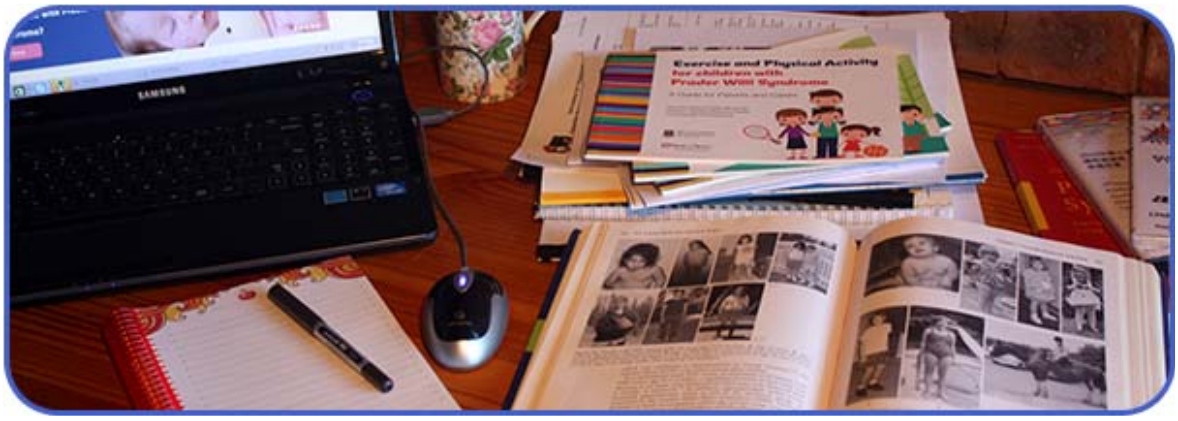
# People With Strength

Newsletter for parents by parents

Volume 17 Issue 2 **KNOWLEDGE & UNDERSTANDING** June 2014

**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

PWSA (SA) acknowledges Jochie Prinsloo for his assistance with the photo.

We trust that the information at your disposal will help you to understand this complex genetic condition. Let us always remember: behind the syndrome, behind the mass of information, is a sensitive human being, a person in his or her own right with an individual personality and a complex emotional life, who deserves to be accepted for what he or she is.
www.praderwilli.org.za

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## WHAT IS PRADER-WILLI SYNDROME?

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## FROM THE CHAIRPERSON

Dear Readers

It's winter and time for our June newsletter.

The theme of our letter is the importance of knowledge. I want to direct your attention to the list of educational articles and websites appearing in this newsletter. They cover a wide range of subjects on PWS. The child or adult with PWS is a complex individual and he or she cannot be approach according to a fixed recipe or pattern. Numerous characteristics are describe in literature which vary from individual to individual. We implore parents to gain as much knowledge as possible on PWS. We don't have all the answers, but try to trace the characteristics of your child in the information provided. You may be able, eventually, to understand him or her better or even fully. Understanding the problem may contribute to a more effective management of that problem. But keep searching for knowledge and answers. I trust that everybody will find something useful in all these articles. Sources are supplied for further reading. The PWSA (SA) will gladly supply information for those readers who don't have access to the internet.

The March 2014 edition was aimed at the family and friends who are involved in the caring of persons with PWS. I noticed an article in the newsletter of the PWSA (UK) which deals with the sibling issue again. They give good advice on how to establish positive and supporting relations between siblings and the person with PWS. Please read it.

Marietjie Jansen responded to the invitation in the March 2014 edition and wrote to us about her granddaughter, Wilmien. They went through a very difficult time in finding effective schooling for Wilmien. Marietjie stepped in and is now providing home schooling.

Good dental care is often addressed in the literature. Individuals with PWS have problems with saliva production resulting in dental cavities, loss of enamel and severe tooth wear. A few useful hints are given in the article *Enamel erosion and severe tooth wear in children and adults with Prader-Willi syndrome.* Do pay attention to this article.

The importance of exercise in the daily programme of a person with PWS is constantly highlighted. You can get your child active in a playful way without him of her experiencing it as exercise. Hints from Latham Centers' website are published in this newsletter. These exercises should become a way of life.

The life expectancy of a person with PWS is regularly under discussion. We know about two ladies in South Africa who are in their forties. A lady in Britain will be 68 this year. Read more about Shona Duncan's life and her interests.

The Famcare Committee of IPWSO submitted an article assembled by Janice Forster and Linda Thornton. Their subject is *Story-telling*. Attention is given to the common lie, wishful thinking and the fabrication of facts in the form of a story. Hints are given on the handling of this type of behaviour. The article will help parents to understand this behaviour.

Please take note of the following:

- Membership fees for 2014 are now payable.

- A friendly request: the next newsletter will appear in November. We invite you to share the joys in your life and tell us about your loved one with PWS. Parents, brothers, sisters or even a grandma or granddad can contribute to our newsletter.
- Lectures by Dr Janice Forster at the Red Cross War Memorial Children's Hospital in CAPE TOWN on FRIDAY, 3 OCTOBER 2014.
- Don't miss out on *IPWSO News*.
- The Mashilo family joined PWSA (SA) in May 2014.
- This newsletter is also electronically available in A4 format.

The next important item on the calendar is the Annual General Meeting on Sunday, 24 August 2014. Please remember this date and come and meet some other members. More information will be sent by mail or e-mail.

Warm greetings
Rika du Plooy

*Thanks to Francis Morrisson, a volunteer who did the translation*

**Any piece of knowledge I acquire today has a value at this moment exactly proportioned to my skill to deal with it. Tomorrow, when I know more, I recall that piece of knowledge and use it better.** Mark van Doren

## VAN DIE VOORSITTER

Liewe Lesers

Die winter is hier en dit is tyd vir ons Junie-nuusbrief.

Die klem val in hierdie nuusbrief op die belang van kennis. Wat ek graag onder ons lesers se aandag wil bring is 'n omvattende lys van opvoedkundige inligting. Die lys van artikels en webwerwe dek 'n wye verskeidenheid aspekte van PWS. Die kind of volwassene met PWS is so 'n komplekse wese en daar is geen vasgestelde resep of patroon wat gevolg kan word nie. Neem ook in ag dat eienskappe by persone met PWS verskil. Ek wil ouers en betrokkenes aanraai om hulself met kennis oor PWS te bemagtig. Antwoorde het ons nie altyd nie, maar soek die eienskappe van jou kind in die inligting en probeer om hom/haar werklik te verstaan. Begrip dra by tot meer effektiewe bestuur van die probleem, maar dit bly 'n voortdurende soek en groeiproses. Ek vertrou dat daar vir elkeen iets waardevols sal wees. 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.

Die Maart 2014 uitgawe was gerig op familie en vriende wat betrokke is by die persoon met PWS. In die *PWS news* van PWSA (UK), het ek 'n artikel raakgeloop wat weer eens die kwessie van die sibbe aanspreek. Baie goeie wenke word gegee oor hoe om positiewe en ondersteunende verhoudings daar te stel. Lees dit gerus.

Baie dankie aan Marietjie Jansen wat op die uitnodiging in die vorige nuusbrief gereageer het en oor haar kleindogter geskryf het. Sy vertel van die opdraende stryd wat hulle gehad het en steeds het om te sorg dat Wilmien effektiewe onderrig ontvang. Marietjie het oorgeneem en doen tuisonderrig.

In die literatuur word dikwels aandag gegee aan tandversorging. Ons is almal bewus van die taai en te kort aan speeksel by die persoon met PWS. Hierdie faktore lei tot gaatjies, verlies

aan tandemalje, ens. In die artikel *Enamel erosion and severe tooth wear in children and adults with Prader-Willi syndrome,* word handige wenke gegee. Gee gerus hieraan aandag.

Die belang van oefening in die daaglikse program van die persoon met PWS word oor en oor beklemtoon. Hoe kan jy jou kind met pret aktief kry sonder om dit soos oefening te laat voel? Wenke vanaf Latham Centers se webwerf word geplaas. Probeer dit gerus en laat dit 'n lewenswyse word.

Dikwels word navraag oor die lewensverwagting van die persoon met PWS gedoen. In Suid-Afrika weet ons van twee dames wat al hul veertigste verjaardae gevier het. In Brittanje is die oudste dame vanjaar 68 jaar. Lees gerus meer oor Shona Duncan se belangstellings.

Die Famcare komitee van IPWSO het 'n volgende artikel gereed. Hierdie artikel is saamgestel deur Janice Forster and Linda Thornton. Die artikel handel oor 'n ander aspek wat by persone met PWS voorkom, nl. *Story-Telling.* Aandag word gegee aan die gewone leuen, wensdenkery en konfabulasie of dan versinsel van feite in 'n storievorm. Wenke word ook gegee hoe om hierdie vorm van gedrag te hanteer. Die feite in hierdie artikel sal ouers beslis help om hierdie vorm van vreemde gedrag beter te verstaan. Lees gerus hierdie artikel.

Neem ook asseblief kennis van die volgende:

- Ledegeld vir 2014 nou betaalbaar
- 'n Vriendelike uitnodiging……die volgende nuusbrief verskyn in November. Skryf gerus oor julle gesin of dalk 'n spesifieke aspek van jou kind met die sindroom. Dit kan 'n ouer wees, broer of suster of dalk 'n oupa of ouma.
- Lesings deur Dr Janice Forster by die Rooikruis-kinderhospitaal, KAAPSTAD op VRYDAG 3 OKTOBER 2014.
- Moenie *IPWSO News* oorslaan nie
- Die Mashilo familie het Mei 2014 by PWSV (SA) aangesluit.
- Hierdie nuusbrief is ook elektronies in A4-formaat beskikbaar.

Die volgende belangrike gebeurtenis op die kalender van die PWSV (SA) is die Algemene Jaarvergadering. Teken asseblief Sondag 24 Augustus 2014 aan en kom kuier gesellig saam. Hou die pos of epos dop vir meer inligting.

Vriedelike groete
Rika du Plooy.

## NOTICE AND INVITATION

**THE PRADER-WILLI SYNDROME ASSOCIATION**
TAKES PLEASURE TO INVITE YOU TO THE ANNUAL GENERAL MEETING ON
SUNDAY, 24 AUGUST 2014 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 24 AUGUSTUS 2014 Vriendekring Rolbalgronde, Groenkloof, Pretoria
Inligting aan lede volg

# DIE PAD SAAM MET MY SPESIALE KLEINDOGTER

**Geskryf deur Marietjie Jansen**

*Wilmien loves knitting*

Ek deel graag die volgende aangaande my kleindogter Wilmien Dormehl wat tans 11 jaar is en op 5 jarige ouderdom met Prader-willi-sindroom gediagnoseer is. Ek is haar ouma en tuisonderrig haar sedert Graad 1. Sy het saam met haar sussie Marelie, wat twee jaar jonger is, begin skoolgaan. In hierdie tyd is Wilmien met PWS gediagnoseer en is toe in 'n klassie vir Leerders met Spesiale Onderwys Behoeftes (LSOB) geplaas, saam met twee seuntjies! Aanvanklik het dit gelyk of dit goed gaan. In haar tweede jaar op skool het sy ‘n nuwe juffrou gekry en hierdie verandering het Wilmien baie omgekrap. Sy was in trane en kon die aand nie slaap nie.Vir die eerste keer het sy bed natgemaak. Al hierdie probleme en die feit dat die LSOB klassie doodgeloop het, het veroorsaak dat ons na ‘n ander skool moes soek.

Ons soektog en met baie genade, het ons by die Bergie Dwergie skool uitgekom. Dit is die Gr RR en R klassies by laerskool Bergland - 'n departementele hoofstroomskool. Alma Ferreira was Wilmien se juffrou en het ons dilemma en frustrasie verstaan, omdat haar suster ‘n dogtertjie met Downsindroom gehad het. Die juffrou het wondere met Wilmien uitgerig en sy was twee jaar in Bergie Dwergie. Wilmien het op hierdie stadium nog glad nie gepraat nie, maar het alles verstaan wat die juffrou sê. Gereelde vergaderings is met die juffrou en die departementshoof gehou en om rede Wilmien se spesifieke probleme is sy nie op dieselfde wyse as die ander kinders ge-evalueer nie.

Die volgende stap was Graad 1 in laerskool Bergland. Hier het die prentjie heeltemal verander en het Wilmien die eerste kwartaal gedruip. Wilmien het baie van haar juffrou gehou, maar skielik was daar geen vordering nie. Hier is sy soos die res van die klas ge-evalueer, terwyl sy in graad R op haar eie prestasies ge-evalueer is. By die huis kon sy haar woordjies skryf, maar by die skool het sy net ‘n klomp letters gekrabbel. Vir my was dit duidelik dat die volgende gebeur: haar juffrou lees die woorde - Wilmien moet die inligting prosesseer en ook onthou hoe om die letters te maak. Die juffrou trek al by die derde woord dan sukkel sy nog met die eerste woord! Die gevolg was dat sy net iets op die papier gekrabbel het.

Hier het die pad doodgeloop. Op ‘n vergadering met die hoof, departementshoof en ouers is die ouers meegedeel dat die skool nie 'n inklusiewe skool is nie en het hulle aanbeveel dat Wilmien na ‘n spesiale skool – Kamagugu - moet gaan. Hierdie skool is 'n skool vir kinders met fisiese gestremdhede en is nog 'n paar kilometer verder. In Nelspruit is daar geen ander skole wat op die kind met spesiale onderwysbehoeftes gerig is nie.

Op hierdie punt het ek besluit om in te gryp en het met tuisonderrig begin. Tot ons almal se voordeel is die feit dat ek 'n gekwalifiseerde remediërende onderwyser is en Wilmien wel kon help. Dit was so bevredigend as Wilmien iets kon regkry en sê: “Yes ouma, ek kan!" Sy sukkel nog om te lees en is nou op Graad 2 vlak. Om sinne te maak is ook nog ‘n probleem. Sy het 'n mate van disleksie asook apraksie. Sy sukkel om haarself uit te druk en ontvang steeds spraakterapie.

Ons het verlede jaar aan praktiese aspekte begin aandag gee soos om te leer brei en hekel, asook naaldwerk. Wilmien geniet die handwerk en vorder goed. Sy het al blokke gebrei en na aanleiding van idees uit haar breiwerkboek, het sy vir haar ander ouma en tannie uil-deurstoppers gemaak. Sy het nou ook uit haar eie 'n serp gebrei. Wiskundige prakties word hier ingebring – Wilmien moet 'n begroting opstel van wat al die benodigdhede kos en wat sy daarvoor kan vra indien sy dit sou verkoop. Ek beoog dat sy op so 'n manier vir haarself 'n inkomste kan genereer. Dit is vir my baie hartseer as ek van ander kinders met PWS lees wat so goed aanpas in hoofstroom of spesiale skole! Ons hier in Mpumalanga het regtig 'n probleem. Die hoofde van skole stel dit ook duidelik dat hulle 'n kind nie mag weier nie en noem ook dat hulle jou kind nie kan help nie. Die kind is dus net 'n nommer, sit maar in die klas en moet hulself besig hou – dit is wat met Wilmien gebeur het!

Ons kom deur elke dag met baie Genade, Geduld en Liefde. Wilmien het ook haar "tantrum" dae, maar dikwels as ek weer teruggaan en kyk wat gebeur het, besef ek dat dit my skuld is. Ek het iets verander sonder dat ek vooraf vir Wilmien daarop voorberei het. Sy is 'n dierbare dogtertjie en wil so graag presteer! Die breiwerk vul op die oomblik hierdie behoefte!

Sterkte aan almal wat 'n kindjie met spesiale behoeftes het. Dit is nie altyd maklik nie, maar met die hulp van die Vader kom mens deur elke dag.

**PLEASE RENEW YOUR MEMBERSHIP**

Please consider renewing your membership to the Prader-Willi Syndrome Association (SA). Your financial support enables PWSA (SA) to continue to support parents and to promote knowledge and awareness of the syndrome.

PWSA (SA) would like to thank those members who already paid membership fees. Thank you for your loyalty and support.

## THE ROAD TOGETHER – WITH MY SPECIAL GRAND-DAUGHTER

**Written By Marietjie Jansen**

*Margie Deegan, a member of PWSA (SA) did the translation. We appreciate her support and the time she spent.*

I would like to share the following story about my grand-daughter, Wilmien Dormehl, who is 11 years old. At the age of 5 she was diagnosed with Prader-Willi syndrome and, after a series of "hit and misses" as far as schooling was concerned, I took on the special task of home-schooling her from Grade 1.

Wilmien and her sister, Marelie, who is two years younger, started pre-school together. During this time Wilmien was diagnosed with PWS and was then placed in a class for Learners with Special Educational Needs (LSEN), together with two little boys. Initially, it looked as though everything was going well but in her second year a new teacher was appointed – needless to say, this change upset the applecart and Wilmien could not sleep at night and cried constantly and, for the first time ever, she started bed-wetting. As a result of the negative effect on Wilmien and the fact that the LSEN class came to an end, we had no option but to look for alternative schooling.

Our search eventually led us to the *Bergie Dwergie* school, which is the RR and R classes at Bergland Primary School – a departmental mainstream school. Alma Ferriera was Wilmien's

class teacher and understood our dilemma and frustrations as her sister has a daughter with Down's syndrome. Alma worked wonders with Wilmien, who spent two years at *Bergie Dwergie*. At this stage Wilmien was still not talking but understood everything her teacher said. Regular meetings were held with her teacher and the department head and it was agreed that, due to Wilmien's specific challenges, she would not be evaluated on the same scale as the other learners.

*Marelie and Wilmien*

Then the progression to Grade 1 at Bergland Primary where the picture changed drastically and Wilmien failed her first term. Although Wilmien really liked her teacher, there suddenly seemed to be a sudden decline in progress. Whereas at *Bergie Dwergie* she was evaluated on her own progress, now she was being evaluated against the progress of the entire class. We also noticed that at home she could write down words with ease but at school it would be a jumble of letters. It was clear to me that the following was happening: The teacher would read out words for the class to write down, Wilmien would then have to process this information plus remember how to form the letters. By the time the teacher had reached the third word, Wilmien was still trying to write the first word. This resulted in her writing down whatever came into her head!!

Finally, at a meeting with the department head, the school Principal and Wilmien's parents, it was recommended to her parents that Wilmien be placed at a special school – Kamagugu – a school for children with physical disabilities some distance from home. Unfortunately, in Nelspruit, no other schooling options exist for children with special learning needs.

At this point I decided to grab the bull by the horns and start home-schooling Wilmien – the advantage being that I am a qualified remedial teacher and in a position to help her. It was incredibly rewarding when Wilmien got something right and she'd say "Yes! Ouma – I Can"!! She still has difficulty with reading at Grade 2 level and making sentences is still a challenge as she has a degree of dyslexia and apraxia. She also has difficulty expressing herself and still receives speech therapy.

Last year we started to apply practical work into her day and Wilmien began to learn to knit, crochet and sew. She loves handwork and is progressing well. She's knitted squares and has made her other Grandmother and Aunt Owl door-stoppers from ideas she got from her Knitting Book. She's also knitted a scarf for herself! We have incorporated practical maths into this handwork project by getting Wilmien to draw up a budget showing the cost of all the knitting materials and working out what she could charge for her hand-made items should she decide to sell them. I envisage that she could eventually generate an income for herself in this way.

I must admit that I do sometimes get very heart-sore when I hear about the successes that other children with PWS achieve in mainstream schooling or special schools. In Mpumalanga there is a huge problem, in that schools on the one hand may not refuse a child admission but then tell you that they cannot help your child – so it becomes a "Catch 22" situation where the child is just a number and must sit in class and amuse themselves. This is precisely what happened to Wilmien!

We get through each day with Faith, Patience and Love. Wilmien has her "tantrum" days and more often than not when I go back on the events I realise that it was actually my fault – I changed something without preparing Wilmien in advance. So, each day we both learn! Wilmien is a wonderful child and is so determined to succeed.

I wish everyone who has a child with special needs the strength to walk the road that we do with Wilmien. It is not always easy but, with His help, we manage to get through each day.

**WELCOME**

A warm welcome to the Mashilo family. They reside in MMabatho and Thatoyaone Koketso will be two years on the 4th of July.

**PEOPLE WITH STRENGTH**

All the newsletters since 2002 are loaded on the website of the PWSA (SA). Please view on www.praderwilli.org.za

## SIBLINGS OF YOUNG PEOPLE WITH PWS

**By Erin Deegan – PWSA (UK), PWS Support, Advice and Training**
**PWS news ~ Prader-Willi Syndrome Association (UK) May 2014**

*PWSA UK has worked hard over the years to increase understanding of the syndrome and recognise the needs of those with PWS. It can be so easy to forget the needs of those family members who don't have PWS. Siblings of disabled people often face difficult situations from an early age, increased responsibility and can feel socially and emotionally isolated. So how do these young people continue to have positive and supportive relationships with their siblings?*

A new diagnosis of PWS is devastating for any family. Whilst Mum, Dad, Grandma, Granddad and other relatives begin to understand the syndrome and prepare for the future, often the younger members of the family are unable to understand the complexity of the condition. It is common for them to lack the basic knowledge and information to fully understand the situations they find themselves in. So these young siblings educate themselves through experience and day to day living, developing skills and tactics to help manage the syndrome. Often these experiences can help them to become understanding, patient and compassionate adults. They are likely to develop qualities such as an increased maturity, increased tolerance of others and enhanced communication skills. Having said this, they can face many challenges along the way.

With the lack of information and knowledge about PWS, it could be easy for siblings to take typical PWS behaviour personally and have thoughts such as "my sister doesn't like me because she lost her temper with me". This can, of course, affect children's self-esteem and confidence. In turn, it could also lead to them resenting or disliking their disabled brother or sister, with thoughts such as "my brother always fights for attention until he gets what he wants". It is important that all the family have an opportunity to learn about the reasons behind these behaviours, at a time that you feel is appropriate for your individual circumstances. Explaining PWS to your children can also help to eliminate feelings of favouritism, meaning they will understand that their brother or sister simply needs more help, not that they are more loved.

They may feel a lack of parental attention and that this, along with resources and external services, are aimed towards their disabled sibling. Even when parents do all they can to share their attention out, it's inevitable that the disabled child will receive the most time and attention. With the added stress and pressure on the whole family, parental time will be stretched, making it difficult to get the right balance. Of course, time to look after yourself must also be factored in. There are several ways to tackle this problem, but first off it is important to include the child. Take them along to hospital appointments, allow them to ask questions and give them the opportunity to learn about their brother or sister's needs. You can also include them by explaining who various professionals are so they are familiar with them and understand why they are there. We can often make the assumption that a child will understand the role of a consultant or a social worker.

You can also help your child by showing that you support their feelings. This is like giving them permission to have negative feelings about their brother or sister from time to time. Telling them it is ok to be "annoyed" or "angry" can often make it easier for them to convey their feelings through words rather than actions. This can help to reduce negative behaviours in the home. The ability to express their feelings through words also makes it easier to tackle when they experience feelings of worry or concern for their brother or sister and makes supporting them through this much easier.

It is important for all members of the family, including siblings, to understand that even at times of frustration, anger and worry, there are many positive attributes that your child with PWS brings to your family. Many of our siblings are grateful to have their brother or sister in their lives as they bring happiness and joy to their family. They provide smiles and laughs and are usually loving, caring young people. One has even said that the presence of her sister made her less selfish as she can never feel sorry for herself when she thinks of what her sister goes through every day. The qualities of these young siblings, as well as the qualities your PWS child holds, are some that many of us lack.

## ENAMEL EROSION AND SEVERE TOOTH WEAR IN CHILDREN AND ADULTS WITH PRADER-WILLI SYNDROME

**Prader-Willi Syndrome Association of Wisconsin, Inc.**
**By Barbara Dorn R.N. and Dr. Kimberly Wachter, D.D.S.**
**The Gathered View ~ Prader-Willi Syndrome Association (USA) September-October 2013**

For years, it has been noted that individuals with Prader-Willi syndrome (PWS) have problems with saliva production resulting in dental caries/cavities, loss of enamel and severe tooth wear. Recent studies on "Salivary Flow and Oral Abnormalities in Prader-Willi Syndrome" as well as the study on "Severe Tooth Wear in Prader-Willi Syndrome: A Case-control Study" done by Saeves, Nordgarden, Espelid and Storhaug (2010 and 2012) from Oslo, Norway continue to support and validate these findings. The challenge now facing those with PWS is what to do to prevent and/or manage these problems. The following resource provides an overview of these dental problems along with some possible approaches.

## ENAMEL EROSION is the wearing down of the protective coating (enamel) of the teeth.

When enamel wears down, microscopic channels in the tooth open up and become exposed. Most people with this problem complain of discomfort and pain. However, since persons with PWS have an altered pain response, they may not experience this sensation.

**Common Causes:**

- Abnormal salivation
- Diet high in sugars and acid

## EXAMPLES OF FOODS AND BEVERAGES WITH HIGH ACIDITY:

Soft drinks, sport drinks, fruit juices, lemonade, coffee, tomatoes, strawberries. (Citric acid, phosphoric acid, and ascorbic acid are a few ingredients to watch for in foods.)

- Grinding of the surfaces of teeth
- Health conditions including gastric reflux
- Medications (aspirin, antihistamines and some vitamins)

**Signs:**

- Translucency of the enamel
- Darkening or discoloration of the teeth
- Pain or cold sensitivity (may not always be reported in persons with PWS)

**Prevention:**

- Drink water. Limit soft drinks, sport drinks, juices and coffee in moderation. Dilute juices with water.
- Swish your mouth out with water or brush your teeth after drinking or eating items high in acid
- Don't sip on beverages for extended periods of time.
- Do not drink or eat high acid beverages/food in the evening after supper.
- Use a straw to keep acids away from teeth.
- Brush with fluoride toothpaste. Minimize swallowing. (Too much fluoride can cause problems like enamel fluorosis. This condition can occur in children and causes defects in the enamel of the teeth. Ingesting large amounts of fluoride can be toxic and cause nausea, stomach pain and/or vomiting. It may not be quickly detected in persons with PWS).
- Get regular dental checkups and cleanings – at least every 6 months.
- Talk to your dentist about daily fluoride mouthwash if there is a history of cavities (Use as a rinse; do not swallow). If needed – apply with Q-tip.
- Ask the dentist if sealants may be helpful in preventing enamel erosion and tooth decay.

**Treatment:**

- Once lost, enamel cannot be replaced. Treatment of tooth enamel loss depends on the underlying problem.
- If a person with PWS is found to have a sudden loss of tooth enamel, testing should be done to determine if a medical condition is present. (May require special diet and medication intervention).

## SEVERE TOOTH WEAR / GRINDING (also called bruxism)

Occurs most commonly at night. When teeth grinding occurs on a regular basis, teeth can be damaged and other oral complications can arise - often seen in both children and adults with PWS.

**Causes:**

- Exact cause is unknown.
- Often contributed to stress and/or abnormal bite

**Treatment:**

- Mouth bite guard – best if individually fitted by a dentist. Used most commonly while the person sleeps. May require incentive program for compliance.
- Avoid or cut back on foods and drinks that contain caffeine, such as colas, chocolate, and coffee.
- Avoid alcohol. Grinding tends to intensify after alcohol consumption.
- Discourage chewing on pencils or pens or anything that is not food. Minimize chewing gum. It allows jaw muscles to get used to clenching and makes it more likely to grind teeth.
- Work with person with PWS to learn not to clench or grind his/her teeth. Have him/her practice relaxing jaw muscles.
- Try holding a warm washcloth against the cheek in front of the earlobe.
- Include plenty of water. Dehydration may be linked to teeth grinding.

## EMPOWERING PARENTS

**Taken from the website: www.empoweringparents.com**

"You don't have to attend every fight you're invited to."

What Does **My Child** Need From Me **Right Now?**

There is always **HOPE** for your child But hope without a **PLAN** is fruitless.

BE A CALM PARENT. **Create a Space Between Your child's action and Your Reaction.**

## EDUCATIONAL MATERIAL FOR PARENTS, CAREGIVERS AND OTHERS

Undiagnosed and unmanaged, Prader-Willi Syndrome causes morbid obesity. Empower yourself and learn more about treatment and management strategies. Let's improve the quality of life of all who are impacted by PWS.

### FROM THE PEN OF FAMCARE Famcare is a project of IPWSO: famcare@ipwso.org

SKIN PICKING IN PEOPLE WITH PRADER-WILLI SYNDROME – MARCH 2013
COPING WITH CHANGE IN PEOPLE WITH PRADER-WILLI SYNDROME – June 2013
THE BASICS OF A HEALTHY ADULT LIFE – Nov 2013
"GOOD HEALTH" CHECKLIST – March 2014d
STORY-TELLING – June 2014
Articles are available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

### FIRST STEPS PARENT PACKAGE WWW.FPWR.ORG

You may have recently received a diagnosis of Prader-Willi syndrome for your child or loved one, or maybe you are just finding us on your journey, and are ready to reach out for guidance. Please join us in taking your "First Steps" following a diagnosis! You are likely feeling overwhelmed, and worried about what your child's future might hold. This is completely normal, and it will get better.
The Foundation of Prader-Willi Research (FPWR) invites you to contact them. By sharing some basic information, you will be granted access to a wealth of current, informative documents, as well as photos and videos of children with PWS.
Download your FIRST STEPS PACKAGE from www.fpwr.org

### DVD: FOOD, BEHAVIOUR AND BEYOND

**Pittsburgh Partnership and PWSA (USA)**

This comprehensive DVD 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.
**Please contact Wilna Basson** at 012 991 3399 or e-mail: bassons@iafrica.com

### BIRTHDAY CARDS

We would like to thank Magdaleen Kloppers and Karin Clarke, the friendly ladies who are responsible for the 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 card. A friendly request – please acknowledge receipt by SMS either to Karin or Magdaleen.
**Karin (English cards): 083 681 6842**
**Magdaleen (Afrikaans cards): 083 663 7234**

## DVD: UNDERSTANDING THE STUDENT WITH PRADER-WILLI SYNDROME

### PWSA (USA)

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

## PRADER-WILLI SYNDROME TEACHER TIPS ON YOUTUBE

### http://www.youtube.com/watch?v=Y5LhSePDvqk

A PWSA (USA) video with Elizabeth Roof presenting **Tips for Teachers** is available on YouTube. Elizabeth Roof is a M.A. - Senior Research Specialist at the Vanderbilt Kennedy Center, USA. This fantastic resource provides very helpful information and practical strategies for teachers who have students with PWS in their classroom. It has two parts:
1) The Video and 2) A related Tool Box for Teachers
We encourage you to send the video link and the attached Tool Box to your child's teacher and please let other families you know in the PWS community about this amazing new resource. We want as many teachers as possible to use the video and tool box to better understand how to effectively support students with PWS.

## 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

## FOOD SECURITY – BASIC CONCEPTS

### Pittsburgh Partnership, Specialists in PWS

Successful Behaviour Management of PWS means that uncertainty about food must be eliminated as much as possible. The planning and availability of meals must be discussed

with the person with PWS in order to diminish any uncertainty and stress. The basic concepts *Pittsburgh Partnership* uses to ensure success in maintaining both weight and behavioural control are ***no doubt, no hope*** and ***no disappointment****. C*heck lists for the family and workplace are given to assist and guide you in the correct procedure of FOOD SECURITY.
Available from www.pittsburghpartnership.com
*People With Strength* March 2012
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## 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.
**PWSA (SA) members: contact Wilna Basson** at 012 991 3399 or e-mail: bassons@iafrica.com

## LOCKING THE CABINETS AND REFRIGERATOR FOR PRADER-WILLI SYNDROME

### By Allen Heinemann

One of the questions that is often asked as the child with Prader-Willi syndrome gets older is, "How can we keep our child from sneaking into the refrigerator or cupboards and getting at food?" It's hard enough to have to keep food out of sight and put away, but then to have to constantly keep one eye toward the kitchen to catch someone before they get into food can be very stressful. It becomes necessary for reducing everyone's stress level and to have the food locked up. Allen Heinemann discusses common solutions.
Available at: www.pwsausa.org
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## BEYOND THE DIET – FOR THE DIETICIAN

### By Janalee Heinemann - Prader-Willi Syndrome Association (USA)

Important realities for a dietician to consider are given: the food-foraging drive; how to lock food away; how many calories? Advice is given on educating the extended family and community, as well as the support to parents. Dietician's help, support, and compassion are needed to make the long road to weight control a little easier.
Available at: www.pwsausa.org
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## DIFFERENCES BETWEEN THE GENETIC SUBTYPES OF PRADER-WILLI SYNDROME

### By Dr Suzanne Cassidy

Articles on genetics are usually not read because they are too long and complicated. Don't skip Suzanne Cassidy's article on the different genetic subtypes found in PWS. She explained why different genetic changes cause PWS. A deletion, maternal uniparental disomy and an imprinting defect are also explained. It is indeed interesting and readable. Much is still to be learnt about genetics and PWS.

*People With Strength* November 2012
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## 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

## GROWTH HORMONE FOR ADULTS WITH PRADER-WILLI SYNDROME

**By Barbara Y. Whitman, Ph.D**

The benefits of growth hormone (GH) replacement therapy for infants and children with Prader-Willi syndrome in the USA are discussed.
Dr Whitman joined with several other teams of researchers and conducted a study of GH for 40 adults with PWS, ranging in age from 19 to mid to late 40s. This study and another conducted in Sweden are now completed and results are being published. The procedures, pitfalls, risks and benefits are discussed. Website: www.pwsausa.org (Medical View)
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## CHARACTERIZING AND MANAGING BEHAVIOUR IN PRADER-WILLI SYNDROME

**Kevin Jackson. Ph.D., CBA. ARC of Alachua County, Gainesville, Florida**

Kevin Jackson wrote: "This series of articles is based on my training as behaviour analyst and on my thirteen years of experience in working with children and adults with PWS in family settings, schools, and as director of behavioural services for a large residential treatment program specialising in PWS. The concept and interventions discussed here have been extremely effective in addressing the behaviour of individuals with Prader-Willi syndrome.
Website: www.pwsausa.org

## FINDING PSYCHIATRIC HELP FOR YOUR CHILD

**Prepared by the *Pittsburgh Partnership*, Specialists in PWS**

This article was prepared to provide guidance to parents and other care providers in seeking out psychiatric services. Parents are guided how to choose a psychiatrist for their child. Important requirements are given like what to look for in a consultant and how to prepare for the consultation in order to make the most out of the time.. It is not necessary for the treating psychiatrist to have experience with Prader-Willi syndrome. The role off the psychiatrist is also clearly outlined. Available from www.pittsburghpartnership.com
*People With Strength* June 2013
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## MANAGING PRADER-WILLI SYNDROME: A PRIMER FOR PSYCHIATRISTS

**Prepared by the *Pittsburgh Partnership*, Specialists in PWS**

This guidance is offered for psychiatrists and psychologists who are facing the challenge of treating a very complex patient with an unfamiliar condition, Prader-Willie syndrome. The material here is based on our clinical experience of inpatient and outpatient crisis intervention with hundreds of cases of persons with PWS of all ages.
This comprehensive 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
It is also available from chairperson@praderwilli.org.za

## PSYCHIATRIC ALERT FOR PSYCHIATRISTS ON PRADER-WILLI SYNDROME

**Prepared by the *Pittsburgh Partnership*, Specialists in PWS**

Many families caring for children and adults with PWS will seek psychiatric treatment for problems such as mood liability, tantrums, skin picking and repetitive behaviours. Although the psychiatrist is usually asked to prescribe psychotropic medication, the psychiatrist's role as a consultant to the treatment team is equally important. The multidisciplinary team may include behavioural, educational, residential, and occupational specialists. Most psychiatrists will not have treated more than one or two cases with PWS. More important than previous experience is a willingness to learn about the clinical features and management of PWS. Clinical experience with other developmental disabilities is helpful, but it is important to know that all persons with PWS display impaired judgment regardless of IQ and verbal language skills. A concise summary of things the psychiatrist needs to know before prescribing medications are also given.
Available at: www.pittsburghpartnership.com.
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## PSYCHOTROPIC MEDICATION TIP SHEET FOR PATIENTS WITH PWS FOR HEALTH CARE PROVIDERS

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

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
*People With Strength* March 2012
Also available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## INFORMATION FOR SCHOOL STAFF: SUPPORTING THE STUDENT WHO HAS PWS

**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

## SPEECH AND LANGUAGE DISORDERS ASSOCIATED WITH PRADER-WILLI SYNDROME

Excerpted from: Management of Prader-Willi Syndrome (Third Edition), Chapter 9.
Barbara Lewis
Available only via email from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## TO: LAW ENFORCEMENT PERSONNEL REGARDING CHILDREN WITH PRADER-WILLI SYNDROME. PWSA (USA)

In this article Prader-Willi syndrome (PWS) is being explained to the law enforcement person involved. Because of the unique behavioural and emotional characteristics related to this rare syndrome it is imperative that you read the following information in order to better understand the situation you've encountered. PWS is a genetic disorder that causes the brain to function in a way that is not typical of other children. This brain dysfunction can cause problems that often lead to the involvement of law enforcement.
*People With Strength* November 2012
Available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## TO: LAW ENFORCEMENT PERSONNEL REGARDING ADULTS WITH PRADER-WILLI SYNDROME. PWSA (USA)

Behavioural instability can be a feature of PWS. However there are other unusual behaviours that can be the cause of police attention. These behaviours, while actively discouraged by parents and caregivers, should also be treated as a manifestation of the person's disability rather than a criminal act. Appropriate responses include working with the parent(s) and or caregivers to develop positive behavioural solutions in each situation to discourage repeat incidents in the future.
While each person with PWS is unique, tips are given to help to create a more successful response to a person with PWS who is in need of police assistance.
*People With Strength* November 2012
Available from **the chairperson PWSA (SA):** chairperson@praderwilli.org.za

## BULLYING AND CHILDREN AND YOUTH WITH DISABILITIES AND SPECIAL HEALTH NEEDS

Bullying is unwanted, aggressive behaviour among school aged children. Children with disabilities or other special health needs may be at higher risk of being bullied. There are specific ways you can support these groups. Children with autism spectrum disorders are at increased risk of being bullied and left out by pears. What parents can do is also addressed. (From an American perspective).
www.stopbullying.gov (Considerations for Specific Groups)

## 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**

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. *This article is posted on the medical section at* www.pwsausa.org.
*People With Strength* June 2013
Also available from chairperson@praderwilli.org.za.

## USEFUL WEBSITES

| | |
|---|---|
| Prader-Willi Syndrome Association UK | www.pwsa.co.uk |
| Prader-Willi Syndrome Association USA | www.pwsausa.org |
| International Prader-Willi Syndrome Organisation | www.ipwso.org |
| The Foundation for Prader-Willi Research (FPWR) | www.fpwr.org |

**THOSE IN THE EASTERN and WESTERN CAPE - PLEASE DIARIES THIS DATE**
Dr Janice Forster, who is part of the Pittsburgh Partnership (USA), specialists in Prader-Willi Syndrome will visit Cape Town on Friday 3 October 2014. Dr Forster will address professionals as well as parents at the Red Cross Children's Hospital, Cape Town.
If you are interested in these special lectures and want to receive more detailed information, please contact: the chairperson of the Prader-Willi Syndrome Association (SA): chairperson@praderwilli.org.za

## TIP OF THE WEEK: SNEAKING IN EXERCISE

**Website of Latham Centres - www.lathamcenters.org look for *Tip of the Week***

One of the best ways to get your child to exercise is to not call it exercise! Getting active and having fun together doesn't have to be a chore and once the habit of being active is formed it will be hard to break. Here are some ideas for exercise that you can sneak in without anyone knowing it's good for them!

1. Have an outdoor scavenger hunt. This can be on a nature walk, the beach or just during a neighbourhood walk. Kids will be more likely to keep moving if they are distracted with the hunt.

2. Get competitive. We have had great success with the use of pedometers. The person with the most steps at the end of the week is the winner. Their competitive nature kicks in and over time we found that even the prize for the winner was unnecessary. They simply wanted to be the winner.

3. Go to a playground. Most towns have community playgrounds and they are highly underused. Don't discount the exercise value of climbing, swinging and many of the upper body building equipment found on playgrounds.

4. Impromptu games of catch and tag are wonderful ways to get exercise in and have fun at the same time.

5. Anything that can be turned into a contest; who can get to the mailbox the fastest, who can throw the farthest, who can carry the most books. Literally anything that has a winner will be met with enthusiasm.

6. Walk for charity. Our kids have such sweet spirits and are typically very willing to help out someone in need. During the nice weather there is some kind of charity walk happening almost weekly.

Whatever you choose to do, be sure that you are equally as involved and excited. Your child will model after your attitude towards being active. Standing with a stop watch and telling

them to get their exercise in for the day will last about 5 minutes, if you're lucky. Exercise can be fun, but shh…., don't call it exercise!

Patrice Carroll - Manager of PWS Services

## IPWSO NEWS

The International Prader-Willi Syndrome Association (IPWSO) is a non-profit organisation, founded in 1991, whose mission is to raise the quality of life for people with Prader-Willi syndrome and their families all over the world. They encourage families and professionals to join together to help one another. www.ipwso.org

**NEW:**
**For professionals and scientists**

A new publication from Nova Biomedical, entitled "Prader-Willi Syndrome - Congenital Disorders; Laboratory and Clinical Research", edited by Charlotte Hoybye, is now available via the website of IPWSO www.ipwso.org. For a short time there is a 20% reduction when ordering this book. US$140

You can view a chapter from the book, "Prader-Willi Syndrome: The First 50 Years" by Martin Ritzen, and find a list of contributors, at IPWSO's website.
This is an excellent resource book in 6 sections with many contributors.
The sections cover:

1. Introduction to PWS
2. Genetics in PWS
3. The Clinical Picture of PWS
4. Specific Issues in PWS
5. Specific Treatment Strategies in PWS
6. Patients and Parents' Supporting Organisations

A list of the 37 contributors, including contact details, is listed in the book, thus giving interested readers contact with the specific expert.
*IPWSO's scientific advisory board is available for your questions and answers via our email:* information@ipwso.org *and already has helped with enquiries ranging from specific drug treatment and side-effects, to gastric issues in PWS. All ten of our experts from around the world give freely of their time to help. We are always grateful for their input.*

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

Read about a mother who had a bad PWS appointment experience. So, she decided to make a short list of some of the many anonymous people to whom she is grateful for making her family's PWS journey so much easier. Visit the blog and read her list.
You are welcome to communicate directly with the Communications Director of IPWSO, Linda Thornton. She will welcome your comments. http://ipwso.blogspot.com

## 9TH INTERNATIONAL PWS CONFERENCE, 2016

The next international conference will be held in Toronto, Canada in 2016.

## STORY TELLING IN PWS

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

*Many thanks to Janice Forster and Linda Thornton for all they have contributed to this article.*

**A parent writes:**

The situation arose over the then-imminent wedding of a fellow participant in the exercise class for whom Gemma had developed a strong affection. She had listened to the evolving discussions in the class about this lady's plans to marry, from concept to detail and had over some months articulated to me the hope that she would be invited to the wedding. As it happened, no-one from the exercise class was invited. Plans firmed for a venue, menus were chosen, and some intense fitness work was happening so she could 'fit into her wedding dress and be looking great' for the big event. We put Gemma's constant repetitive talk about the wedding progress and hopes for an invitation down to the frequently-demonstrated perseveration associated with PWS, probably not understanding how intense this particular wish was. The bride-to-be also kindly took Gemma out for lunch one day, taking time off work to do so. More such catch-ups were promised, though none had occurred. (and still haven't).

One afternoon after a morning exercise class, Gemma came to me saying that she had just remembered Jane, the exercise therapist, had spoken to her that morning reminding Gemma not to forget to RSVP to the bride about her invitations. Gemma clarified that the bride wanted to pick her up from work (at Secret Garden Nursery) one day the following week and take her out for dinner, leaving her partner at home to look after their daughter (an unusual event, apparently). Gemma also was invited to the wedding, and to travel down with the bride, partner and daughter on the Friday preceding the wedding, stay overnight with them, and they would bring her back home on the Sunday after the wedding, if that was all OK.

Gemma then sent an email to the bride accepting these kind invitations. Later that night I received a phone call from the bride saying that she was a little concerned because the discussions/invitations had never happened. I also spoke to the receptionist, whom Gemma was certain had overheard the conversation and had supposedly reminded her not to forget to reply. The receptionist assured me that neither conversation had happened. Both the exercise therapist and bride expressed their affection for Gemma, but were concerned for her well-being.

Gemma was so certain the conversations had happened, clearly 'remembering' every word, expression and her excitement at what had been said. It was this 'clarity' that made it so hard for her to accept that it didn't happen and caused her to wonder if she was going mad. I am not sure the psychologist and (subsequently) psychiatrist that we visited were able explain to her satisfaction why it had happened, and to this day I think she still occasionally wonders if it did. She worries less about her sanity these days because there have been no similar episodes since, but it took a while.

Story telling is common for individuals with PWS, especially among higher functioning people like Gemma. Not all story telling results in such complex stories. There are several variations of this behaviour in PWS.

First, there is the simple lie. Typical children lie to escape punishment or to avoid doing something they don't like doing or don't want to do. Lying in PWS is often about food. For

example, a young person with PWS might tell his teacher that his mother did not feed him breakfast, so that he will get another one at school. Also, people with PWS of all ages are likely to lie when confronted with theft of food or other items, even if they are caught in the act. The act of lying implies that a person is telling a falsehood; that is, they know that what they are saying is untrue. Sometimes a person with PWS will make a statement that you know is false, but they believe that it is true at that point in time. For example, a young woman steals a CD from a peer at the work program. When confronted by her mother, she says that she did not steal the CD because it was hers. The young woman argues with certainty that the CD is hers, until her mother finds the same CD in the young woman's collection.

The second form of storytelling is *wishful thinking*. This is the type of story that Gemma told. She wanted to be included in her friend's wedding. Her story is a fantasy, but she believed that it was true. She did not have the capacity to test the reality of this story. She was not able to see the content from a perspective other than her own. She was emotionally invested in it, and she wanted it to be true.

The third form of storytelling is *confabulation*. Confabulation is defined by dictionaries as*: the production of fabricated, distorted or misinterpreted memories about oneself or the world, without the conscious intention to deceive* or *the replacement of a gap in a person's memory by making-up stories that he or she believes to be true.* In addition to faulty memory, there is another form of confabulation which is similar to the puzzle assembly abilities among individuals with PWS. In this form, pieces of information that the person has heard are put together into a story. The information can come from conversations, news releases or movies. Because the person is assembling this information into a story, they believe that it is true.

Confabulation is different from lying because the person is making up and telling stories that he or she genuinely believes to be true. In turn, the person believes that you, the listener, should also believe that what they are saying is true. The person tells the story in a clear and consistent manner, and the content of the story is believable in that it *could* actually have happened. Although the story itself is false, some of the information contained within the story is true, and this is what makes the story believable. The story is put together from facts, memories, or what the person has heard, read or seen. Confabulation can range from the subtle changing of a story, to a quite bizarre invention.
The details in these stories may be contradictory, but suggesting this to the story-teller can cause problems. The story-teller will object to your contradictions no matter how clearly you present them, and if an argument ensues, you will not win. Also, the story-teller can become very upset, anxious, and often becomes the 'victim' so that the listener 'can no longer be trusted'; is no longer their friend; never believes them; is calling them a liar' and so on.

Some of these stories can lead to more difficult situations, especially when the content of the story involves physical or sexual abuse or emergency calls. When the story-teller is so convinced they are telling the truth and they are believed, higher authority intervention occurs, involving the police or emergency medical personnel. For example: a boy took his parents cell phone and called police early one morning saying that he had awakened to find that everyone in his family had been killed by an intruder; he had escaped by hiding in the bathroom, but the intruder was still in the house. When his mother woke-up in the morning and opened the front door to pick up the paper, she was greeted by the police emergency team! Another example is a young man who claimed that he had been sexually molested.

There was no evidence of abuse (and there may not be even in real cases of sexual abuse) and the timing of the event in the story was impossible, but the young man was so insistent and believable, the accused staff person lost his job anyway.

Sometimes, it is difficult to decide if a story is confabulation or delusion. Confabulations are usually consistently and accurately repeated within a given context, but they might change from listener to listener. Over time, the person may admit that the story was untrue or deny ever telling it. Delusions are fixed false beliefs related to psychosis; they do not change over time unless the underlying condition is treated. Examples include: a psychotic man who believed that he was being controlled by a she-devil who was pinching him repeatedly; a boy who believed that the characters in a TV show or video game were real, and he could converse and interact with them. Some stories have grandiose content such as a middle aged woman saying that she had three sets of triplets, or a young man who stated that he was a championship ballroom dancer. Grandiose delusions are associated with mood elevation and usually resolve when the mood disorder is appropriately treated with medication.

Other stories revolve around religious themes or follow the death of a loved one. Stories of seeing angels or having conversations with loved ones who have died are not considered to be pathological. Usually these stories bring comfort to the grieving person.

***How do you investigate the "event" when your person with PWS tells you?***

When confabulation indicates abuse, neglect or danger, it must be investigated. The first step is to make sure that he/she is safe. Second, if at all possible, you should take in all the information and react with neutral emotion. How these stories are managed is critical to prevent them from being told again; situations involving police and emergency medical staff are usually highly reinforcing to people with PWS. Make note of the time frame of the story; there may be inconsistencies that indicate doubt. Supporting evidence is essential. Don't lead the person on by asking questions or suggesting additional information. For example, if the person says, "My Daddy touched me last night." Ask, "How did he touch you?" or "Where did he touch you?" Don't ask, "Did he touch you between the legs?" No matter how outrageous the story is, always consider the possibility that the person is telling the truth!

- Pay attention to the details of the story and write them down.
- Have someone else ask the person with PWS how their day is going. If he/she has had a traumatic event, they are more likely to tell the same story the second time to a different person.
- Most people who have had a traumatic event will show emotional distress; the story teller may appear to be detached.
- Alert the parent or guardian about the situation.
- Proceed with an evaluation by police or medical personnel, if indicated.

***Why do people with PWS make up stories?***

The precise reason why people with PWS tell stories has not been scientifically investigated, but clinicians and parents alike, have some theories about why it occurs. Some scientists believe that story telling is a developmental phenomena, and people with PWS remain more child-like in their thinking and behaviour. Others believe that the short term memory problems typical of people with PWS cause them to make up details and interfere with their ability to test the reality of those details.

One aspect of the way people with PWS think (*a cognitive trait)* causes them to see things only from their own point of view. They *can* accept another person's viewpoint, but *not at the same time* that they are expressing their point of view, especially if they are emotional invested in the story. At a later date, they might admit that the story they told is untrue or say they never told it. But when they are telling a story, they really do believe that it is true. That's why they are so convincing and it is so difficult for them to admit that the story is made-up. Another cognitive trait is their ability to take little pieces of data and create a whole picture - like putting a puzzle together. They can overhear a conversation, get tiny bits of information and make quite a convincing story. The story is most effective if there is an audience. Usually there is a lot of attention directed toward the content of the story and the person who tells it. Attention encourages the story-telling because of the interest it is given, even though it is false.

Sometimes the person with PWS has something to gain from telling the story. A person with low self esteem might tell a story to sound important. Or, a person with PWS might tell a story to access food or money or just attention. Sometimes stories cover up responsibility or guilt, like the man who was charged for breaking and entering a service station and stealing chocolate. He told police that the window was left open, and he thought someone might be inside committing a burglary, so he climbed in to investigate. That's when he saw the chocolate and took it.

***What is the best way to help the person overcome their tendency to 'spin' the truth?***

Janice Forster MD, (Developmental Neuropsychiatrist of the Pittsburgh Partnership and advisor to FAMCARE and IPWSO) suggests the following strategies:

First, in order to understand the nature of the problem, ask these questions:

- Is this pattern of behaviour typical of the person with PWS?
- Is the person high functioning?
- Does the person have a lot of freedom in his/her life?
- Has the person been diagnosed with bipolar disorder or is he/she receiving any medications that might cause grandiosity or mood activation?
- Has the person been diagnosed with a psychotic disorder?
- Are the parents the legal guardians, or does the person consent for him/herself?

***Managing this behaviour is challenging - here are some helpful tips:***

1. **Reduce his/her degree of freedom.** He/she may have access to too many people and too much information. This may enable him/her in a negative way. If the information for the stories is coming from the computer, leisure time can be limited or supervised.
2. **Take away the audience.** This means that you have to alert every one that he/she comes in contact with, that he/she is a storyteller. They should listen but always react in a neutral way. Although the truth should be doubted, there is always the possibility that a story could be real. Then, try to redirect the person to a safe topic that everyone knows is true, like "Tell me about your sports card collection" or "Tell me about the first time that you went to a football game?" Or, if in a day program or school environment, teach staff an intervention "That's an interesting story, but I'll have to check with your parent," or, "It must make you feel important to think that you're related to a sports star." It's not helpful to ignore, and it's not helpful to attempt to dissuade. Helping him or her to save face is important.
3. **Turn the content of the story into an asset**. If he wants to be a sports star, or she wants to be a film star, then they need to live the life-style, including weight loss and exercise.

(Special Olympics can be a wonderful way to provide an appropriate level of competition, together with appropriate exercise.)

4. **Use social stories.** Social stories are a helpful tool for teaching morality like why it's a good thing to always tell the truth and why there are consequences for lying. Most confabulators are creative people, so maybe they can write some social stories with you.
5. **Alert the police.** Be proactive with the police. Tell them that story telling is part of your child's repertoire. Give them your telephone number so they can call you if they become involved. If your son or daughter already has experienced the police, I would strongly advise you to obtain guardianship, or advocate for them. As intelligent as they may appear, they will not be able to advocate for themselves, although the court may find them competent.
6. **Consider therapy**. If your person with PWS is high functioning, he/she may be dealing with feelings of inadequacy due to having PWS, so their stories are all about being someone else who doesn't have the syndrome.
7. **Adjust medications** if they are contributing to the situation.
8. **Punishment never works.** When a person with PWS truly believes their thoughts are based on reality we must remember to be aware of their sensitivity to "blame." Overreacting to their stories, ridiculing their confabulation or blaming them for "causing trouble" will not help the situation, no matter how embarrassing or far reaching the implications of the story have become.
9. Remaining calm and supportive while providing the person with PWS the security they need to express his/her thoughts, then gently redirecting the conversation allows them to be heard without enhancement.

## DID YOU KNOW? The oldest lady in the Britain is 68 years

### A summary from Hi to Shona Duncan – the oldest lady in Britain with PWS? PWS news ~ Prader-Willi Syndrome Association (UK) May 2014

Hi, my name is Shona Duncan. I will be 68 years old this year and I've been told that I may be the oldest person in Britain with PWS. I live in a small residential home in Edinburgh where I am the only resident. Before coming here I lived with my mum, but at the age of 50 I thought it was time I left home and this care home was the obvious choice for me as I had been coming here for respite weekends for many years. It's also near my mums and I visit her regularly.

Over the years I've had many opportunities to try new things and learn new skills. I have also been able to do the things I enjoy most - being involved with animals and craft work!

I have a person centred plan that myself and my key worker review annually and set out goals for the coming year. One of my goals this year is to knit dog coats to order and the money I raise from these I donate to the Dogs Trust.

I like coming to the centre five days a week (Longstone Resource Centre). I get to socialise with my friends and enjoy trips out and about, sometimes in the local community and sometimes day trips further afield. My weight is not much of an issue anymore as I am well supported both at home and in the resource centre.

Despite using a 'walker' to give me confidence with my mobility, I manage to enjoy a full and active life. I love music and I have a vast collection of DVD's and CD's. I love Elvis and Cliff but

my absolute favourite is Daniel O'Donnell – I am his number one fan and I go to see his shows every time he comes to Edinburgh.

## ARTICLE ON PWS IN TOP 100

### The Gathered View ~ Prader-Willi Syndrome Association (USA) September-October 2013

A review in the journal *Pediatrics, Official Journal of the American Academy of Pediatrics,* identified and analyzed the top 100 most frequently cited articles published in journals dedicated to pediatrics between 1945 and 2010. An article about Prader-Willi syndrome was 53rd on that list!

Although PWS was first described by Swiss doctors Prader, Willi and Labhart in 1956, there were no criteria for a diagnosis until 1993. That year a gathering of professionals particularly involved with PWS gathered for a consensus conference at the Western Look resort in Tucson, Arizona. PWSA USA provided a grant to support this effort.

From the conference came the article: Prader-Willi syndrome—consensus diagnostic-criteria. *Pediatrics.* 1993; 91(2):398–402.

Participating in that conference and authoring the article were Dr. Vanja Holm, Dr. Suzanne Cassidy, Dr. Merlin Butler, Dr. Jeanne Hanchett, Louise Greenswag, Ph.D., and Barbara Whitman, Ph.D. All of the authors are—or were—members of PWSA (USA) Advisory Boards.

This is a tremendous honour, given that this article made the top 100 out of over 450,000 articles published in 191 pediatric journals between 1945 and 2010.

## THE PRADER-WILLI SYNDROME ASSOCIATION OF SOUTH AFRICA

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

*We invite everyone involved with persons with Prader-Willi syndrome, including parents, family members, friends, professionals, caregivers and other interested persons, to become members of the association.*

### 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 chairperson.
chairperson@praderwilli.org.za or 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: SURNAME, please

## PLEASE CONTRIBUTE AND HELP TO MAKE A DIFFERENCE!

Your donation, large or small, enables PWSA (SA) to continue to support parents and to promote knowledge and awareness of the syndrome.
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).
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.

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

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

## NEW: EDUCATIONAL PACK FOR TEACHERS

Prader-Willi Syndrome Association (UK)

A very comprehensive summary of the different aspects the teacher will come across when confronted with a child with PWS in his/her class. Useful tips are given to teachers. Parents with young children are advised to provide this info to their child's teacher as soon as they start nursery or primary school. Contact Erin Deegan at EDeegan@pwsa.co.uk
Also available from the chairperson@praderwilli.org.za