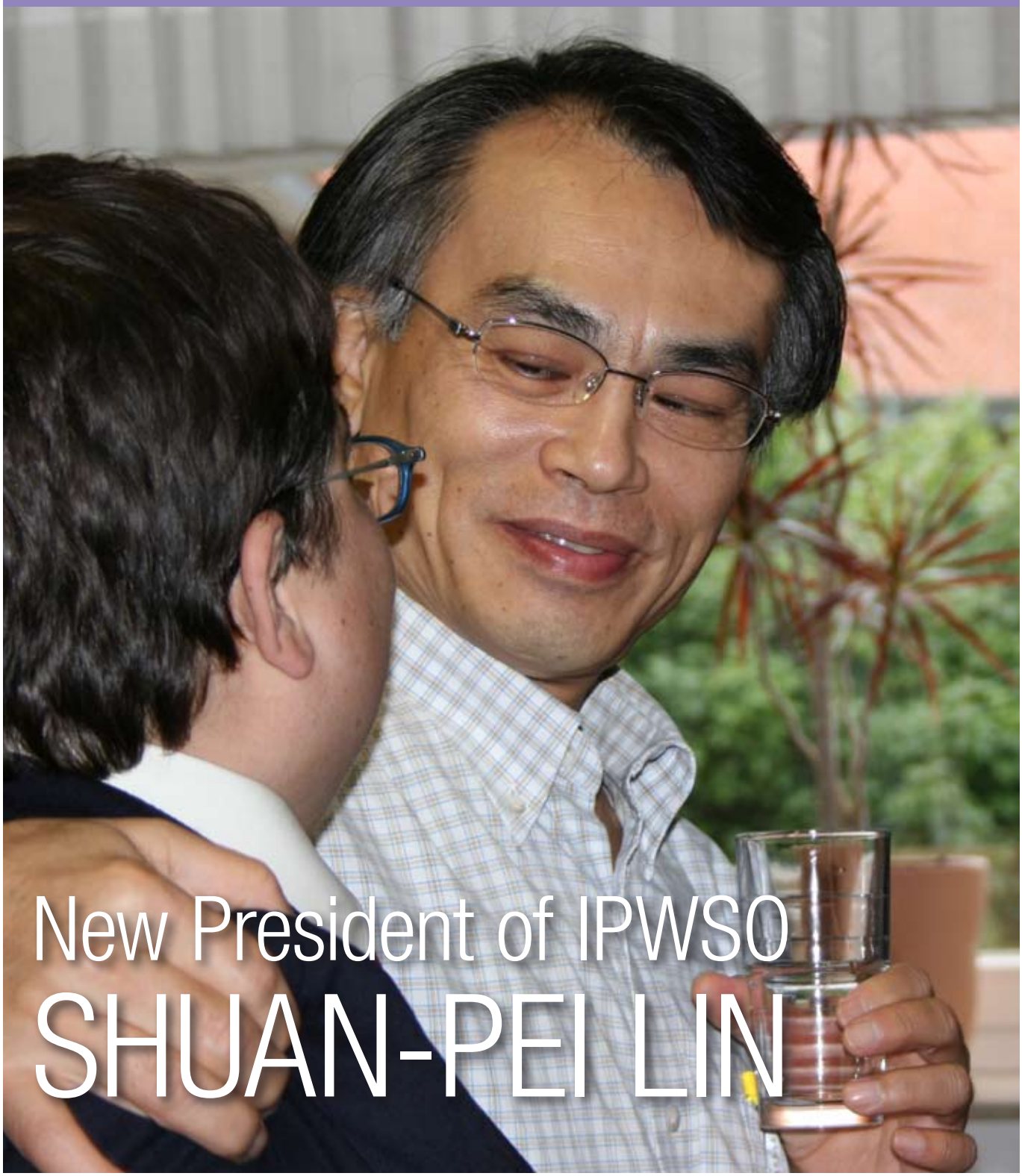
Vol.2 September 2009

WAVELENGTH



New President of IPWSO

SHUAN-PEI LIN

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 (and within the parameters of the syndrome), 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.

From the Editor:

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

The Second International Caregivers' Conference was held in July, in Herne, Germany. This second conference followed the same concept as the first: a series of lectures and workshops where professional caregivers from as many as 14 countries throughout the world, gathered to talk, discuss, share, and finally to hammer out the best possible guidelines for 'best practice' in the care of those with PWS. You will find some reports of this inside this edition of Wavelength.

At the same time your Board held full and comprehensive meetings over a period of three days, examining in detail our current situation and looking forward to our future. There is a short report on p. 4 outlining the decisions taken by the board. We also discussed the forthcoming 7th International IPWSO Conference, 22-24 May, next year in Taipei. There was a great deal of enthusiasm for the conference and programmes are rapidly coming together. It is always a time of great excitement, determination, and pressure to make sure that the conference will be of benefit to scientists, researchers, professionals, and not least, to parents and caregivers. Our international conferences have always set a high standard and have a wonderful feeling of partnership and camaraderie with the mix of professionals and parents under one roof.

Even in this time of global recession, swine flu, and extreme weather patterns, we have a job to do that just doesn't change – the care of, education of, and responsibility for all our children and adults with PWS. Come what may, this need to provide the best possible care will never change; research will always continue, our children will always need us. Let's continue to share our knowledge, improve our welfare, and provide the best possible support we can, from country to country, town to town, person to person.

LINDA THORNTON
National Director, PWSA (NZ)

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Cover picture:
Shuan-Pei Lin togethr with Daniele Fornasier

PRESIDENT'S MESSAGE

I bring you greetings from Taipei, Taiwan, the home of the next international PWS Conference. You will find more information on this in the newsletter, but I want you to know how delighted we are to be hosting this conference, and I know you will be excited by the programme we are developing together.

At our board meetings in Bad Oeynhausen, Germany, we met for the first time since Pam's death and it was heartening to know that the progress we are making in IPWSO would be as she would have wished. The second International Caregivers' Conference in Herne, was a tribute to Pam's ability to recognise the importance of such a gathering when, back in 2006, she, Norbert Stuntebeck and Hubert Soyer and others who were at the Cluj IPWSO conference, discussed what could be provided for all caregivers in the world caring for adults with PWS to give them the best possible lifestyle they could manage. This was the birth of the first Caregivers Conference which Pam attended last year. This year, we had a special Pam Eisen Lecture, given by Professor Tony Holland, which looked at the management of PWS over the years and concluded with a wonderfully positive attitude for the future. In Taipei, we will be having our third Caregivers' Conference with a specially designed programme building on the knowledge for best practice guidelines already started at the last conferences.

We are looking forward very much to seeing you, and greeting you at the seventh international PWS Conference in Taipei!

Warm regards to you all,

Shuan-Pei Lin
President and Chairman

News from the IPWSO Board

The board had a series of meetings recently both at the 2nd Caregivers' Conference in Herne, Germany, and afterwards in Bad Oeynhausen where we were wonderfully hosted by Norbert Stüntebeck and his staff at Wittekindshof and had the opportunity of visiting the residential home of 8 people with PWS and being shown around one of the busy workshops, and later hosted at dinner where the Mayor of Bad Oeynhausen was the guest of honour. The warm hospitality and eagerness to show us both the working and living environments was most appreciated by us all, and made a welcome break in our two days of meetings.

As this was the first meeting since the death of our President, Pam, the board invited our auditors, our legal adviser, and the consultants who had been at the Caregivers' Conference. We felt it was important to share our combined wisdom, opinions, and gain good advice for our future.

It was an excellent decision and the enthusiasm and good-will this generated gave us a huge boost. The board discussed many ways to give IPWSO a professional profile, more input into, and support of, future international conferences, and better ways to share and disseminate information among countries

IPWSO Board had the opportunity of visiting the residential home of 8 people with PWS in Wittekindshof and Bad Oeynhausen.

Linda Thornton from IPWSO Board shaking hands with one of the residents. In the picture also Norbert Hödebeck-Stüntebeck from Wittekindshof and Janalee Heinemann also from IPWSO Board.

David Gordon

Board recommendations:

and associations. We took a long, hard look at our responsibilities and roles, we restructured and increased our board size with two co-opted members (Jackie Waters, UK, and Janalee Heinemann, USA), created new sub-committees and task groups to strengthen our scientific advisory board and look at creating new advisory boards. Other recommendations and changes are listed below:

Board restructure (and responsibilities):

Shuan-Pei Lin
President and Chairman
Linda Thornton
Vice President and Secretary:
Jackie Waters
Assistant Secretary
Jan Haanaes
Honorary Treasurer
Dorica Dan
Assistant Treasurer
Tiina Silvast
Graphic Designer
Janalee Heinemann
IPWSO conference coordinator and Medical Information Coordinator

Executive Director:
Giorgio Fornasier
Appointed auditors:
Beppe Quaglia,
Verena Gutmann
Legal adviser:

Board Members:

The board recommended that the board should comprise at least 8, but not more than 10 members, so we will be looking for nominations at our 2010 General Assembly.

IPWSO internet site:

The board recommended an increase its website capacity to enable more information to be downloaded. IPWSO's website is currently hosted by one of our parent members, Steve Lundh, whose effort over the years has been monumental and is gratefully acknowledged.

IPWSO Conferences :

The board recommended that for future conferences (2013 conference and thereafter) IPWSO takes ownership of the conference in such a way that it will co-host with the successful tender country and have direct input into organising committees and programmes; and will also help financially.
A new tender document is currently being drafted.

PWS Country/Regional Conferences:

Where countries are holding PWS conferences of their own, or in their own region, IPWSO will, if invited, help fundraise for these events and, in turn, will have its logo prominently displayed and have the opportunity for its own stall promoting the work of IPWSO.

Annual Report:

IPWSO will produce an Annual Report with audited accounts at the end of each financial year (31 December) and this will be posted on the IPWSO website

Registration:

The board has recommended that, in the future, IPWSO will be registered as a charity in countries only where major fundraising or financial transactions are undertaken.

Parent and Scientific Delegates:

The board has recommended that the number of parent delegates from each member or associate member country can be up to two. Scientific delegate representation would remain at one per country. There is no change to the voting rights; this remains at one vote per constitutionally registered country.

Country Membership of IPWSO:

The board recommended that IPWSO work directly with country Associations as well as parent delegates. Medical and scientific information that has, in the past, been sent only to parent and scientific delegates, will also be sent to Associations for reprinting in their newsletters, or websites. Invoices for membership will also be sent directly to Associations. Although IPWSO has no restrictions in offering information to anyone supporting a person with PWS, to become a subscribed member of IPWSO requires that a country has its own constituted PWS Association. This also entitles that country to vote and take part in the General Assembly.

2010 Conference "East Meets

Second Caregiver's Conference in Herne, Germany

During her term as President of IPWSO, Pam Eisen was committed to the idea that the caregivers of persons with PWS should have a "home;" a conference of their own where they could share ideas and receive support for the challenges that they face. The First Caregiver's Conference in Herne in 2008 was so successful that Pam decided that the next caregiver's meeting could not wait until 2010 in Taiwan. So she proposed to have the Second Caregiver's Conference in 2009. Pam's death in November of 2008 only strengthened this resolve. With much work and good fortune, it became possible to have the conference at Akademie Mont Cenis in Herne on July 6-8, 2009.

The purpose of this conference was to continue the task of developing guidelines and standards (best practices). This conference was designed to be smaller and more focused in its work. There were two work groups devoted to gaining broader consensus for "Environmental Structure of Living" and "Behavior Management." There were two new work groups focused on "Interpersonal Relationships" and "Self Determination."

Caregivers were welcomed warmly to Herne by Mayor Mrs. Erika Wagner, Linda Thornton from IPWSO, our on-site host from Diakonische Stiftung Wittekindshof Irene Stenzig, Hubert Soyer from Regens Wagner Absberg and Volker Holzkämper from PWSA Germany. There were general educational lectures for everyone on the topics of Self Determination and PWS (Leopold Curfs), Behavior Management (Janice Forster), Environmental Structure of Living (Hubert Soyer), and General Health in PWS (Suzanne Blichfeldt). There were also two new lectures that in the future will become part of every Caregiver's conference: The Pam Eisen Memorial Lecture and the International Lecture.

Janalee Heinemann gave an emotional tribute to her long time friend, Pam Eisen. She presented pictures of Pam's life

There were two work groups devoted to gaining broader consensus for "Environmental Structure of Living" and "Behavior Management." There were two new work groups focused on "Interpersonal Relationships" and "Self Determination."

and travels for IPWSO. Through these pictures and her words, as only a good friend could say, she brought Pam close to all of us in our hearts and memories. Then, Giorgio Fornasier, past president of IPWSO and current CEO, presented the pictures and the history behind the IPWSO song, Ich Auch (ME TOO) that he and Pam wrote together. He led the group in song, involving all of the participants in this special moment created by standing hand in hand together, feeling like one big PWS family. Then Janalee reviewed the distinguished career of the person selected unanimously by the IPWSO Board to give the memorial lecture.

The first International PWS Lecture was presented by Professor Jeyachandran from India.

The first Pam Eisen Memorial Lecture was given by Tony Holland. This outstanding lecture, dedicated to Pam and her daughter Gabriella, contrasted the amount of knowledge that was available at the time that Pam's daughter Gabriella was born with the information that is known for parents today. Dr Holland traced the development of our knowledge about PWS from 1956 to the present, identifying the most important research findings and how they have affected clinical care. (This lecture has been transcribed and will be published in a separate format.)

The first International PWS Lecture was presented by Professor Jeyachandran from India. His lecture reviewed the history of India over the past 5000 years, integrating the development of medical care and the current health system of India today. There are over one billion people living in India. Over the next ten years, we will see considerable progress in the management of PWS, as well as the services available for persons with PWS throughout India.

IPWSO Board and Consultats. In the back: Jan Haanaes Norway (left), Linda Thornton New Zealand, Susanne Blichfeldt Denmark, Tony Holland UK, Giorgio Fornasier Italy, Norbert Hödebeck-Stuntebeck Germany. In the front: Janice Forster USA (left), Jackie Waters UK, Tiina Silvast Finland, Hubert Soyer Germany and Leopold Curfs Netherlands.

Near the end of the conference, there were presentations for everyone summarizing the best practice statements from each of the four work groups. The response to these guidelines was offered from the perspective of the caregiver (Renate Staufenberg from Regens Wagner), the provider (Professor Stamitzke, director of Wittekindshof), and the parent (Linda Thornton).

The 2009 Herne conference was smaller (80 attendees) but more intense; each work group was given twice to increase the number of participants and to build consensus. There were more countries represented this year (17). Attendees were caregivers, providers,

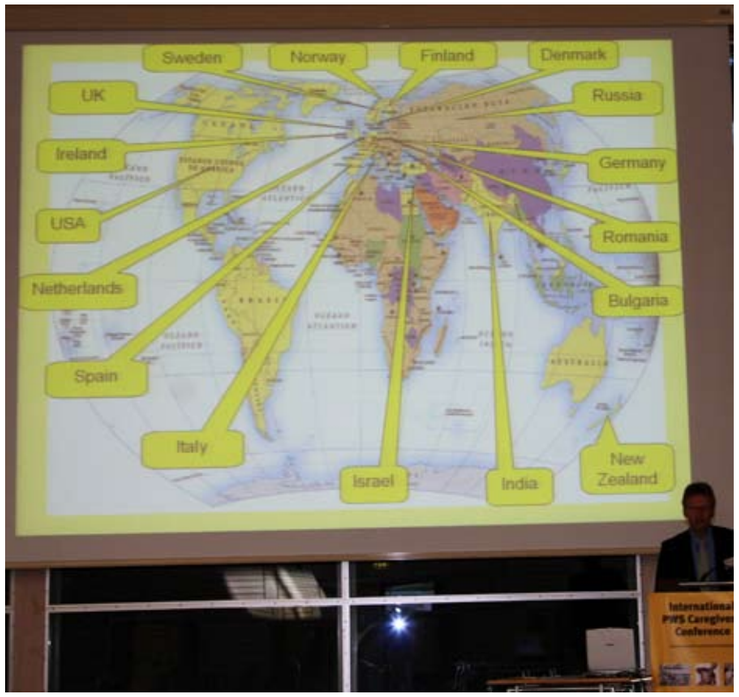
The first Pam Eisen Memorial Lecture was held by Dr Tony Holland from UK. Dr Holland traced the development of our knowledge about PWS from 1956 to the present

There were more countries represented this year (17). Attendees were caregivers, providers, psychologists, psychiatrists, physicians, teachers, and physiotherapists.

psychologists, psychiatrists, physicians, teachers, and physiotherapists. This year at the Akademie Mont Cenis, there was the same opportunity for attendees to build collegial relationships outside the work group experience, enjoying good food and excellent beverages! The group attended the barbecue again this year, and the rain could not dampen our spirits! The cultural venue was a trip to a coal mine museum that offered not only local history but an archeological adventure!

The 2009 Herne experience was most memorable, characterized by hard work and good times. It provided an unequaled opportunity to develop guidelines that will have an impact on the world of PWS for many years to come. This was the first conference without our dear Pam, but we know that she was with us in spirit, and we know that she is smiling! The Caregiver's Conference is her wonderful legacy. We look forward to the Caregiver's Day at the Seventh IPWSO conference next year in Taiwan when many of the best practice guidelines will be presented.

Respectfully submitted,
Conference organizers: Norbert, Hubert, Janice and Leopold, with special thanks to Linda Thornton, IPWSO



PWS Association in India

The Indian Prader-Willi syndrome association was initiated with the dream of late Pam Eisen in the year 2004 during the annual PWSA meet at United States. After a long wait and lot of struggling it was founded and formed in India by two parents. Finally the first Medical conference was successfully held in Mumbai, in January 2008.This was attended by about eight PWS families from across the country and Giorgio Fornasier of IPWSO who has been our pillar ever since Pam left us.

Our Association has also participated in two prestigious medical conferences held in 2008 in Mumbai and 2009 in Bangalore. The stalls were attended by around 2000 paediatricians who were briefed about the syndrome as well as our association. We have successfully diagnosed several PWS cases by conducting tests in Italy and supervised by Dr Jalan's clinic and by Dr P. Raghpathy. Now our association has registered about 15 PWS cases and we provided information and help to individual families as need be. Hopefully we are even more successful at our next PWS meeting to be held early next year - something we are looking forward to greatly.

Best regards
Shikha

President of PWSA India Mrs Shikha Harlalka (left), Dr. Susan Casside USA, member and founder of PWSA India Mr. Rohit Kumar and Giorgio Fornasier IPWSO.

Sultan from Kazakistan

Sultan has a very kind heart, he is compassionate to people and loves animals. He likes to read, write, paint, and knows how to use the computer.

"The health status of the Sultan (my nephew) who has Prader-Willi syndrome is serious. He weighs 156 kg, and is aged only 16 years. During the day he stays in the apartment, sometimes walking weakly, sometimes sleepy, eats supervised, and when he's awake, he calls friends on the phone. At night while asleep, he finds it difficult to breathe, snoring, often waking, as though he is suffocating. His blood pressure is 100/40 and 130-160/40 and suffers tachycardia, and poor joints, foot cramps, poor eyesight, etc.

Emergency physicians and medical institutions do not help, do not know the disease, have rejected it. Kazakhstan did not know this disease and has no recorded cases of this syndrome, we are the only ones. We only got the diagnosis by accident when a visiting American pediatrician, Kazakhstan happened to see my nephew, Sultan, when he was 6 years old.

Doctors are indifferent, even to the general ill people, not to mention any disability. We have show and tell the doctors a description of Prader-Willi syndrome, and explain to them the health of the Sultan, but the doctors in Kazakhstan cannot help us and do not want to, and do not want to know. They look to him as a monster.

Besides, we live in the city of Ust-Kamenogorsk, where almost every day the tubes emit harmful chemicals from steel producers. We do not have a satisfactory Ecology here.

Sultan basically is looked after and cared for by his grandmother. She is 59 years old. She tries to make him comply with the diet, but with so little food, he gets very hungry and cries, becomes very anxious and tries to secretly find any food in the kitchen. The doors in the kitchen, we are closing with locks.

In our Sultan is a very kind heart, he is compassionate to people and loves animals. He likes to read, write, paint, and knows how to use the computer. He loves watching TV and can even work out some things. But does not know the time (hour, midday, morning, evening), does not know how to count money, cannot write down basic sums without a calculator.

For the provision of assistance during sleep in obstructive apnoea, Kazakhstan has no breathing apparatus; they say that the device doesn't exist. Oxygen cannot help, and there is no medication according to the physician pulmonologist. Oxygen is only used in intensive care in hospitals. Pulmonologist advised Sultan to give tablets Teófilo overnight.

We do not know what to do. But the health of the Sultan is extremely difficult.

We wish you dear IPWSA staff to communicate and write.

Thank you for helping to Giorgio Fornasier, Linda M. Gourash, Janalee Heinemann."

[Along with our consultants, we are offering support, information, and assistance to Sultan and his family. It is cases like this that desperately need the help of our combined knowledge to support Sultan, his family, and the medical advisers in Kazakistan.]

Askar Sadykov, uncle of Sultan

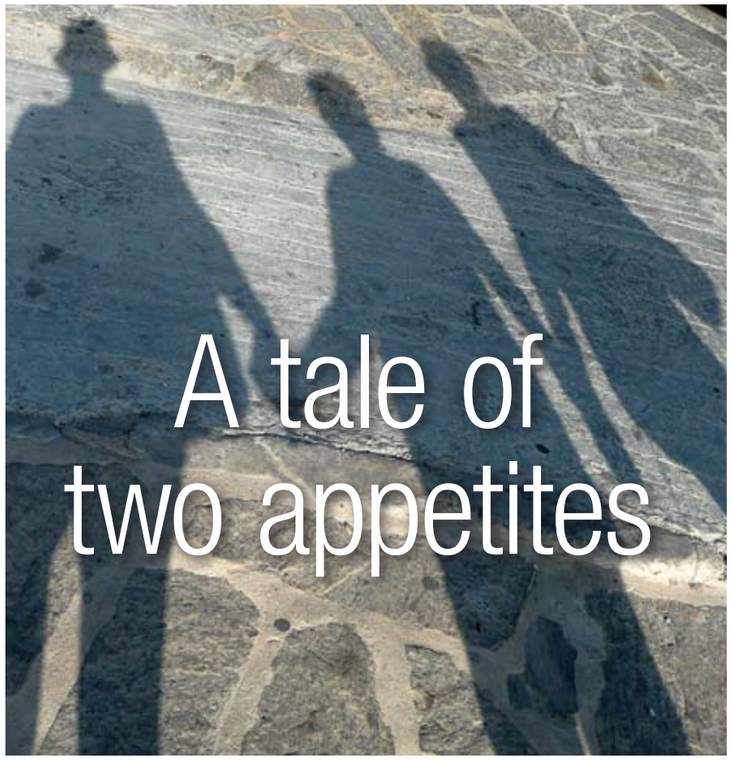
PWS Association in Slovakia

The Slovak Prader-Willi Syndrome Association (SPWSA) was found in September 2006 thanks to support of the pharmaceutical company Pfizer. Our main objective is to bring together families with children who have PWS, organize meetings, share knowledge and information and thus make life easier for both the families and children. We publish information on our web site, print information leaflets and cooperate with other national PWS organizations, especially Czech national association with which we also plan some activities in the future. In October 2006 SPWSA became a regular member of the international organization IPWSO.

The Slovak Prader-Willi Syndrome Association currently has 19 member families. These are all families who have a child affected with PWS. The youngest child is 8 months old and the oldest one is 20. Most of the younger children receive the growth hormone treatment, which in Slovakia is indicated and covered by the insurance company. However, the older children and adolescents receive no treatment. Therefore, it is our focus to spread the information about the syndrome, possibilities of its early diagnosis and treatment around the pediatricians, pedagogical staff, occupational therapists, other professional staff and also general population.

We plan to organize meetings of the member families on a regular basis (twice a year), with the participation of professionals who will present to us the latest techniques and news in PWS management.

In the long - term perspective SPWSA plans to establish a group home or sheltered workshop for the children with PWS and other disabled children, where they could live more independent life under the supervision of skilled professional staff.



A tale of two appetites

By Jackie Waters (UK)

When I was a young child I was what is now known as a "difficult feeder". I barely ate anything and became full very quickly. As a teenager I ate chocolate and sweets in an effort to become fatter - but large meals were just impossible - I dreaded going to restaurants because of the embarrassment of not being able to make more than slight inroads into a meal. I wasn't anorexic - I truly wanted to get fatter, but physically I couldn't do it.

So when I had my daughter with PWS I was delighted to find that she would eat anything I put in front of her - she wasn't diagnosed till she was 5. However, my own perceptions of portion size meant that I didn't massively overfeed her anyway. When the doctor diagnosed her, she said to me, "No more biscuits and no more cakes." And as a child she stuck to that. Her weight was always good and she never sneaked food.

When she reached 16, she had more independence, starting helping with her meals etc. Now 31, she has total independence around meals and resents any interference on any level. Her weight is on a fairly steady upward curve - the more she eats, the more she wants. We seem to be on a runaway train that has no brakes - she refuses to go into residential care.

And so it is with me. After the age of 45 I gradually began to eat more ... and more ... until now I am approaching the mild obesity level. Trying to cut down and resist food is virtually impossible! My mother and grandmother both followed the same path - very thin as younger women, followed by a descent into obesity in late middle age.

I guess the moral of this story, if there is one, is that appetite feedback mechanisms are a very delicate balance and can change over time. And if our physical controls stop working, then we have to introduce self-regulation and/or environmental controls to stop it. How much more so is this the case with PWS!

Regulation of Weight in Prader-Willi Syndrome

Theoretical and Practical Considerations

Pittsburgh Partnership, Specialists in Prader-Willi Syndrome; www.pittsburghpartnership.com
Linda M. Gourash, MD, PWSA-USA Clinical Advisory Board, PWSA-USA Board of Directors
Janice L. Forster, MD, PWSA-USA Clinical Advisory Board, IPWSO Scientific Advisory Board

Edited by Jackie Waters, PWSA (UK) Services Director, IPWSO Board member

Introduction

These questions have been raised by nearly everyone who comes in contact with a person with Prader-Willi syndrome:

• What is the defect in weight regulation that sets PWS apart from typical obesity? • How is the problem to be managed? • Does every person with the syndrome require the same approach?

The answers are complex and require more than short term observations of persons with PWS. Observers who have had long term experience with the syndrome have come to understand the following:

• The defect is severe and life threatening;
• Short term behavior with respect to food does not predict long term weight regulation capability;
• Behavior with respect to food is highly dependent on an individual's past experience and current opportunity.

Theoretical Considerations

We do not yet know the precise defect(s) leading to weight dysregulation in PWS. However, by examining how weight is regulated in typical persons, we may better appreciate the situation with PWS.

There are 3 categories of factors that potentially control body weight over time:

Self-regulation
Environment
Physiology

The least potent of these categories is self-regulation (the voluntary calorie restriction and deliberate calorie expenditure); it has a minimal role in control of body weight (Peters et al. 2002).

The current worldwide epidemic of overweight and obesity is generally attributed to a change in the environment (Hill et al. 2003; WHO 2000). Never before in human history has food been so inexpensive, convenient and tasty. Statistics reveal that most persons (greater than two thirds of us) gain excessive weight over time when environmental constraints on the food supply are no longer operative, and physical activity is optional rather than necessary for our survival. Research on general obesity is painting a dismal picture about our ability to learn to regulate our body weight under these environmental conditions, battling an inherited, physiologic tendency to conserve energy and to gain weight.

PWS and the Rest of Us: Orders of Magnitude

What separates the slow steady weight gain of the typical overweight person from the rapid weight gain possible in PWS is physiology. It is easy to underestimate the physiologic defect in PWS with respect to body weight regulation. Typical persons who are morbidly obese are thought to be taking in less than 100 excess calories (kcal.) per day or less than 2-5% excess over their daily energy usage. The net effect is an annual weight gain of well under 10 pounds per year. This appears to be what typically occurs in susceptible persons.

Other persons, those of fortunate genetic makeup maintain a perfect daily energy balance and a healthy weight year after year. This weight regulation is a not a voluntary process. A number of complex neurohumoral (neural and hormonal) signaling systems are involved in slowing or preventing weight gain. These include messages to the brain that make further consumption of calories unappealing, nauseating or even painful. These signals have been shown to be delayed, weak, and short lived in persons with PWS allowing them to consume comfortably 6 times their actual calorie needs and, in documented cases, far more.

Further, typical overweight persons have two mechanisms available to slow their weight gain: they expend more calories than lean individuals due to an increased body mass and muscle mass; and their metabolism becomes less efficient, thus wasting calories (Leibel et al. 1995). Neither mechanism is available to persons with PWS. Their muscle mass is smaller than normal, and their hypothalamus is less responsive and likely does not recognize and adjust their metabolism when they are gaining weight.

Due to their smaller muscle mass, persons with PWS are much more limited in their ability to increase their calorie

expenditure through exercise; they have no hope of achieving an energy balance by that means alone. Even with exercise, they will gain weight eating a normal diet.

Some clinical observations of responses of persons with PWS to food and fasting have been intriguing. Persons with PWS appear to enjoy food more. Further, they may find foods or even inedible items that are unpleasant for others to be enjoyable. Some clinicians have interpreted this willingness to eat distasteful items as evidence of their "ravenous hunger". But there is a different interpretation. Rather than assuming extreme hunger at the beginning of a meal, there is reason to believe that the defect involves the failure to respond normally at the end of a meal. The defect appears to be in the sense of satisfaction or fullness (called satiety) that normally comes with eating. Both behavioral observation studies and more recent brain imaging studies have demonstrated abnormalities in the response of persons with PWS following the ingestion of food.

The brain activity of typical and PWS persons has been localized and quantified while they viewed food pictures during the fasting state and following a meal. Holsen et al. (2006) found that study subjects with PWS not only responded to food stimuli more than healthy weight controls, but also the magnitude of response was greater after a meal than before the meal.

There are paradoxical clinical observations as well. Families have reported that their child with PWS can be relatively indifferent to fasting as long as they are assured of the arrival of the next opportunity to eat. Prolonged fasting as a result of a medical condition has also been noted to result in less, not more, interest in food. And, of course, as every parent knows, young infants with PWS universally appear to be indifferent to hunger and to eating.

Discussion

Neural and humoral (nerve and hormone) signals control both short term energy intake/use as well as long term body weight regulation; they provide tonic continuous feedback to prevent or slow weight gain. Persons with PWS appear to have abnormalities in both short term and long term regulatory functions. An imbalance of short term energy intake allows excessive calorie intake meal by meal, day by day. Leptin, a messenger produced by fat tissue, is present in normal levels in PWS, but it fails to deliver the message that for the rest of us stabilizes weight over time. The brain's apparent insensitivity to this messenger even at high levels allows persons with PWS to rapidly accumulate a large amount of adipose tissue (fat). Ghrelin, the "hunger hormone", is often elevated in persons with PWS but there is reason to believe that the brain is relatively insensitive to this messenger, as well. It is not at all clear that ghrelin is driving the excessive intake. The difference between persons with PWS and other weight gaining persons may only be a matter of degree, but the degree of abnormality is extreme.

Direct clinical and research observations, together with reports from caretakers and families, suggest that both hunger and satiety signals are blunted or diminished in PWS (along with other types of feedback such as feelings of disgust and pain). The neuro-imaging studies, while very preliminary, appear to be pointing to a similar conclusion. What drives the excessive food intake may not be hunger so much as the reward of eating. This enhanced reward associated with eating may be the result of the delayed and diminished satiety signals that would normally balance the reward of eating.

It's lunch time at PWS summer camp in Finland in 2006.

Food is good and everybody is happy.

Practical Considerations

Self-regulation is not new to PWS. Persons with PWS must regulate their impulses all the time. They self-regulate when they do not grab food off your plate. They self-regulate when they accept a disappointment without a meltdown. They self-regulate when they know they are being watched. They self-regulate when they put in their daily exercise. Self-regulation is enhanced with a number strategies used by parents and caretakers over the years. Cognitive techniques have been used with persons with PWS to help them express their feelings, agree to behavioral contracts, accept their differences and work within their limitations.

But none of this comes easily. Persons with PWS do not have good judgment. They have difficulty identifying realistic long-term goals and keeping those goals in mind while they pursue the short-term steps to attain those goals. They function best when caregivers assist those executive functions by breaking down goals into manageable steps, supplying emotional support, and providing immediate rewards and consequences based upon daily behavior.

All of this comes at a cost. Self-regulation requires effort from the person with PWS and a long term commitment from those around him/her to provide the consistent supports needed for optimal function. The level of support required for optimal function can never be withdrawn. Optimal function will deteriorate with diminished support. Maximal function (including an individual's involvement in his or her own life decisions) requires a greater, not lesser, investment of time and effort from everyone involved in the person's care.

Resources available in both the short term and the long term must be taken into account. For this reason, self-regulation and independent function are not synonymous.

For persons with PWS, uncertainty and opportunity related to food consumption are a constant source of stress. Thus, persons with PWS display anxiety and other stress symptoms when they are responsible for their own food regulation. Conversely, stress and related behavior problems can be managed by reducing uncertainty about food. This is the basic premise for the concept of FOOD SECURITY. This term refers to the psychological state of the individual, not the presence or absence of locked access to food in the environment.

Persons with PWS can be content provided their expectations and their actual life experience are concordant. A discrepancy between expectations for increased independence and the inability to sustain that independence without weight gain can be a chronic source of stress. Frequently, this stress precipitates behavioral crises.

Practical Applications

1. The Principles of FOOD SECURITY ("No doubt, no hope, no disappointment") can provide a working paradigm for evaluating a person's capacity for self-regulation. This approach requires caretakers to see food issues from the subjective point of view of the person with PWS. Food security requires different measures for different individuals. The strategy examines the degree to which the person's expectations about food are managed:

No Doubt: the person knows how much food (portion control), what kinds of food (advanced menu planning), and when (daily schedule) the food will be served.

No Hope: the person knows that there will be no opportunity to obtain additional food; this usually require the use of locks to secure food access, the use of supervision in food accessible areas or situations, and the training of caretakers on how to manage the person's opportunity to buy/steal/pilfer/trade or manipulate others to obtain food items.

No Disappointment: the person does not experience an emotional let-down due to false anticipation or unfulfilled expectations about food.

"Doubts" and "hopes" may be fact-based or not; it is their existence in the mind of the individual with PWS that matters. When these are minimized, stress is reduced, and behavior is nearly always noticeably improved.

2. Past behavior around food issues appears to be the best predictor of future behavior and provides clues to the measures that may be required top develop an individualized plan for full food security. For example, some persons may never go through garbage cans, break locks or steal, while others can be quite obstreperous in this regard. Persons who have been willing to break societal rules or go to greater lengths to obtain food may be less likely to tolerate situations in which there is a chance to access food.

3. Some degree of self-regulation in a PWS-dedicated facility is not predictive of successful adaption to a mixed residence. No other medical condition requires the long term, extremely low calorie intake required of persons with PWS. Some adults maintain their weight on as little as 600 calories per day even with an exercise program. Typical calorie needs are 1000-1200 calories per day. A mixed environment leads inevitably to exposure to the greater amounts and types of foods permitted to other residents who may be consuming twice as many calories per day as the person with PWS, even if they are themselves on a calorie restricted regimen. Mixed residences have been successful when food is inaccessible to all residents.

4. Theoretically, all persons with PWS who are exposed to unsupervised and unlimited food access are at risk for gorging, choking, gastric distention and necrosis leading to rupture, shock and death (Wharton et al. 1997; Stevenson et al. 2007 persons with a history of gorging or with a past history of episodes of abdominal distention should be considered at special risk. If self-regulation in the presence of food is to be attempted, the amount of extra food that is potentially accessible must be limited.

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East Meets West
A New World for Prader-Willi Syndrome

FIRST ANNOUNCEMENT

Seventh International Prader-Willi Syndrome Scientific Meeting

May 20 & 21, 2010

The Seventh International Prader Willi Syndrome Scientific Conference will be held on the above dates in Taipei, Taiwan under the auspices of the International Prader Willi Syndrome Organisation (IPWSO) and in collaboration with the Taiwan Rare Disease Foundation.

STEERING COMMITTEE: Chairs Professors Leopold Curfs (NL) and Suzanne Cassidy (USA). Committee members: Professors Shuan-Pei Lin (Taiwan), Anthony Holland (UK), Daniel Driscoll (USA), Tomoko Hasegawa (Japan), Duangrurdee Wattanasirichaigoon (Thailand), and Stewart Einfeld (Australia)

CONFERENCE AIMS: This Scientific Conference will continue the traditions of previous international PWS meetings by bring together clinicians and researchers from around the world to present and discuss new research findings relevant to 1) our understanding of PWS, 2) new treatments and support strategies, and 3) policy and practice development.

The Scientific Conference will take place over one and half days and will be held in close association with a conference for parents and professionals, thereby facilitating the exchange of ideas and the dissemination of research findings. Those attending the Scientific Conference are encouraged to stay for the further day of the parents and professionals conference, which starts on the afternoon of the second day of the Scientific Conference and continues for an additional day, the theme of this conference is East Meets West: A new world for Prader-Willi syndrome. The Scientific Conference will take the form of peer-reviewed submitted presentations introduced by key note presentations. Contributions from diverse academic perspectives are welcomed including medical, biological, behavioural and sociological studies and investigations using mouse models. The conference presentations address three broad themes:

Genetics and animal models
Clinical and behavioural issues
Brain studies

The organisation of this IPWSO Scientific Conference aims to foster inter-disciplinary discussion and the exchange of ideas, thereby aiding in the development of new research and collaborations. Abstract submissions on research into all aspects of PWS are solicited. All those attending are encouraged to stay for the full Conference Éast Meets West: A new world for Prader-Willi syndrome. Further details will follow.

Enquiries: Contact Leopold Curfs (HYPERLINK "mailto:Curfs@msm.nl" Curfs@msm.nl) or Suzanne Cassidy HYPERLINK "mailto:cv.sc@sbcglobal.net" \o "mailto:cv.sc@sbcglobal.net" cv.sc@sbcglobal.net

Instructions for Abstract Submission

Deadline for abstract submission is January 15, 2010

Your abstract should be typed on a single page with font size of 12, Times New Roman with 2.5 cm margins. The maximum size of the entire text should be 24 cm x 17 cm. Please follow the formatting shown in the sample abstract on the next page. The title should be at the top of the page in all capital letters and bold type and be centered on the page. Skip a line, then on the next line, in regular type, provide the names of the authors, with the name of the speaker underlined. Skip a line, then on the next line list the affiliations of all the authors, in the same order as they appear in the author list. Then skip a line and begin the abstract. You may use whatever other formatting is needed in the body of the abstract. If possible, please divide the abstract into labeled sections for Introduction, Methods, Results, and Conclusions. Please send an e-mail to curfs@msm.nl if you have problems with the form, of if you do not have access to Microsoft Word. Please save in a format readable by Word 2003 (i.e., .doc, not .docx). Use "save as" to save your abstract using your first and last name as the document title. You may submit multiple abstracts (please number them), but be prepared to present all that are accepted.

Please also send a cover email or letter with each abstract including the following information for the presenting author:

- Name
- Mailing address
- Telephone number
- Fax number
- E-mail address
- If you have a strong preference for slide talk or poster

DEADLINE FOR SUBMISSION OF ABSTRACT is January 15, 2010

Notification of acceptance will be provided by March 1, 2010. Depending upon the number of submissions, there will be both platform and poster presentations

Please email to:
Suzanne Cassidy: e-mail: cv.sc@sbcglobal.net

SAMPLE: IPWSO 2010 Scientific Conference Abstract Submission

STOMACH NECROSIS: A LETHAL COMPLICATION OCCURRING IN SLIM INDIVIDUALS WITH PRADER-WILLI SYNDROME

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INTRODUCTION: People with Prader-Willi syndrome (PWS) have hyperphagia that leads to obesity if uncontrolled. Their well-documented ability to consume very large amounts of food without feeling discomfort is due, at least in part, to their characteristic high pain threshold and high vomiting threshold. Control of food intake combined with exercise can prevent or treat the obesity, and many affected individuals today are not obese. However, the hyperphagia cannot yet be controlled medically. We here report the occurrence of unexpected mortality in adults with PWS, most of whom are slim, who develop gastric dilatation and necrosis sometimes leading to rupture and death, often following an eating binge.

METHODS: In recent years there have been a number of efforts to identify the various causes of death in individuals with PWS. The Prader-Willi Syndrome Association (USA) is conducting a large-scale questionnaire survey of families.

RESULTS: Thus far, their database contains a total of 178 deaths, of which 6 individuals had suspected or documented gastric necrosis and rupture. Age range is 17-46 years. All were relatively slim. There were 4 males and 2 females. An additional 7 individuals had a suggestive clinical course but no confirmatory documentation. Typically, there was an episode of binging associated with a holiday or special event followed several hours to a day later by complaints of stomach pain and sometimes evident abdominal distention and/or vomiting. Since complaints were not strong, they were often ignored. Death was sudden with the cause being sepsis when documented.

DISCUSSION: Wharton et al. (1997) reported 4 adults (3 F, 1M) with PWS having acute gastric dilatation and necrosis, all of whom were slim. One died. Four others had dilatation without necrosis. In an international series of 27 people with PWS who died, Schrander-Stumpfel et al. (2004) reported two adult males (one obese) died with abdominal pain, one of whom had autopsy-proven gastric dilatation. Gastric dilatation and necrosis also occurs in people with anorexia nervosa following an eating binge. The pathophysiology is controversial, with both vascular occlusion and thinning of the mucosa in a shrunken stomach as proposed explanations. In individuals with PWS, abdominal complaints and dilatation should prompt rapid evaluation and consideration of possible gastric necrosis and rupture, especially following a binging episode in those who are slim or have lost weight over a long period, whether or not complaints seem adequately strong.

Working for your Country, Working for the World

The role of the board is to act as the governing body of IPWSO, within the IPWSO Constitution, to establish policies and objectives, to look to the future well-being of the organisation by ensuring good financial management, leadership and accountability to its members. Board members are appointed for a 3 year term, with an optional additional 3 years after that. At this next General Assembly, we will be looking for new board members with a desire to work for, and uphold the principles of IPWSO as stated in our Mission on p.2.

Serving as a board member can be one of the most challenging and rewarding of volunteer assignments. While appointment or election to a board is an honour, board members have important legal and fiduciary responsibilities that require a commitment of time, skill and resources. Prospective board members need to ask basic questions about this commitment before agreeing to stand for election. Good guidance can be found online about joining a charitable board, but also you can ask questions of any one of the current board members.

In May, 2010, IPWSO will be holding its 7th General Assembly. Similar to an annual general meeting, but held every three years in conjunction with our international conference, this is a time when all parent and scientific delegates gather to look at the work the IPWSO board has been doing over the past three years, and to elect new board members. It's an important time for IPWSO as the organisation can only ever be as good as its board of directors.

Questions you should be asking are:

Do I understand how IPWSO works, or do I need more information?
How do IPWSO's current programs relate to the mission?
Does the organization have a strategic plan that is reviewed and evaluated on a regular basis?
Ask questions about the organization's financial status
Is the financial condition of the organization sound?
Does the board discuss and approve the annual budget?
How often do board members receive financial reports?

Ask questions about the structure of the board

How is the board structured?
How often does the board meet?
Are there descriptions of the responsibilities of the board as a whole and of individual board members?
Are there descriptions of board committee functions and responsibilities?
Is there a system of checks and balances to prevent conflicts of interest between board members and the organization?

Ask questions about individual board members' responsibilities

What are the ways that you think I can contribute as a board member?
How much of my time will be required for meetings and special events?
How are committee assignments made?

IPWSO
INTERNATIONAL PRADER-WILLI SYNDROME ORGANISATION

If you think you have the time, commitment and dedication to help us make a difference in the world, you should talk first to your PWS Association. They should then contact the Secretary, Linda Thornton, and put forward the name of their nominee. We will require a brief biography and a photograph. Please give this serious consideration – we need your help and support.