a meal

These are exactly the same things that make a person with PWS anxious. The difference is, I can communicate my anxiety; I can control my anxiety.
We need to be able to help the person with PWS control their anxiety so that they can communicate this and at the same time, feel safe.

What can we do to help:

• Not being able to make themselves understood clearly
Clues:
- Shouting/screaming ("You're not listening to me! Go AWAY! I don't like you")
- Insults, swearing

Help by sitting down with the person and talking quietly, asking questions like "I'm sorry – I just didn't understand what you wanted. Can you tell me again and I'll listen really carefully?" "How can we make this work together?"

• Not being able to understand you
Clues:
- Repetitious language
- Stubbornness and refusal to do something
- Insistence they know what they're doing
- Insistence that someone else has told them what to do

Help by sitting down with the person and taking time to explain something clearly, but put in lots of good detail so that they feel part of the decision-making. A longer answer (to them) is so much better than just a short one. They love to know detail because this helps them understand their own world better.

- Having boundaries shifted or new rules made

Tell the person well in advance if the rules or boundaries are going to be changed and explain why this has happened. Tell them the reason behind the change. Explain your reasoning and include them in the decision-making process.
If other people are involved (caregivers, teachers, parents), make sure they also know the process/rule/decision.

Anxiety around food:

People with PWS have anxieties which are increased around food. You and I have a choice around food: they do not.

• Not being able to have the meal ready on time
Clues:
- Upsurge in anxiety, repetitious questioning, frustration, anger

Help by explaining why the meal is going to be late, maybe involve the person in preparation of meal - supervised - so that they can be a helpful part of the process; give them something important to do like setting the table.

Not having "enough" food

"He's got more than me!" Comparing food with others and insisting that they have more than them, can make a person feel frustrated, cheated, angry. It may be necessary to go back to the menu, to weigh and measure out the food in front of the person so that they do not think it has been "thieved" before it has reached them. Portions are very important to a person with PWS. You can learn to bulk out a plate of food with many more low-calorie foods rather than a small portion of meat, pasta, or other high-calorie foods. More is always better to a person with PWS.

Anxiety can form many different behaviours. Here are some other common ones in PWS:
• Non-compliance
• Frustration
• Argumentativeness
• Accentuated compulsive behaviours
• Denial
• Perseveration (repetitive questioning)
• A drop in the level of communication or concentration
• Muddledness

If we can intervene at the point when anxiety presents itself in a behaviour we know will escalate; if we can re-direct the person, if we can help relieve the anxiety by identifying what it is, then we have a very good chance of stopping the escalation in behavioural outbursts. Remember: you can't win an argument, so don't buy into one. If the person starts to argue, leave the subject well alone, even if it means telling them you are going to leave the room, but you will come back shortly.

Behaviour is ...

Rewarding good behaviour

By constantly praising good behaviour when we see it happening, we increase a person's sense of self-worth, happiness, and contentment. The more we praise, then the more the person wants to do well for us. What's fair to expect – what's set to fail – what needs to be rewarded? When working with a person who is unable to control anxiety, it is absolutely vital that you ask yourself the following:

What is fair to expect?

- remember that you are judging 'fairness' from their perspective, not yours
- remember you are not there to 'win' the argument, you are there to solve the problem
- trying to understand the situation from the other person's perspective can be difficult and can seem as though your authority is being undermined. It isn't. You are skilfully solving a problem that might have otherwise erupted into a huge blow-out.

What is set to fail?

Failure in this case is looking at the other person's capabilities, not yours eg. Taking responsibility to change a person's routine for their own good, is not necessarily going to seem like that to them. Taking time to explain your reasoning and involving them in the decision-making process means you have a 'win-win' situation. Give and take, compromise in order to reach a solution that suits you both. Reminding the person of their extreme behaviour during a past outburst is not going to stop it from happening again. Remember that they are not able to control their behaviour when it has escalated.

What needs to be rewarded?

This can be the simplest thing - catch the person doing something 'good', or doing the right thing - rewards can also be simple, a 'thank you!' a hug, or a nod & a wink to show your approval can work wonders.

When the person comes back to you and apologises. This is a huge hint that the topic now be dropped for good! Thank them.

Are rewards needed?

Yes. Some sort of recognition for a job well done, or for having got through a good day - just a simple acknowledgement, a few words will show the person they have succeeded.

What rewards work? (do not use food as a reinforcer)

Praise, acknowledgement, mentioning it to others.

For greater rewards, make sure the reward is obtainable and reasonable (not *"You'll be able to get a new wardrobe if you lose weight"*, but *"once you reach x kg we'll go and look for a new t-shirt"*. Identify your reward carefully, or you may find you'll be up for more than you bargained for!

Working with a person who has challenging behaviours such as Prader-Willi syndrome, can be immensely frustrating, disempowering, and often hurtful. It may often seem that you are 'giving in' and allowing the person to take control. Managed skilfully, you will always win if you do it together.

Please Listen to This Shout Straight From the Heart

EIKO SHOJI, Japan. Mother of a person with PWS

Stop nagging me!?
Why must you always be angry with me?
What do you want from me?
I can scream back at you too
You're making me want to break out

You always yell at me!
My mind goes blank and then I want to run away,
So I end up laughing it all away
Why can't I be understood?
I know what I'm doing, but I can't stop myself.

I'm always misunderstood!
I can talk back, I can even argue
But I'll be yelled at again,
Since I don't know enough to really talk
Please, I need a clear explanation

Can't you see the pain in my heart!
I can't tell what's happening around me,
I'm glared at and scolded day by day,
Leaving behind scars and awful memories
My heart is crying for help

I do love people though!
The ones that try to understand me
The ones who listen to me
And the ones who look gently at me and smile warmly
With them by me, I can be at peace

But, what I really, really, want,
Is to be watched and looked after
Is to be free of this worry and fear,
That's why, I'm begging you to keep your eyes on me
Please promise that you'll watch out for me.