People With Strength

Newsletter for parents by parents

Volume 16 Issue 2 **AGGRESSION** June 2013

Prader-Willi Syndrome Association of South Africa Non-profit Organisation No. 035-837-NPO, PBO Exemption No. 930 016 853, PO Box 2399 Brooklyn Square 0075, www.praderwilli.org.za

8th International Prader-Willi Syndrome Organisation Conference

Cambridge July, 2013

Thank you IPWSO and PWSA (UK) for the 8th International PWS Conference. Being the parent of a child with PWS can be a daunting prospect. Sometimes the responsibilities seem overwhelming, even though we deeply love our child. We are therefore so grateful to the professionals, researchers, caregivers and volunteers who support us and share this responsibility with us. Thanks everybody who spends time and energy to contribute to the well being of the person with PWS.

CONTENTS

WHAT IS PRADER-WILLI SYNDROME?

- Genetic disorder
- Floppy baby (low muscle tone)
- Feeding difficulties
- Cognitive impairment
- Increased appetite
- Obesity
- Food related behaviour problems
- Obsessive-compulsive features

Chairperson:	Rika du Plooy	rikadup@mweb.co.za	012 344 0241
Vice-chairperson:	Janet Legemaate	legemaate@absamail.co.za	031 767 4493
Secretary:	Brian Legemaate	legemaate@absamail.co.za	
Medical Advisor:	Dr Engela Honey	engela.honey@up.ac.za	012 319 2269

FROM THE CHAIRPERSON

Dear Readers

A fantastic fall has spoiled us all, but winter has eventually arrived and that is less comforting.

Some positive feedback was received from readers on the content of the March 2013 newsletter. You may still remember that this newsletter was all about the younger child and special educational needs.

In this newsletter I want to concentrate on an aspect which can be very traumatic for parents: the aggressive behaviour of a person with PWS. During a short period I have received three enquiries about aggressive behaviour which was displayed by young adolescents. This type of behaviour can even led to the damaging of property and attacks on persons. Interesting, though, is the fact that all three cases were boys round about 15 years of age. Please note, girls aren't excluded from aggressive behaviour.

When your child is still young many aspects of PWS are quite disturbing and it is no fun to hear or read about it. I acknowledge that one must take special note of everything available in this regard. The content of this newsletter is therefore not only of importance to parents with older children, but also to parents who want to know more about the correct handling of the younger child. The correct strategies even from a young age are very important. To cure incorrect behaviour in a later stage is not easy. It can make life much more complex for everybody concerned. To avoid negative behaviour also entails that parents should be finely tuned to their child or young adult with PWS as human beings. Listening to and honest communication on their level is as important as with any other child.

Famcare was since the beginning of the year busy preparing an article to support parents in the handling of predicted change and sudden change in the life of a person with PWS. By experience parents know that routine must strongly feature in their households. They also know how rigid their child can be regarding his/her actions and thoughts. They like routine, boundaries and uniformity. Change leads to uncertainty which, in turn, leads to anxiety which manifest in different ways. It is also important to remember that frustration is caused by a feeling that they are not understood correctly and by the fact that they do not have the ability to express themselves verbally – especially in adolescents. This may also lead to misbehaviour. It is important to acknowledge the feelings of a person with PWS. Time must be set aside for communal planning and problem-solving. The reason for misbehaviour is not always clear. It is advisable to take time and trace it back to the cause. Please read *Coping with Change* the article from Famcare which was written by parents and informed professional people. Also available on http://www.ipwso.org/support/famcare/

Read the article by a couple who was confronted by their 15 years old son's extreme aggressive behaviour. Their calm and loving son became unmanageable. I appreciate the fact that they were willing to share their trauma with us. Their motivation for this was that other parents may learn from their experience. Other members who experience similar problems are welcome to share their hardships. Please respond to the Segals' plea for advice. South Africa's dilemma is still to find a permanent refuge for our children.

Also read the remarks by two parents and Georgina Loughnan who give us illuminating feedback on their experience and handling of aggressive behaviour.

When the behaviour of a person with PWS is such that it can be dangerous for other people, the person should be admitted to a hospital or a clinic. Care by a psychiatrist is essential. It is advisable that parents submit information on the syndrome and the management of behaviour to the professional people and caregivers. Wherever possible the PWSA (SA) will also help to supply appropriate information to parents. Enquiries can also be directed to Dr Engela Honey, the medical advisor of the PWSA (SA).

The article PRADER-WILLI SYNDROME: FINDING PSYCHIATRIC HELP FOR YOUR CHILD explains psychiatric symptoms and how the symptoms can communicate distress. Useful guidelines are given in order to help the parent to prepare for a visit to a psychiatrist. Please take time to read this as well.

Take note of a recent article, *Gastroparesis: The Newest Threat*. It appeared in the latest edition of *The Gathered View,* PWSA USA. This article may be disturbing for the reader, but by reading it you may make the world of persons with PWS a safer place. According to the article the stomach of a person with PWS empties slowly and not fully. Weakened muscles in the stomach wall may be the cause. This condition may lead to infection which may cause the death of the person. This may be a suitable article to give to your doctor. It is also electronically available from chairperson@praderwilli.org.za.

Look at our potpourri of information. Sources are given where obtainable. For those readers who do not have access to the internet, the PWSA (SA) will gladly supply the necessary information.

The 8th International PWS Organisation Conference will take place from 18 to 21 July in the United Kingdom. The General Assembly of IPWSO takes place on the Sunday where I will represent the members of PWSA (SA). Many thanks to PWSA (SA) who is sponsoring my visit to Cambridge.

Also take note of the following:

- Visit the blog of IPWSO – interesting piece on Michela, a young lady with PWS who will marry Roberto later in the year.
- *IPWSO Medical Alert Booklets* which is still available.
- Welcome to Alwin and Paula Carstens of Malelane.
- Membership fee for 2013 is now payable.
- This newsletter is available electronically in A4 format when requested. Please e-mail it to everybody who may be interested in the content.

The next newsletter will be published by the end of November 2013. The next important event is the Annual General Meeting on Sunday, 25 August 2013. More particulars will be sent by mail or email. Best wishes until then.

Greetings

Rika du Plooy

Thanks to Francis Morrison, a volunteer who did the translation.

VAN DIE VOORSITTER

Liewe Lesers

Ons hier in Pretoria is bederf met heerlike herfsweer, maar die winter het nou toegeslaan.

Die Maart 2013 nuusbrief het gunstige reaksie ontlok en soos lesers sal onthou was die inhoud hoofsaaklik op die jonger kind in die skoolomgewing gerig.

In hierdie uitgawe wil ek graag spesifiek aandag gee aan 'n aspek wat vir ouers baie traumaties kan wees en dit is aggressiewe gedrag wat by die persoon met PWS kan voorkom. Ek het in 'n kort tyd drie navrae oor aggressiewe gedrag by jong adolosente ontvang. Die realiteit is dat die gedrag beskadiging van eiendom en aanvalle op persone ingesluit het. Wat dit interessant maak, is dat in al drie die gevalle, dit seuns van ongeveer 15 jaar was. Let wel, hierdie gedrag is nie net beperk tot seuns nie, maar kom ook by die vroulike geslag voor.

Wanneer jou kind klein is, is baie aspekte van die Prader-Willi-sindroom ontstellend en is dit nie lekker om daarvan te hoor of daarvan te lees nie en is dit makliker om dit liefs te vermy. Ek besef net meer en meer dat dit goed is om van ALLES kennis te neem. Die inhoud is dus nie net vir ouers met ouer kinders nie, maar ook veral gerig op die korrekte hantering van die jong kind. Korrekte strategieë van jongsaf is so belangrik – om verkeerde gewoontes later af te leer is baie moeilik en maak die lewe vir almal ingewikkeld. Hierdie voorkoming behels dat ouers fyn ingestel moet wees op die menswees van die kind/jong volwassene met PWS. Net soos met ander kinders is luister en eerlike en opregte kommunikasie op hulle vlak, so belangrik.

In hierdie tyd was die Famcare komitee besig om 'n artikel saam te stel met die doel om ouers te ondersteun in die hantering van voorspelbare en skielike verandering in die lewe van die persoon met PWS. Alle ouers weet hoe belangrik roetine in PWS huishoudings is en hoe rigied ons kinders in hulle optrede en denke kan wees. Hulle hou van roetine, grense en eenvormigheid. Verandering bring onsekerheid wat weer kan aanleiding gee tot angstigheid wat op verskillende maniere tot uiting kan kom. Wat belangrik is om te onthou, veral by die adolosent, is dat die frustrasie wat volg uit die gevoel dat hulle nie verstaan word nie en ook nie oor die vermoë beskik om hulle gevoelens verbaal uit te druk nie, ook aanleiding tot wangedrag kan gee. Dit is belangrik dat die persoon met PWS in die saak geken word en dat daar tyd sal wees vir beplanning en gesamentlike probleemoplossing. Die oorsaak van die gedrag is nie altyd duidelik nie, maar dit is goed om op die spoor terug te gaan en te soek na die oorsaak. Lees gerus Famcare se artikel wat saamgestel is deur ouers en ingeligte professionele persone. Ook beskikbaar op http://www.ipwso.org/support/famcare/

Lees gerus die weergawe van 'n ouerpaar wat gekonfronteer was met erge aggressiewe gedrag by hul 15 jarige seun. Hulle rustige en liefdevolle seun het onregeerbaar geraak. Ek het waardering dat hulle die trauma met ons deel. Hulle motivering was dat ander ouers moontlik uit hulle ondervinding kan leer. Lede en ander wat ook in dieselfde bootjie was of steeds is, is welkom om hulle ervarings te deel. Enig iemand is welkom om op die Segals se pleidooi om raad te reageer. Die dilemma in Suid-Afrika is steeds om geskikte sorg of 'n permanente heenkome vir ons kinders te kry.

Lees gerus net na hierdie artikel insiggewende terugvoer deur twee ouers en Georgina Loughnan oor hulle ervaring en hantering van aggressiewe gedrag.

Wanneer die persoon met PWS se aggressiewe gedrag sodanig is dat hy of sy vir hom- of haarself of vir ander 'n gevaar inhou, is dit belangrik dat die persoon in 'n kliniek of hospitaal opgeneem word. Die sorg van 'n psigiater word in sulke gevalle aanbeveel. Dit is belangrik dat ouers inligtingsmateriaal oor die sindroom en veral oor die hantering van gedrag aan die professionele persone en versorgers voorsien. PWSV (SA) sal ouers vir sover dit moontlik is, van die nodige inligting voorsien. Navrae kan ook aan Dr. Engela Honey, die mediese raadgewer van die PWSV (SA) gerig word.

Die artikel *PRADER-WILLI SYNDROME: FINDING PSYCHIATRIC HELP FOR YOUR CHILD* verduidelik wat psigiatriese simptome is en hoe dit ook 'n vorm van kommunikasie is. Handige wenke word gegee om die ouer te help om voor te bery vir die besoek aan 'n psigiater. Lees dit gerus.

'n Onlangse baie belangrike artikel handel oor *Gastroparesis: The Newest Threat.* Hierdie artikel het in die jongste uitgawe van *The Gathered View,* PWSA (USA) verskyn. Die versoek is gerig dat die artikel wyd versprei moet word. Ook leesstof wat vir die leser ontstellend kan wees, maar as hierdie inligting die lewe van ons kinders met PWS veiliger kan maak, is dit die moeilte werd om dit te lees. Volgens die artikel het 'n groot persentasie van persone met PWS 'n maag wat stadig en nie goed leegmaak nie. Dit blyk dat die verswakte spiere in die maagwand die rede daarvoor is. Hierdie toestand kan tot infeksie lei wat die dood tot gevolg kan hê. 'n Goeie artikel om aan die huisarts te gee. Die artikel is op aanvraag elektronies beskikbaar vanaf chairperson@praderwilli.org.za.

Kyk gerus na die potpourri van inligting. Bronne word gegee - vir daardie lesers wat nie toegang tot internet het nie, sal die PWSV (SA) graag help om die inligting beskikbaar te stel.

Soos lesers weet vind die *8th International PWS Organisation Conference* van 18 – 21 Julie 2013 in die Verenigde Koningkryk plaas. Die Algemene Vergadering van IPWSO vind die Sondag plaas, waar ek die lede van die PWSV (SA) sal verteenwoordig. My dank en waardering aan die PWSV (SA) wat my besoek aan Cambridge borg.

Lees ook meer oor die volgende elders:

- Besoek gerus die blog van IPWSO – interessante stukkie oor Michela, 'n jong dame met PWS wat Oktober hierdie jaar met Roberto in die huwelik tree.
- *IPWSO Medical Alert Booklets* is steeds beskikbaar
- Baie welkom aan Alwin en Paula Carstens van Malelane.
- Ledegeld vir 2013 nou betaalbaar
- Hierdie nuusbrief is ook elektronies in A4-formaat beskikbaar – epos dit gerus aan almal wat mag belangstel

Die volgende nuusbrief verskyn eers weer einde November 2013. Volgende belangrike gebeurtenis is die Algemene Jaarvergadering wat Sondag 25 Augustus 2013 gehou word. Hou die pos dop vir meer inligting. Tot dan.

Vriendelike groete
Rika du Plooy

We hope you find the newsletter of the PWSA (SA) interesting and helpful.
If you are not a member of PWSA (SA) please consider a donation to the association.
It will be a great help in supporting families country wide.

NOTICE AND INVITATION

THE PRADER-WILLI SYNDROME ASSOCIATION
TAKES PLEASURE TO INVITE YOU TO THE ANNUAL GENERAL MEETING ON
SUNDAY, 25 AUGUST 2013
Vriendekring Bowls Club Grounds, Groenkloof, Pretoria
More info to members to follow

KENNISGEWING EN UITNODIGING

DIE PRADER-WILLI-SINDROOMVERENIGING
NOOI U VRIENDELIK UIT NA DIE ALGEMENE JAARVERGADERING OP
SONDAG 25 AUGUSTUS 2013
Vriendekring Rolbalgronde, Groenkloof, Pretoria
Inligting aan lede volg

THIS HAPPENED TO US……

I want to thank Ivan and Brenda Segal who shared their traumatic experience. We need to realize that what happened to them isn't an isolated case, but also happened to other parents worldwide. They trust that other parents and carers will learn from their experience and emphasised that adolescence is a challenging time for the child with PWS.

Nothing and nobody could have prepared me for what happened recently, very recently! I am still in shock as I write.

For 14 or so years we have been able to, more or less, manage our son with Prader-Willi syndrome's weight as he is very aware of his syndrome and of healthy choices. Usually after much negotiation we come to a compromise about the food issue. His love of diet sugar has become a problem to deal with as he knows it's not fattening and wants too much.

Being blessed with a large family there is always something on the go. Whether it's a wedding or a celebration of a new baby, or siblings going overseas to study or the many cultural holidays we celebrate, always with the customary abundant food. During this busy time we failed to pick up that our son wasn't coping so well and was getting used to take more and more to eat than usual. Slowly our "sweet child" was becoming more and more out of hand without it being addressed.

When all the family activity subsided and I started to restrict him, things were not going his way with the result that he became increasingly difficult. He refused to go to school and did not listen to us at all. He also became destructive and even crushed a pair of his sibling's sunglasses and refused to pay for the damage he caused. Family also reported that he took food when I wasn't at home. I now, in hindsight realized that there were many signs that should have warned us.

To cut a long story short - after staying up late one night to play computer (not usual for him) he assured us he would go to school the following day. Unfortunately the following morning it didn’t happen and things went from bad to worse. I locked him in his room (something I’ve never done before, but the housekeeper was petrified) while I took the others to school. When I got back (which wasn’t long) he had broken the lock so I couldn’t open the door. He then broke part of the door, the lock and handle. After he came out he took a large breakfast looking very pleased with himself. He played computer again, and after about an hour he wanted a snack. I tried to stop him and he became violent. If it weren’t for my other son and gardener who pulled him away from me it could have turned out very badly….. He was unreasonable after that - taking a hammer and smashing a window.

The gardener was amazing with him and managed to calm him somewhat. In between all this he drank one and a half litres of grape juice and half a two litre coke. I called an ambulance and they came to our rescue and managed to take him willingly to the Rand Clinic. Much to his dismay and surprise he ended up staying at the clinic for one and a half weeks showing aggression in the beginning. It took five security men to hold him down when I left! He improved and was allowed to return home on condition that he attends school. (He had been doing well at school previously).

When our son came home, his arguments about food started again. He occasionally went to school and when he refused to go to school he just slept and ate and gradually started to become aggressive again. He has been on medication since he left the clinic. He now came with a new thing which was most upsetting. He was too lazy to go to the toilet, took a bottle to urinate in and often messed all over the place.

We arranged to have him institutionalized at the Selwyn Segal Home, an institution for the Jewish mentally handicapped. On his arrival there he threw the coffee over the social worker and was very insolent. It was reported that he threw plates from the kitchen on the floor and also punched out some windows. That evening he broke a cupboard to get to a resident’s yoghurt. The aggression escalated and it was most of the time about wanting more food. The social worker said she couldn’t believe his strength. At that stage he was admitted to the Rand Clinic again and he still is there at present.

We don’t know how to proceed, as we can’t take our son home and we don’t know if the Selwyn Segal will accept him again. We would appreciate if you could recommend any institutions that are geared for our children. We can only pray and hope that with medication, things will be OK.

What I wanted to stress is that we never took adolescence and even “normal” teenage behaviour into account. In hindsight I see that he was depressed and even a little envious, and with the added stress of puberty and being a teenager it escalated into behaviour that was frightening for everyone. We hope to never witness such again and to try preventing it from happening to anyone else.

Thank you to Rika du Plooy and the Prader-Willi Association (SA) for all their help, support and advice. We are desperate to hear of any advice or suggestions.

We hope to have our well behaved, helpful young man back that we so dearly love.

The boy was dismissed from the Rand Clinic as the Medical Aid allows only 21 days per year. However, the boy was re-admitted to Selwyn Segal.

REMARKS AND ADVICE FROM TWO MOTHERS AND A PROFESSIONAL

From Lesley the mother of David, 46 years

Due to firsthand experience over what seemed a longish period of time, I can perhaps offer something.

It first happened at our house when our son was about 18. It shocked us all, including his younger sister and brother, more than other behaviours because it was so loud and dangerous and horrible to clean up.

Whether the breaking of a window is due to a recognisable change or inability to cope with life as the person with PWS finds it at a particular time, once it happens it can happen again and again when the person can't cope. When our son moved into a residential setting when he was 25, a huge change for him, windows were broken there and this happened over a longish period of time until the cycle was broken by gradually replacing glass with clear unbreakable glass alternatives. It worked. This was costly and inconvenient but most certainly absolutely necessary to break that cycle of behaviour. In our experience, not all the windows in a house have to be replaced, just the ones previously targeted and the ones in close proximity to those.

> *I want to suggest two very minor things - but important to me as a parent. Self worth or self esteem is difficult for our people with PWS to have but it will improve with good management. The second one is simply an encouragement for parents and carers to not feel guilty or hopeless when things go wrong and difficult behaviours happen - try sound management suggestions again and again. Lesley*

A well designed *Behaviour Intervention Plan* with preventative strategies such as in the *Coping with Change* document can help to avoid or defuse difficult situations, but there has to be acceptance by people managing the person with PWS that situations can escalate when least expected. Even though all the management plans are in place, it seems that once glass has been broken there may be repeat occasions by that person due to the 'remembered' sound or impact of breaking glass.

So there has to be a plan in place to break the cycle of breaking glass! Just as there have to be plans in place for breaking the cycle of other undesirable behaviours.

In our case, there have been no repeats of breaking glass for some years.

From Linda the mother of Francie, 29 years

I have had a lot to do with aggression in PWS. Lots from my daughter and during the training sessions I presented - it is a topic that comes up very frequently.
Mostly, you can connect it to anxiety where changes occur. Usually they were unable to talk about it, understand it, or deal with it. Anxiety becomes heightened; behaviour becomes erratic and often dangerous.

My daughter who is now 29, and much more compliant and less likely to have a melt-down, had a HUGE one the other day. The staff said she just "got up in a bad mood" and it went from bad to worse all day. They could do nothing to help her because she couldn't talk about the thing that was bothering her - and the reason for that was because she probably wasn't even aware of it. Anyway, she took a knife to the padlocks and tried to get more food; she threatened the staff ("I'm going to kill you") and then smashed the ceramic stove top. The staff did all the right things; called for help, stayed out of her way, monitored her for safety, etc.

What was going on for Francie was this: a week before, one of her friends with PWS died very suddenly (ruptured stomach). I took Francie to the funeral where she was really very good - she talked to Paul in his coffin, she wrote a message and left it on the coffin, she talked to his parents, and to anyone else she knew. She was - I thought - fine. But she wasn't. She had kept asking me every day if I knew what had caused Paul's death. I didn't tell her; I just said that his mother was going to give me a copy of the autopsy when it came. I only guessed at his demise, I didn't really know. This disturbed Francie and she became over-anxious. She couldn't express her anxiety and I didn't know just how anxious she had become.

I am quite sure this outburst related to her fear of dying the same way as Paul had done.

Because she seemed to be so OK about it, I didn't worry about things and just thought I'd done the right thing by taking her to the funeral. In the future, I will make sure that she has a counsellor to talk to about this kind of grief/change/anxiety.

From Georgina Loughnan a professional at The Prader-Willi Syndrome Clinic, Camperdown NSW, Australia

In our clinic, we see aggression, frustration and anger developing in boys from 13 – 18 years – often for the first time. What we see is similar to non PWS teenage boys, but the severity and lack of ability to express their anger in words (also common to other teenagers!) seems to increase the intensity of, otherwise, PWS behaviour. This is also a period of time when many males are started on testosterone. Is it the testosterone that causes this increased aggression or is it a greater awareness of the limitations that having PWS places on their lives? The gap between them and their peers widens and the bad behaviour is often repeated. (In our clinic the testosterone is started at a very low dose and gradually increased, but yes, it is known to increase aggression in some males on larger doses).

I think when property is damaged during an event of aggression – the event needs to be discussed once the person has completed the “anger period”. As Damien Jones advised – asking “what caused you to be upset?” and “how could we all do things differently to help you through such an event?” and just quiet talking and lots of listening – is most helpful. However, I think what needs to be added is the acknowledgement that damaging other people’s property is not acceptable and needs to be paid for – even if it takes a small amount of money each week for a few years to do so. We can’t sell our people with PWS short. They like responsibility and although the responsibility of paying for damaged goods may not prevent them repeating the action in the future – it often helps reduce the amount of damage caused next time. When they pay for the damage it in some way should mean they go without something that the money would have been for. Consequences are important but obviously need to be suitable.

BAIE WELKOM

‘n Hartlike welkom aan Alwin en Paula Carstens van Malelane, wat Mei 2013 by PWSV (SA) aangesluit het. Hulle dogtertjie Lareine het in Meimaand een jaar oud geword.

COPING WITH CHANGE IN PEOPLE WITH PRADER-WILLI SYNDROME

Famcare is a project of IPWSO: famcare@ipwso.org

Most people with Prader-Willi syndrome (PWS) have difficulty coping with change. This varies from person to person and can be a real problem for those families affected. Change can be anything from a substitute teacher at school, to a different route being taken in a car trip or a new cup being used. Change can also occur suddenly and unexpectedly, as in an electrical blackout. A bad reaction to change may result in the person with PWS refusing to comply with requests, routines or plans and can quickly escalate into perseveration (repeated questions or comments), arguments and aggression. "Shutdown" is another typical response to the anxiety associated with change. These responses can be stressful for families as they often occur at the most inconvenient times.

Why do people with PWS have difficulty coping with change?

The problem is thought to be linked with the inability to "switch" attention from one thing to another. People with PWS generally find it more difficult to switch attention. Researchers from the UK have shown that the greater difficulty a person with PWS has in switching their attention from one thing to another, the more resistance they show to change. It is also known that people with PWS tend to prefer repetitive routines and often exibit ritualistic or inflexible behaviours. Although varied in their reactions to change, they all feel "safe" with set schedules and expectations. If a change can be predicted in advance, it is possible to use strategies to avoid much of the anxiety that may otherwise result from the change occurring.

What works to avoid anxiety related to change

These are simple and practical strategies that help to minimise the reaction to *predicted* change:

- Discuss a back-up plan in advance. For example, you could agree to buy pears if there are no apples at the fruit shop. The fact that this is discussed in advance can prepare the person for a different situation to the one they are expecting.
- Thoroughly plan all details and double-check things with the person with PWS and with others. Occasionally the person with PWS has an expectation that you are not aware of. For example, they may expect that you will be stopping for a cup of coffee on the way home, however if you are not aware of that and drive directly home, that would be perceived by them as a change. So it is always helpful to go over the details of any plans, beforehand.
- Agree on some rules for an outing. For example, a rule that "If there is any yelling for any reason we will return home." Before the outing reminds the person of the rule, using positive terms e.g. "your best behaviour will mean we can finish the trip and won't need to come home early". A set consequence of bad behaviour can be hard to enforce at the time, however, going home once can stop a lot of problems in the future.
- Praise the person whenever change is accepted graciously.
- Use picture-boards or written routines. Any change to routine is added to the board in advance. For example, a schedule could be written for the week. Doctor's appointments or variations in the normal week can be written on it. If the person

with PWS has any problems with the plans they can tell you in advance so, hopefully, any problems can be discussed and a solution can be negotiated.

What works to limit reaction to an unpredicted, last minute change of plans?
At times, unpredicted and unplanned change does occur. If you are fortunate enough to be able to respond quickly you may be able to reduce anxiety.

Below are some strategies that parents and carers have found successful in similar situations

- Give a solution before explaining the change. For example if a favourite coffee shop is closed for renovations, start by saying, "We are going to have coffee today at another coffee shop. That'll be different, won't it! We can ask them to make it just how you like. The coffee shop we like will be open again next week but it is closed today so we can't go there today." The last piece of information given in this example is the fact that the shop is closed and the fact that they will still get a nice coffee, comes first.
- Give a visual example of the problem or the change. For example if you were planning to move a cupboard into a room and it did not fit, showing a person with PWS the cupboard outside the door is better than trying to explain the problem in words. Simple sketches can at times be more easily understood than words.
- Be patient and calm as it may take some time for the change to be processed and then accepted.
- Try to point out the positive aspects that are involved in the change.
- Expressing shock at the change, yourself, and asking the person with PWS to help in the situation can diffuse their reaction to the change if they are given some responsibility within the event.

Managing change is an important part of managing the overall well-being of a person with PWS. Effective management of change can reduce anxiety and improve outcomes in other areas such as health and behaviour management.

Even 'good changes', such as a holiday, can be a sources of stress for the person with PWS and need to be managed as a change to routine. Perseveration, increased anxiety and worry about what will happen while they are away and so on, are often seen. Not telling the person with PWS about something 'good' coming up for them is an option, but not always a good solution.

Working out changes with the person with PWS, so that a better result might be achieved next time, can be useful. For example: " Next time, if you think your cat has been in a fight and might have an abscess, do you think it would be better to go straight to the vet instead of trying to fix it yourself?" If the answer is 'yes', then you can incorporate it into their guidelines – write it up on their pin-board, and make sure they know the new plan!

After an argument, or a 'blow-out', and when the person with PWS has become quiet and even remorseful, ask them what *you* can do to help them next time something like this upsets them. You may be surprised at their answers and often they can lead to positive results. For example: "Please leave me alone for a while," "Please listen to me," "Please don't treat me like a child," These sorts of agreements can often prevent a future argument or blow-out.

It's all about compromise

As the person with PWS grows older, their behaviours may mellow. There is an expectation from the person themselves, as well as parents, siblings and family friends that they will be treated more and more like an adult. As parents and caregivers, we know this is not always possible, but we can sometimes be a little more flexible and trusting in certain areas. It is difficult to let go, and there will be times that the trust between you will be broken, but if you compromise, safely, often you can reach an agreement. As PWS lack of judgement and poor emotional control are not visible when they are calm and happy always prediscuss consequences.

Practising increased, safe responsibility can facilitate positive behavioural change.
For example: Carrying a set amount of money for a specific, preplanned event can encourage self confidence and trust. However, success with this will take much preparation, explanation and negotiation. Having a "back-up" agreement to cover times when such a compromise does not work successfully is essential, such as: "If you spend the money instead of using it for what we've discussed it will be best for you not to take the money next time. I will then continue to organise the payment so you are not tempted to spend it on food." People with PWS vary in personality and cognitive ability. Some people can cope with more responsibility than others. If monetary responsibility is not necessary it is best avoided so as not to set the person up for failure.

A parent says: The main thing about compromise is that if you are going to *take something away* from a person, you must always give something back. *For example:* "Unfortunately I cannot let you keep a dog in this house because we really don't have room. But, you can keep a bird in a cage or a goldfish in a bowl, and we could always arrange for you to help walk some of the poor dogs at the SPCA who never get a chance to go out".

Appealing to their own sense of judgment ("we don't really have room") and getting them to see that fact; giving something back (goldfish, or bird, for example) and appealing to their sense of importance, of doing something for others (walking someone else's dog), seems to work.

Preparing people with PWS for change gives them the opportunity to cognitively and emotionally process what is to happen and how the change will affect them. It may take time and effort to prepare people for change but the results are usually worthwhile. However, if the person with PWS is stressed for a reason unknown to you or they are aware of anxiety you may be experiencing, difficult behaviour can occur despite your use of all of the above suggested strategies. Don't feel like they or you have failed. Keep applying positive proactive strategies. In time, improvements will be noticed.

BIRTHDAY CARDS

Every child or adult with PWS on the membership list of PWSA (SA) is due for a birthday card. Please inform us if your child doesn't receive a card. We would like them to experience the excitement of a birthday card. A friendly request – please acknowledge receipt by SMS either Karin or Magdaleen.

Karin (English cards): 083 681 6842

Magdaleen (Afrikaans cards): 083 663 7234

FINDING PSYCHIATRIC HELP FOR YOUR CHILD

Janice L Forster, MD Linda M Gourash, MD ***Pittsburgh Partnership,*** **Pittsburgh, USA**

Introduction

Psychiatric symptoms can emerge in anyone when the ability to cope with stress has been compromised. Coping mechanisms can be adaptive or maladaptive. Coping strategies can be learned but they are more likely determined by temperamental characteristics called personality traits. Temperament is both genetically determined and shaped by interpersonal experiences. Individuals with developmental disabilities have intellectual deficiencies, brain differences, and/or physical and sensory handicaps which affect their temperament, their capacity to relate to others in their environment, and their ability to acquire coping strategies. They are not only more likely to experience stress, but they are also less likely to be able to cope with stress adaptively. In these individuals psychiatric symptoms may be the way in which they communicate their distress. Internalizing behaviours (anxiety, depression) and externalizing behaviours (frustration, disruption and aggression) are common psychiatric symptoms which indicate distress.

Psychiatric symptoms may also emerge as a manifestation of an underlying psychiatric disorder. Psychiatric diagnoses are made when a specific cluster of symptoms occurs over a long enough time period to alter an individual's level of functioning in their daily environment. The most common psychiatric disorders are anxiety and depression. Another common set of diagnoses originate in childhood and are classified as disruptive behaviour disorders. The most severe psychiatric disorders are indicated by the presence of psychotic symptoms defined by hallucinations, delusions, and disordered thinking. In general, the predilection for a particular psychiatric disorder can be familial, genetically determined, associated with specific chromosomal abnormalities, or caused by brain injury. They are also frequently triggered by stress. Certain temperamental traits are also prodromal.

Psychiatric evaluation can:

- delineate symptoms
- identify stressors
- recognize the strengths and weaknesses in coping strategies
- indicate the presence of an underlying psychiatric disorder

Recommendations for intervention may include:

- prescriptions for environmental change
- alterations in interpersonal interaction through behavioural modification
- education to teach new coping strategies
- medication to alter the individual's threshold for response to stress
- medication to treat an underlying psychiatric disorder

The Psychiatrist's Role

It is the role of the psychiatrist to obtain a thorough data base including developmental, behavioural, family and medical histories. This may involve interviews of family members, teachers, staff and/or other caretakers, as well as a review of records. The psychiatrist will also interview or observe the patient to obtain a mental status examination. This series of questions and assessments allows the psychiatrist to evaluate the patient's attention span, concentration, motor function, speech and language, interactional abilities, mood and affect, thought patterns and problem solving abilities. The mental status examination provides a window through which brain function is indirectly explored. The psychiatrist organizes the data and synthesizes a case formulation delineating the nature of symptoms, their etiology and their significance. A differential diagnosis is made, and further diagnostic testing is recommended. Ultimately, multimodal treatment interventions are suggested. Depending upon the expertise of the psychiatrist, some of the treatment recommendations may be referred to other professionals on the mental health team.

PWS Intervention Pyramid

It is not necessary for the treating psychiatrist to have experience with Prader-Willi Syndrome. Because this is a rare disorder, there are few clinicians who have had the opportunity to see large numbers of patients with PWS. Rather, parents should seek a credentialed psychiatrist (ABPN certified in either Child and Adolescent Psychiatry or General Psychiatry) who has an interest and experience in caring for persons with developmental disabilities and who is willing to listen, read and learn about the syndrome.

Above all, parents should seek a psychiatrist who is expecting to follow the patient over time and who requests office visits and follow up phone calls whenever prescribing a new medication or when changing a medication dose. Typical follow up may be in 1-3 weeks initially and at least monthly thereafter with an office visit depending on the nature of the symptoms and the type of medication prescribed. Some medications require blood testing when first prescribed, when the dose is changed and at less frequent intervals thereafter, perhaps every 3-6 months.

This article was prepared by the *Pittsburgh Partnership*, specialists in PWS to provide guidance to parents and other care providers in seeking out psychiatric services. Clinical information for the consulting psychiatrist is available in a separate article **Managing Prader-Willi Syndrome: A Primer for Psychiatrists.** This article and more information can be downloaded from www.pittsburghpartnership.com. It is also available from chairperson@praderwilli.org.za

GASTROPARESIS: THE NEWEST THREAT

by Lisa Graziano, M.A., PWCF Executive Director
Janalee Heinemann, M.S., PWSA (USA) Director of Medical Affairs
Ann Scheimann, M.D., M.B.A., Gastroenterologist and PWS Specialist

An urgent request from the PWSA (USA):

USA
PRADER-WILLI SYNDROME ASSOCIATION
Still hungry for a cure.

Please share this important information with families that may not be PWSA (USA) members or do not go to the PWS internet sites. This article will be posted in the medical section at **www.pwsausa.org**

If you're a member of the Prader-Willi California Foundation and the national Prader-Willi Syndrome Association (USA), then you have read about or discussed at a meeting of some sort this thing called gastroparesis. There have been articles about the fact that it exists, alerts about it (*Medical Alert: Gastrointestinal Issues in Individuals with PWS*), and now a peek at the likely incidence rate. What we haven't yet received enough information about is how do we know if our child/adult has it and what can we *do* about it. This is the focus of this article.

Please know that this will likely be a difficult read for those of us who care for and love someone with PWS. Knowledge is power, however, and so if the information contained within this article helps inform care providers enough that they can keep someone with PWS safer, then the read is worth it.

In 1999 *The Gathered View* included an article about the discovery by PWS specialist Rob Wharton, M.D., of what he termed Acute Idiopathic Gastric Dilation. What Dr. Wharton saw in his patient was that for some unknown (idiopathic) reason the stomach (gastric) was quickly (acute) pushed out (distended), causing the stomach tissues to die. If not immediately treated with surgery, this condition may lead to death.

Over the following years, particularly with closer examination by PWS/GI specialist Ann Scheimann, M.D., it has become clearer that a great number of other people with PWS have a stomach that empties too slowly. In fact, Dr. Scheimann now believes **it is highly probable that *a significant number of* people with PWS have some degree of a slow emptying stomach**. The medical name of this disorder is gastroparesis: the muscles in the wall of the stomach work poorly and prevent the stomach from emptying properly. As a result, food stays in the stomach longer than it should. Over time, the volume of accumulated food in the stomach can cause the stomach to become full. Like a balloon that has too much air, the stomach of someone with PWS that contains too much food can respond in one of two ways: it will rupture or the food will push so hard against the stomach lining that it "compresses and weakens" the cells in the stomach. Both of these conditions cause massive internal infection and can quickly lead to death. (*Please note that there has typically been a prior eating binge with most incidents of GI necrosis and death.)*

Other important factors to consider are that some medications such as narcotic pain relievers and anticholinergic medications (group of bronchodilators) can also cause the stomach to empty too slowly (as well as cause dry mouth symptoms). Abnormally high blood glucose (sugar) levels or undetected hypothyroidism can also slow stomach emptying; therefore, it is important to control blood glucose levels and screen periodically for hypothyroidism.

The symptoms of a slow emptying stomach are primarily nausea, vomiting, abdominal fullness after eating, and/or pain. But for persons with PWS who often have a blunted pain threshold and an absent vomit reflex, symptoms of gastro-paresis or Acute Idiopathic Gastric Dilation can be extremely difficult to detect.

At the same time the stomach empties too slowly, the bowel intestinal tract seems to empty too slowly. This means that digested food that the body turns into waste product and must eliminate from the body as faeces/stool is not entirely eliminated, leaving too much stool in the intestinal tract.

Many parents and care providers believe that because their child or adult has a bowel movement every day, this means they don't have a slow emptying bowel. This is not necessarily true. Even with a regular daily bowel movement, the intestinal tract may not empty appropriately. As the colon becomes more backed up with retained stool, the ability to evacuate stool is less effective. Over a long period of time, continuous, constant hard pushing has resulted in some people with PWS experiencing rectal prolapse. (The feeling of constant fullness and pressure on the anus or itching of the skin from irritation from bile acids present in the stool can contribute to reasons that some people with PWS insert their fingers into their anus or pick at it.)

As the colon becomes more impacted with retained stool, emptying of the stomach commonly slows down. This means that the risks of gastric rupture or dilation are dangerously elevated.

How to Detect Gastroparesis and Slow Emptying Bowel

How do we know if the individual with PWS we're caring for has gastroparesis or a slow emptying bowel? What are the signs? What are the symptoms? What do we look for? The answers are, unfortunately, that there probably aren't many easily recognizable signs or symptoms.

Because the abdominal core muscles are generally weaker in persons with PWS, the stomach can often appear to be more rounded. If food is not emptied quickly enough, the stomach can look rounded (distended) and feel "too firm" to the touch. On the other hand, for those who are taking growth hormone medication and are therefore leaner, the stomach can already feel "firm" to the touch.

The definitive test to identify delayed stomach emptying is the Gastric Emptying Study which measures the amount of time it takes for food to empty from the stomach and enter the small intestine. The test is done in the nuclear medicine section of a hospital. The patient fasts overnight and eats a breakfast that contains a tiny amount of radioactive material. The patient then lies flat and still on an exam table under a large "arm" that measures the amount of food particles that evaporate from the stomach over a period of time; generally four hours is the appropriate amount of time for the emptying study following a mixed meal of liquid and solids. There are no side effects from a gastric emptying study; the radioactive material is not absorbed into the body and is eliminated in the stool. The test can be difficult for kids under the age of 10 to complete because it is critical that the person lie perfectly still throughout the duration of the test. It is important to make certain that other factors such as constipation and/or thyroid disease are well controlled prior to completion of the test.

Treatment Strategies

1. As with all treatment of PWS symptoms, the first approach is to always provide Food Security: a) a healthy, low-calorie, low carbohydrate diet; b) meals and snacks served at structured times/sequences throughout the day; and c) all access to food restricted.
2. Request from PWSA (USA) information about GI issues in persons with PWS.
3. If there are GI concerns present, consider consultation with either a paediatric or adult gastroenterologist, dependent upon age. Provide the physician with your GI issues documents.
4. Discuss the pros and cons of a Gastric Emptying Study.
5. Discuss the use of medications such as metoclopramide (Reglan) and erythromycin to improve stomach emptying.

6. Discuss an assessment for stool build-up (e.g., palpitation, x-ray). The Bristol Stool Chart can be used to screen/track progress with management of constipation. Normal stools should be Bristol Class 4 (See Table on website) http://en.wikipedia.org/wiki/Bristol_Stool_Scale
7. Discuss the use of over-the-counter medications such as Miralax to improve stool elimination and over-the-counter probiotics to help regulate the balance of helpful organisms (micro-flora) in the intestines.
8. If there are challenging issues for your primary GI specialist physician, suggest the GI specialist contact a PWS GI specialist by contacting PWSA (USA).

We continue to learn more about the gastrointestinal and bowel emptying issues of PWS. We have a lot of questions and some theories, but no definitive answers. As we do, we will inform you. Janalee Heinemann.

A POTPOURRI OF INTERESTING INFORMATION

Undiagnosed and unmanaged, Prader-Willi Syndrome causes morbid obesity. There are, however, treatment and management strategies that save lives and improve the quality of life of all who are impacted by PWS.

THE RED YELLOW GREEN SYSTEM FOR WEIGHT MANAGEMENT (RYG)

The Children's Institute, Pittsburgh, USA

Six colourful pages illustrating the concept of the RED YELLOW GREEN system for weight control. Foods are divided into different groups based on their nutrient content and calorie levels and it provides a generous amount of food that is pleasing to the eye. The illustration is catching and it is easy to understand and even a very young child can participate and make choices, increasing the chance of good dietary compliance.
PWSA (USA) website: www.pwsausa.org and search for RYGBook or go to Products.

HELP OUR CHILDREN AND HELP OTHERS TO UNDERSTAND PWS BETTER!

DVD: FOOD, BEHAVIOUR AND BEYOND PWSA (USA)

This comprehensive DVD, a joint project of PWSA (USA) and IPWSO, is being used around the USA and overseas to train staff working with persons with PWS. It is an excellent learning tool for parents. It addresses *Nutritional Basics, Food and Behaviour, The Behaviour Toolbox, Cognitive and Behavioural Traits and To Medicate or Not to Medicate.* Parents will be able to review sections over and over again as needed and share with family, teachers and babysitters. Having years of hands-on intensive experience, Dr. Gourash and Dr. Forster's lectures on the management of PWS have received rave reviews from both parents and providers. Members, parents and other interested people are welcome to order above mentioned DVD (free of charge) from the PWSA (SA). Please contact Wilna Basson at 012 991 3399 or e-mail: bassons@iafrica.com

DVD: UNDERSTANDING THE STUDENT WITH PRADER-WILLI SYNDROME PWSA (USA) Strategies for Success

This DVD provides teachers with an introduction and overview of the issues associated with PWS and to demonstrate and described pragmatic strategies that school staff can use in supporting the success of a student with PWS in the classroom. Done from an USA view, for upper grade and lower grade students, but contains valuable strategies which can inspire school staff to create their own successful strategies.
Please contact Wilna Basson at 012 991 3399 or e-mail: bassons@iafrica.com

MANAGING PRADER-WILLI SYNDROME: A PRIMER FOR PSYCHIATRISTS

Prepared by the *Pittsburgh Partnership*, Specialists in PWS
Janice L. Forster, MD Linda M. Gourash, MD

This article summarizes the most essential information for psychiatrists or psychologists who are new to Prader-Willi syndrome and includes principles of Food Security and management strategies for common presenting problems. It is a detailed resource and is available at: www.pittsburghpartnership.com. Also available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

PSYCHIATRIC ALERT FOR PSYCHIATRISTS ON PRADER-WILLI SYNDROME

Prepared by the *Pittsburgh Partnership*, Specialists in PWS
Janice L. Forster, MD—Child and Adolescent Psychiatrist and Linda M. Gourash, MD—Developmental Pediatrician

Concise summary of things your psychiatrist needs to know before prescribing medications.
www.pittsburghpartnership.com. Also available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

PSYCHOTROPIC MEDICATION TIP SHEET FOR PATIENTS WITH PRADER-WILLI SYNDROME FOR HEALTH CARE PROVIDERS

By: Elisabeth M. Dykens, Ph.D., and Elizabeth Roof, M.A., L.P.E., Vanderbilt University, Nashville, TN

Many guidelines used to treat people with intellectual disabilities in general also apply to those with PWS. Behavioural interventions are a critical feature of treatment in PWS; they should be tried before psychotropic medications are used. Medications commonly used in people with PWS are discussed. Feedback from parents and person with PWS is important.
PWSA (USA) website: www.pwsausa.org and search for Psychiatry. Also available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

INFORMATION FOR SCHOOL STAFF: SUPPORTING THE STUDENT WHO HAS PRADER-WILLI SYNDROME

Compiled by Barb Dorn, Crisis Counsellor PWSA (USA)

All students with PWS are individuals. Each has varying strengths and needs. Common behaviours often seen in students with PWS are covered and appropriate management strategies are given. Available at www.pwsausa.org/Educator/InfoSchool.htm. Also available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

TWO BOOKLETS FROM THE UNIVERSITY OF QUEENSLAND, AUSTRALIA

Exercise and Physical Activity for children with PWS by Kristy Reid & Peter Davies

A guide for parents and carers

This guide is intended to give you a better understanding of the importance of managing your child's energy balance, exercise and/or activity levels. We hope the information will help you to ensure they are limiting their calorie intake and expending enough energy to prevent excessive weight gain over time. Other members of the community are important in supporting you and your child. We encourage you to share this guide with your extended families, other carers, educators, and people in your community.
Available via email from the chairperson PWSA (SA): chairperson@praderwilli.org.za

Need to know Nutrition for children with PWS by Prof Peter Davies

A guide for parents and carers

This booklet will provide parents and carers with many hints about how to manage the diet of a child with PWS. This is no easy task in a society where we are bombarded with food, snacks and drinks. The authors will guide you from the infant years to the teens highlighting important aspects of managing their diet at each stage and a food exchange system is explained with practical examples. Throughout the booklet, it is stressed that good habits, once established, will be of great benefit in the long term.
Available via email from the chairperson PWSA (SA): chairperson@praderwilli.org.za

NEW! ELEMENTARY SCHOOL POWER POINT

Dale and Dottie Cooper

Creating understanding is one of the keys to developing a more welcoming and successful classroom environment for students with PWS. Rob and Debra Lutz, parents of Isabella, created this PowerPoint to promote understanding and awareness in Isabella's elementary school classroom. In a fun and effective way this PowerPoint presentation helps classmates learn about the syndrome and what to expect when sharing a classroom with a student with PWS. This presentation helps to encourage stronger peer relationships and support for a student with PWS, and it enriches the world and understanding of all students as they learn the important lesson that every person is unique, has challenges, and needs support. We invite you to adapt and use this PowerPoint presentation for your child's classroom.
You can download this from the website of PWSA (USA) and revise it to fit your child.
PWSA (USA) website: www.pwsausa.org and search for Educational Awareness Tools.

A SECOND EDITION OF "GROWTH HORMONE IN PRADER-WILLI SYNDROME"

A reference for families and care providers has now been printed and is available in pdf format from IPWSO's website: www.ipwso.org.
Also available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

THE SOUTHERN AFRICAN ASSOCIATION OF LEARNING AND EDUCATIONAL DIFFERENCES (SAALED) www.saaled.org.za

SAALED in association with the UNESCO chair in teacher education for diversity and development at the Wits School of Education, will host 3 conferences aimed at making education inclusive. The conferences will address exclusionary pressures and practices in education and equip educators and therapists to meet diverse learning needs.
Inclusive education can and does work if education policy is implemented properly and teachers are adequately equipped. An education system that promotes the full participation and inclusion of ALL children maximizes every child's personal development and enable him/her to fully engage in an inclusive society.

There are many kinds of barriers to learning and many forms of learning difficulties and disabilities. No single solution is appropriate for all children. In striving to create inclusion of all learners, policymakers, researchers and educators are charged with the responsibility to identify barriers and dismantle exclusion. Many teachers are inadequately equipped to support the intervention needs of children with learning difficulties and disabilities.

VENUE: The Wits School of Education, Johannesburg
DATE: 1 – 6 July, 2013
CONTACT: Barbi Raymond at md@saaled.org.za
SAALED National Office : Tel. +27 011 325 2406

IPWSO NEWS

GENERAL ASSEMBLY

NOTICE IS HEREBY GIVEN of the 8th General Assembly of the International Prader-Willi Syndrome Organisation (IPWSO) to be held on Sunday 21 July, at 3.00 pm (UK time) at the buildings of the Fitzwilliam College, Storey's Way, Cambridge. The General Assembly is normally held every three years at the International PWS Conference, according to the statute of the organisation. The meeting will be attended by the chairperson of the PWSA (SA).

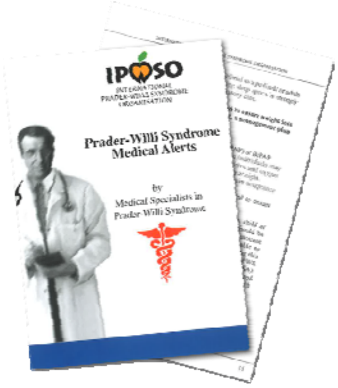
Delegates of the following countries will attend the General Assembly and have the right to vote: Argentina, Armenia, Austria, Belgium, Canada, Chile, Czech Republic, Denmark, Estonia, Finland, France, Germany, Ireland, Israel, Italy, Japan, Korea, Lebanon, Netherlands, New Zealand, Norway, Poland, Puerto Rico, Romania, South Africa, Spain, Sweden, Switzerland, Taiwan, United Kingdom & USA

Delegates will also determine which country will host the 9th International PWS Conference in 2016. There are two contenders: Canada and Israel. Each country will present a 15 minute talk after which delegates will cast a vote.

PLEASE VISIT IPWSO'S BLOG AND BECOME A FRIEND OF IPWSO

Read the different stories as well as the news and views of IPWSO around the world.
Currently on the blog an interesting story about Michela, a young woman with Prader-Willi syndrome, who is getting married to Roberto in October this year.
You are welcome to add comments.

PLEASE HELP: IPWSO MEDICAL ALERT BOOKLETS



Readers are requested to assist the committee to distribute the IPWSO MEDICAL ALERT booklets to professionals who you come into contact with. The booklets are specifically printed in A5 format and the contact details of the PWSA (SA) are included as well as the diagnostic testing procedures for PWS in South Africa. Please inform Janet Legemaate (Vice-chairperson) if you are willing to assist and how many copies you would need. Thank you to those members who already responded to a previous request! Please contact Janet Legemaate at: Tel: 031 767 4493 or 082 737 6144 or email at legemaate@absamail.co.za

THE PRADER-WILLI SYNDROME ASSOCIATION OF SOUTH AFRICA

The Prader-Willi Syndrome Association is a support group and was established by a small group of parents in March 1990.

WHAT IS PRADER-WILLI SYNDROME?

Prader-Willi syndrome is a complex genetic disorder that typically causes low muscle tone, short stature, incomplete sexual development, cognitive disabilities, problem behaviours, and a chronic feeling of hunger that can lead to excessive eating and life threatening obesity. It is estimated that one in 12,000 to 15,000 people is born with PWS. Although considered a "rare" disorder, PWS is one of the most common conditions seen in genetics clinics and is the most common genetic cause of obesity that has been identified. PWS is found in people of both sexes and all races.

ADVICE TO NEW PARENTS:

Your child has Prader-Willi Syndrome – what happens now?
From the website of the PWSA (SA) www.praderwilli.org.za

It comes as a great shock when you heard that your child is been diagnosed with a syndrome you haven't heard of before. It is natural to be upset and you may experience emotions of grief, depression, anger, bewilderment, guilt, disbelieve, denial, rejection or perhaps relief. However, how hard it seems to be, it is now the time to consider the well-being of your child and what your child needs most is your love and devotion.

Get as many information on the syndrome, but always remember that behind the syndrome, behind the mass of information, there is a human being who needs love and acceptance.

Some of the information on PWS might be very disturbing and depressing, but it is important to remember that not all people with PWS experience all of the symptoms and certainly not all at the same time in their lives. Never underestimate your child's abilities.

Share the information with family and friends. It will help them to understand your child's problem and their support can play an important role in the management of behaviour and controlling the food environment.

Establish family habits and routines from the very start that will support your child's diet and behaviour needs.

Seek professional help and please do not try to handle this complex syndrome on your own. Families have to deal with very strenuous circumstances and therefore parents and other family members might need counselling to understand and manage the complexities associated with the syndrome.

Contact a parent support group. Parents of children with the syndrome have experience and understanding of many of the challenges you will face. It implies that you need never face the future alone; there is always somebody who can listen and who can share both in your success and anxieties.

Remember: you can experience a great deal of joy and happiness from your child with PWS. Take life one day at a time!

WOULD YOU LIKE TO JOIN THE PWSA (SA)?

Please contact:

Chairperson:	chairperson@praderwilli.org.za, tel: 012 344 0241 or
Secretary:	secretary@praderwilli.org.za
Visit our website:	www.praderwilli.org.za

COST OF MEMBERSHIP

Registration fee R50.00 (once-off payment). Annual membership fee R200. R220 members outside RSA. **You are welcome to make a direct deposit into the savings account.**
Please ensure that your surname is included as reference on the deposit slip.
Please forward proof of payment to the treasurer. Fax: 012 344 0241

BANK DETAILS OF SAVINGS ACCOUNT

PRADER-WILLI SYNDROME ASSOCIATION (SA)
ABSA BROOKLYN, PRETORIA
Branch number **632005**
Acc. no. **11 364 1800**
Reference: Your **SURNAME**

MAAK 'N VERSKIL MET 'N SKENKING!

Die PWSV (SA) is geregistreer as 'n nie-winsgewende organisasie (Nr. 035-837 NPO) ook as 'n openbare weldaadsorganisaie (PBO Exemption no.930 016 853). Hierdie registrasie hou voordele in vir die donateur en donasies wat aan PWSV (SA) gemaak word is aftrekbaar van die donateur se belasbare inkomste. 'n Amptelike sertifikaat sal vir bedrae groter as R100.00 uitgereik word. Maak gerus 'n direkte inbetaling. Sluit asseblief jou van en selnommer as verwysing in.

WITH YOUR HELP WE CAN MAKE A DIFFERENCE!

PWSA (SA) is registered as a non-profit organisation (No. 035-837 NPO) as well as a public benefit organisation (PBO Exemption no.930 016 853).

The PBO registration benefits donors and all donations made to PWSA (SA) are exempt from income tax. We will issue an official certificate for donations of R100.00 or more.

You are welcome to make a direct deposit. Please ensure that your surname and cell number are included as reference.

THANK YOU TO THOSE MEMBERS FOR PROMPT PAYMENT OF MEMBERSHIP FEES.

MEMBERSHIP FEES ARE NOW DUE
1 APRIL 2013

You are invited to contribute to People with Strength

Please contribute to ***People with Strength.*** Whether you are a parent, medical practitioner, therapist or relation, please send your contributions, questions or suggestions to:

PWSA (SA), PO Box 2399, Brooklyn, 0075 or email: chairperson@praderwilli.org.za

Opinions expressed in *People With Strength* are those of the authors or editors and do not necessarily reflect the views of the management committee of the PWSA (SA).