People With Strength

Newsletter for parents by parents

Volume 18 Issue 2 **CELEBRATING 25 YEARS** June 2015

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 (USA) is celebrating 40 years

The PWS community of South Africa would like to congratulate PWSA (USA) with 40 years of dedicated service to families and people with Prader-Willi syndrome. Since 1983, PWSA (USA) has hosted a National Conference. The 2015 PWSA (USA) National Conference will be held in Orlando, Florida from 4 – 7 November. It will be the largest conference concerning Prader-Willi syndrome in the world! We thank you for your passion to save and transforming the lives of those with PWS. Our best wishes for the road ahead.

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WHAT IS PRADER-WILLI SYNDROME?

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- Floppy baby (low muscle tone)
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Chairperson: Rika du Plooy rikadup@mweb.co.za 012 344 0241
Medical Advisor: Dr Engela Honey engela.honey@up.ac.za 012 319 2269

DIE VOORSITTER

Liewe Lesers

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

Graag wil ek hierdie skrywe begin met ‘n gelukwense aan die *PWSA (USA)* wat vanjaar hulle 40ste bestaansjaar vier. In 1975 is die *Prader-Willi Syndrome Parents and Friends* deur ‘n ouerpaar wie se seun met PWS gediagnoseer is, begin. Nie lank daarna nie, is dit hernoem na die *Prader-Willi Syndrome Association.* Oor 40 jaar was hierdie organisasie getrou aan hulle missie van **Saving and Transforming Lives.** Dit kon hulle bereik deur die toegewyde samewerking tussen die personeel van *PWSA (USA)*, professionele vrywilligers en ouers. Baie dankie dat die PWS gemeenskap oor die wêreld heen kon baat by wat reeds bereik is en nog verder gaan baat by dit wat *PWSA (USA)* vir die toekoms beplan.

Ontmoet Larike Grix wat in Mei haar tweede verjaarsdag gevier het. ‘n Vroeë diagnose en baie navorsing deur die ouers is tot voordeel van Larike. Dit is ‘n positiewe storie oor ouers, wat saam met bekwame professionele persone hierdie dogtertjie help om haar volle potensiaal te bereik. Lesers is baie welkom om met Liezl en Quintin kontak te maak. Eposadres: liezltruter1@gmail.com

Wat is katapleksie? In die artikel oor Larike Grix vertel hulle dat sy op 20 maande met katapleksie gediagnoseer is. Lees die verduideliking oor narkolepsie en katapleksie elders in die nuusbrief.

Vanaf die nuusbrief en webwerf van *PWSA (USA)* is daar interessante inligting oor navorsing asook praktiese wenke. ‘n Waarskuwing word aan ouers en versorgers in *The Gathered View* (June 2015) gerig om asseblief waaksaam te wees wanneer hardlywigheid by die persoon met PWS voorkom. ‘n Baie volledige artikel verskyn op die blog van IPWSO wat dieselfde onderwerp aanspreek en wat ouers gerus moet lees. http://ipwso.blogspot.com
Kontak die voorsitter vir ‘n algoritmiese kaart vir gastro- en ingewandsprobleme by die persoon met PWS.

Soos ons weet is *PWSA (USA)* baie betrokke by navorsing, asook die finansiering daarvan. Lees gerus oor slukprobleme en watter aspekte gekies is vir toekomstige navorsing. Soos ons weet bestaan daar nog nie behandeling vir die oormatige eetlus by persone met PWS nie. Kliniese toetse met verskillende medikasie word tans gedoen om te bepaal watter middel moontlik terapeutiese waarde vir die persoon met PWS kan hê. Op hierdie gebied het dr. Jennifer Miller ‘n belangrike bydra gelewer. ‘n Skakel na die volledige artikel in *Diseases* word gegee. In dieselfde tydskrif is daar ook ‘n artikel oor die gebruik van groeihormoon by die volwassene met PWS.

Dikwels word daar in artikels verwys na versterking van positiewe gedrag – maar, wat behels dit? Ook van die *PWSA (USA)* webwerf wenke hoe om voorkeurgedrag te gebruik sodat die persoon dit wat hy/sy nie wil doen nie, wel doen.

Verder van die Amerikaanse webwerf, “Moets” en “Moenies” uit die artikel W*eight & Behaviour Management.* Baie van die wenke wat gegee word, het ouers en versorgers al gehoor, maar dit is altyd goed om weer daaraan herinner te word. Ouers moet ingestel wees op voorkoming en gereed wees om in te gryp wanneer nodig.

PWSA (UK) het 'n artikel oor aggressiewe gedrag by die persoon met PWS beskikbaar gestel. Dit is 'n baie volledige artikel wat ook aandag aan ontspanningstegnieke skenk, wat asemhaling insluit. Die verwysings wat in die artikel gegee word, is van toepassing op die Verenigde Koningkryk. Lesers kan van die aanbevelings van toepassing maak op ons Suid-Afrikaanse omstandighede. Ons dank aan *PWSA (UK)* vir die baie waardevolle artikel.

Dan, laaste maar nie die minste nie. FAMCARE het weer eens 'n artikel gereed, *Strength in Boundaries.* Die stel van grense of perke laat die persoon met PWS veilig voel en verminder angstigheid. Ouers en versorgers moet daarop let dat grense nooit verslap kan word nie en dat dit ook geld in die volwasse lewe van die persoon.

Neem ook asseblief kennis van die volgende:

- Ledegeld vir 2015 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.
- Moenie *IPWSO News* oorslaan nie
- 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 23 Augustus 2015 aan en kom kuier gesellig saam. Hou die pos of epos dop vir meer inligting.

Opregte groete
Rika du Plooy

NOTICE AND INVITATION

THE PRADER-WILLI SYNDROME ASSOCIATION
TAKES PLEASURE TO INVITE YOU TO THE ANNUAL GENERAL MEETING ON
SUNDAY, 23 AUGUST 2015 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 23 AUGUSTUS 2015 Vriendekring Rolbalgronde, Groenkloof, Pretoria
Inligting aan lede volg

FROM THE CHAIRPERSON

Dear Readers

The winter is upon us and it is time for the June newsletter.

I would like to begin with a message of congratulations to PWSA (USA) which celebrates its 40th anniversary this year. In 1975, the *Prader-Willi Syndrome Parents and Friends* was

started by a couple whose son was diagnosed with PWS. Not long thereafter, the name was changed to the *Prader-Willi Syndrome Association*. Over the next 40 years, this organisation maintained their dedication to their motto of ***Saving and Transforming Lives***. This was achieved due to the synchronicity between the PWSA (USA) staff, professional volunteers and devoted parents. We thank you that the PWS community worldwide has been able to benefit from what has been achieved and from what has yet to be achieved.

I would like to introduce Larika Grix who celebrated her second birthday in May. An early diagnosis and a lot of research by her parents have been of great benefit to Larike. This is a positive story of parents, in conjunction with competent professionals, who assist this little girl in achieving her full potential. Readers are welcome to contact Liezl and Quinton.
Email: liezltruter1@gmail.com

What is cataplexy? In the article about Larike Grix we read about the fact that she was diagnosed with cataplexy at the age of 20 months. Please read elsewhere about the difference between narcolepsy and cataplexy.

The website of the PWSA (USA) and newsletter have interesting information on research, as well as articles with valuable practical tips. A warning to parents and carers is given in *The Gathered View* (June 2015), to exercise caution when a person with PWS experiences constipation. A very concise article appears on the blog of IPWSO which deals with the same topic and we advise parents and carers to read this. (http://ipwso.blogspot.com). An algorithm chart for gastro-intestinal problems in persons with PWS is available from the chairperson.

We are aware that PWSA (USA) is very involved with research and the funding thereof. Please read the articles about problems with swallowing as well as what areas have been chosen for future research. As we know, no treatment for uncontrolled appetite and lack of satiety of people with PWS exists. Clinical trials with various medications which may provide therapeutic options for those with PWS are currently underway in order to determine which medication would have the best therapeutic benefit. Dr Jennifer Miller has made an important contribution in this regard. A link to the complete article in *Diseases* is provided. An article about the use of growth hormone for adults with PWS appears in the same publication.

Reference is often made in articles about the reinforcing of positive behaviour – but what does this entail? The PWSA (USA) website carries tips on the use of preferred behaviour to ensure that in cases where the person does not want to do something, does indeed do so.

Also from the American website, are “Do’s and Don’ts” taken from the article *Weight and Behaviour Management.* Many of the tips given are already known to parents and carers, but it is always good to be reminded thereof. Parents and carers should be conditioned to practice prevention and be prepared to intervene if necessary.

PWSA (UK) has provided an article about aggressive behaviour by people with PWS. This is also a very concise article which gives attention to relaxation techniques, including breathing exercises. The advice and references provided are applicable to the United Kingdom and members are invited to apply the recommendations to South African conditions.

Last, but not least, another article from the FAMCARE committee. This time it is about *Strength in Boundaries.* The use of boundaries gives structure to the person with PWS and

lessens anxiety. Parents and carers should never relax on the set of boundaries. Read and learn more about consistent behavioural management strategies, including firm and clear boundaries.

Kindly take note of the following as well:

- Membership fees are due for 2015
- A friendly invitation is extended to contribute to the next newsletter due in November. You are welcome to write about your family or specific experiences of your child with PWS. This could be from parents, sibling or even grandparents.
- Please don't forget to follow *IPWSO News*
- This newsletter is also available electronically in A4 format

The next important event on the calendar of PWSA (SA) is the Annual General Meeting. Please make a note of Sunday 23 August 2015 and join us for a sociable gathering. Keep an eye on the post and emails for further information.

Sincere Greetings
Rika du Plooy.

Thanks to the Deegan family who assisted with the translation

MEET LARIKE GRIX

June 2014 - more interaction and smiles!

Larike Grix was born on the 21st of May 2013, at Medi-Clinic Nelspruit. She weighed a full 3,08kg and measured 51cm. Initial APGAR and other readings confirmed a healthy baby. The happiest day for any new parents! However, poor sucking and hypotonia got her admitted to NICU shortly thereafter and our long road ahead started. Following two and a half weeks in ICU, Larike was transferred to Unitas, Pretoria under the watchful eye of Dr MM Lippert. By this time, we were exhausted physically and emotionally. Following her first examination Dr Lippert suspected that she may have a rare syndrome named Prader-Willi. Following three days in Unitas, Larike got discharged and we could take our baby girl home for the first time. At that point in time, feeding was one of our biggest challenges, and we quickly developed numerous techniques to improve her sucking.

During her first six months at home she made steady progress. The weak sucking improved at the age of two months, where after our biggest challenge became her hypotonia. At the age of six months she could keep her head steady without any assistance. During this time Larike had her follow up at Dr Lippert and we were handed the test results which confirmed that Larike has PWS. We were devastated as we did not know what to expect and although we knew from her first examination that it might be PWS, we hoped Dr Lippert's initial diagnosis was wrong. Immediately our research went into overdrive. While we did some initial research when Dr Lippert suspected PWS, we now started searching everything we could get our hands on related to PWS on the internet. Perhaps, one of our biggest mistakes was believing everything we researched on the internet as we quickly realised Larike is, as any other child, unique. During this examination, Dr Lippert referred us to Dr Van Dyk, an endocrinologist who practices from Little Company of Mary Hospital in Pretoria.

Larike's christening was a special occasion for her parents and family.

At the age of seven months, Larike's interaction with us and her surroundings improved, she smiled often and could hold her own bottle. When Larike turned eight months, we visited Dr Van Dyk who prescribed Growth Hormone Treatment in the form of Humatrope. Larike was able to sit, unassisted at the age of nine months. Although we believe the growth hormone made a noticeable difference, we knew by then that we had to make certain changes and that we are going to need more assistance and support than the small town we resided in (Lydenburg, Mpumalanga) could offer us. We then made the decision to relocate, with the primary objective of being closer to doctors, occupational therapists, physiotherapists, dieticians etc.

We subsequently moved to Potchefstroom during April of 2014 being some 200 km closer to the said medical practitioners. By 14 months, with the assistance of numerous physiotherapists, occupational therapists, Growth Hormone Treatment and other medical interventions, Larike was able to crawl about. During this time, Larike had scheduled appointments at various practitioners including physiotherapists (twice weekly), occupational therapists (weekly) and daily growth hormone injections (three monthly follow-ups at the endocrinologist). At the age of 18 months, Larike started pulling herself up against the furniture, working her way around the house, slowly building her confidence to one day take that big, unassisted step!

Larike started nursery school in January 2015, in an attempt to develop and improve her social skills. She is a fun loving child between her friends and enjoys the school and friends a lot. Around the age of 20 months, we noticed a peculiar occurrence during, what seemed to be predominantly in the mornings, Larike rapidly losing control of her body for a split second and then revives and continues as if nothing has happened. Initially we thought this was only as a result of not woken up entirely, however after some research we found that this may be quite a serious condition related to epilepsy. A visit to Dr Lippert and a few tests later, revealed that Larike has a condition called "Cataplexy", that according to Dr Lippert is a rare case for 'n child with PWS, but which she will most likely outgrow. At Larike's school she attends 'Kinderkinetika' to help develop her gross motor skills, Smart-Brain to assist with general brain development and the school has services for regular eye and ear tests.

Larike and her occupational therapist

21 May 2015 –
Larike's 2nd birthday!

It was during some of these tests that the audiologist noticed some abnormalities pertaining to Larike's hearing and referred us to an Ear, Nose and Throat Specialist, Dr Tiedt. Four days later, grommets were installed in Larike's ears to improve drainage and her subsequent hearing, and these made a valuable difference! To ensure Larike has all the advantage she requires to walk unassisted, we approached an orthopaedic specialist who examined Larike's feet. He noticed Larike's ankles tilting inwards whilst trying to walk, and suggested custom made inner soles to assist her ankles. Larike is currently in the process of trailing these!

Apart from the various tests, therapies and procedures to ensure Larike has an advantage and the best opportunities in life, she is a healthy, loving and unique little girl whom we love with everything in us!

Currently Larike continuous to go to school and loves her friends and teachers very much! We're continuing with the daily injection of the Growth Hormone Treatment and see the occupational therapist bi-weekly. Larike also started with swimming lessons and is progressing well!

Furthermore, we as her parents came to the realisation that we need to accept Larike as the unique individual that she is, and will be. All that we can do is to ensure she will live her life to the fullest, and to help develop a lifestyle that will benefit her in every way possible.

What we learned so far, is to trust your instincts, and accept others' support. Don't try to take on this adventure on your own; you will be surprised what a little support can do for you.

With regards from Quintin, Liezl and Larike Grix, Potchefstroom.

(Note: While I was busy compiling the newsletter I received an email from Liezl with the exciting news that Larike started to walk on Sunday 14 June 2015! Rika du Plooy)

WHAT IS NARCOLEPSY?

From: National Institute of Neurological Disorders and Stroke

PWSA (SA) thanks Dr Engela Honey for her assistance.

Narcolepsy is a chronic brain disorder that involves poor control of sleep-wake cycles. People with narcolepsy experience periods of extreme daytime sleepiness and sudden, irresistible bouts of sleep that can strike at any time. These "sleep attacks" usually last a few seconds to several minutes.

Narcolepsy can greatly affect daily activities. People may unwillingly fall asleep while at work or at school, when having a conversation, playing a game, eating a meal, or, most dangerously, when driving or operating other types of machinery. In addition to daytime sleepiness, other major symptoms may include cataplexy (a sudden loss of voluntary muscle tone while awake that makes a person go limp or unable to move), vivid dream-like images or hallucinations, as well as total paralysis just before falling asleep or just after waking-up.

Contrary to common beliefs, people with narcolepsy do not spend a substantially greater proportion of their time asleep during a 24-hour period than do normal sleepers. In addition to daytime drowsiness and uncontrollable sleep episodes, most individuals also experience poor sleep quality that can involve frequent waking during night time sleep, and other sleep disorders.

For most adults, a normal night's sleep lasts about 8 hours and is composed of four to six separate sleep cycles. A sleep cycle is defined by a segment of non-rapid eye movement (NREM) sleep followed by a period of rapid eye movement (REM) sleep. The NREM segment can be further divided into increasingly deeper stages of sleep according to the size and frequency of brain waves. REM sleep is accompanied by bursts of rapid eye movement along with sharply heightened brain activity and temporary paralysis of the muscles that control posture and body movement. When subjects are awakened, they report that they were "having a dream" more often if they had been in REM sleep than if they had been in NREM sleep. Transitions from NREM to REM sleep are controlled by interactions among groups of neurons (nerve cells) located in different parts of the brain.

For normal sleepers a typical sleep cycle is about 100 to 110 minutes long, beginning with NREM sleep and transitioning to REM sleep after 80 to 100 minutes. People with narcolepsy frequently enter REM sleep within a few minutes of falling asleep.

Cataplexy

Cataplexy is a sudden loss of muscle tone while the person is awake that leads to feelings of weakness and a loss of voluntary muscle control. Attacks can occur at any time during the waking period, with individuals usually experiencing their first episodes several weeks or months after the onset of excessive daytime sleepiness (EDS). But in about 10 percent of all cases, cataplexy is the first symptom to appear and can be misdiagnosed as a seizure disorder. Cataplectic attacks vary in duration and severity.

The loss of muscle tone can be barely perceptible, involving no more than a momentary sense of slight weakness in a limited number of muscles, such as mild drooping of the eyelids. The most severe attacks result in a complete loss of tone in all voluntary muscles, leading to physical collapse during which individuals are unable to move, speak, or keep their eyes open. But even during the most severe episodes, people remain fully conscious, a characteristic that distinguishes cataplexy from seizure disorders. Although cataplexy can occur spontaneously, it is more often triggered by sudden, strong emotions such as fear, anger, stress, excitement, or humour. Laughter is reportedly the most common trigger.

The loss of muscle tone during a cataplectic episode resembles the interruption of muscle activity that naturally occurs during REM sleep.

A group of neurons in the brain stem halts activity during REM sleep, inhibiting muscle movement. Using an animal model, scientists have learned that this same group of neurons becomes inactive during cataplectic attacks, a discovery that provides a clue to at least one of the neurological abnormalities contributing to human narcoleptic symptoms

It is with sadness that we announce the passing of Andrea Lake in 2014. She was born on the 3rd March 1981. Her family moved to the United Kingdom.
We want to express our sincere condolences to the family.

FROM THE WEBSITE, NEWSLETTER AND E-NEWS OF PWSA (USA)

Celebrating our 40th year!! PRADER-WILLI SYNDROME ASSOCIATION USA
Still hungry for a cure.

MEDICAL ALERT
CONSTIPATION IN INDIVIDUALS WITH PRADER-WILLI SYNDROME

James Loker, M.D., Pediatric Cardiologist, Ann Scheimann, M.D., M.B.A., Gastroenterologist, PWSA (USA) Clinical Advisory Board Members
The Gathered View ~ Prader-Willi Syndrome Association (USA) May-June 2015

Constipation is a common problem in individuals with Prader-Willi syndrome (PWS). It takes longer for food to move through the GI system in Prader-Willi syndrome.* This slower passage of food can lead to serious issues similar to the ones seen related to the stomach (see MEDICAL ALERT BOOKLET UNDER SEVERE GASTRIC DISTRESS and GASTROPARESIS). Outpatient methods used to clear constipation in non-PWS patients may be ineffective due to poor fluid intake and hypotonia. Inpatient regimens frequently use large volumes of fluid which may cause problems. Reliance on these methods may lead to life-threatening conditions such as necrosis and perforation of the colon and subsequent sepsis. Due to decreased muscle tone and altered pain response, individuals with PWS may not have the same clinical exam that a non PWS patient would have. A heavier reliance on imaging may be necessary. Individuals with PWS may be at higher risk for impaction. Rectal examination and enema may be required in addition to oral cleanout regimen. This may also be problematic in some leading to rectal picking.

Patients with PWS having constipation and receiving repeated regimens of oral PEG (polyethylene glycol) solutions for bowel cleansing should be monitored closely for abdominal distension and retention. **Failure of standard constipation protocols to clear the stool in a timely manner, especially in the face of increasing abdominal distension, vomiting, decreased appetite, stoppage of food consumption and/or abdominal pain warrants surgical or GI consultation. Emergent surgical or colonoscopic intervention may be necessary.**

Kuhlmann, et al. (2014) *A descriptive study of colorectal function in adults with Prader-Willi Syndrome: high prevalence of constipation. *BMC Gastroenterology, Apr 4; Vol 14: page 63*

Please visit IPWSO's blog for a very extensive and parent friendly article on *Constipation in PWS,* by Dr Susanne Blichfeldt and Linda Thornton. The article contains some really important information that needs to be understood by not only parents and caregivers, but by the medical profession as well. http://ipwso.blogspot.com

STATUS OF PWSA (USA) RESEARCH GRANTS

As most of you are aware, PWSA (USA) has a long history of funding research, and also programs that accelerate and enhance research on Prader-Willi syndrome – such as the programs we sponsored in 2014 at Obesity Week. We are pleased to announce the funding of the following research grant:

REDUCING THE RISK OF PRANDIAL ASPIRATION AND CHOKING IN PERSONS WITH PWS

Principal investigator: Roxann Gross, Ph.D.

In the past, the increased risk of choking to death in PWS was thought to be primarily caused by gorging and rapid eating behaviours. Thanks to an initial grant from PWSA (USA), research data was obtained revealing the real problem was swallowing problems and poor esophageal function. The most striking finding was discovering the presence of often large amounts of food remaining in the throat and the esophagus after swallowing. Alarmingly, 97% of those studied had no sensation or awareness that the food "didn't go down".

The purpose of this new study will be to provide the medical community with evidence-based recommendations for the evaluation and treatment of dysphasia in persons with PWS. Once completed, this research will significantly advance the understanding of how swallowing problems in PWS can be treated. This work is highly relevant to the current research mission of PWSA (USA) because the results will have a direct and immediate impact on care.

Two more grants have been sent back to the investigator for revisions and resubmission. PWSA (USA) will also be putting out a new request for grant submissions in May. The research committee decided we would put an emphasis on the topics of:

- finding solutions to dealing with gastroparesis in PWS
- sleep issues in PWS and how it plays a role in behaviour
- aging in PWS
- psychotropic medications – what works and doesn't work with PWS
- pulmonary embolus – the cause of 7% of PWS deaths (PWSA current study of death)
- developing a better hyperphagia scale for today's population – including those who have controlled hyperphagia
- motor tics/tardive dyskinesia – reducing the risk; minimizing the impact
- biomarkers for health risk factors in PWS
- postoperative issues and management

At PWSA (USA) we work to integrate what we have learned about the needs of our families through our support programs with research that we think will make an important and practical difference in our children's lives. **Saving lives today – enhancing lives for tomorrow!**

The Gathered View ~ Prader-Willi Syndrome Association (USA) May-June 2015 ***5***

MORE ON RESEARCH

Dr Jennifer Miller, member of PWSA (USA)'s Clinical Advisory Board, wrote the linked published paper, with editing and assistance from Janalee Heinemann, PWSA (USA) Coordinator of Research and International Affairs, and Dr. Theresa Strong, FPWR Scientific Advisory Board chair. The paper titled ***"Medication Trials for Hyperphagia and Food-Related Behaviors in Prader-Willi Syndrome***" has been published in **Diseases** at http://www.mdpi.com/2079-9721/3/2/78

Conclusions of the paper:

"This is a tremendous time of hope for potential treatment of the appetite issues in PWS. Given the fact that the great majority of patients are now diagnosed in infancy, the combination of early intervention and early growth hormone therapy, along with a medication to decrease the appetite issues could result in a much improved quality of life, and increased independence, for individuals with Prader-Willi syndrome."

We want to thank Dr. Miller for her long-term dedication to our children and adults with Prader-Willi syndrome and for her long-term commitment to volunteering with PWSA (USA).

Also in this special edition of PWS in **Diseases** is a paper written by our PWSA (USA) research committee member, Dr. Karen Vogt (an endocrinologist and parent of a child with PWS) and Jill Emerick. The paper is titled ***"Growth Hormone Therapy In Adults With Prader-Willi Syndrome."***

PWSA (USA) IS SPONSORING A GRANT THAT WILL BRING THERAPY TO THE HOME!

PWSA (USA) Joseph McErlane Research Grant
From Janalee Heinemann, MSW, Director of Research & Medical Affairs

Project Title: Evaluating the Feasibility of a Telehealth Intervention of Early Social Cognitive Processes in Children with Prader-Willi syndrome
Principal Investigator: Anastasia Dimitropoulos, Associate Professor of Psychology, PhD. Case Western Reserve University, Cleveland, OH

Relevance of this study:
Current research in typically developing children has shown that pretend play, the ability to play with toys in an imaginative way, is related to important areas of development, such as social-emotional understanding. Research has also shown that individuals with PWS have difficulty in social situations, in emotionally understanding others, and have difficulties with regulating their behaviour and emotions in difficult situations. Since many children with PWS have trouble with social-emotional skills, intervention targeting these skills through pretend play could increase quality of life while also decreasing problem behaviours, as has been shown in children with other developmental disabilities.

Through this grant, they will administer intervention in the participant's home (using telehealth videoconferencing) to increase its accessibility to the PWS population. The goal is the following: (1) to increase the social-emotional abilities in children with PWS; (2) increase parent-child play interactions; (3) reduce the frequency of externalizing behaviours such as emotional outbursts and stereotyped or rigid behaviours; (4) confirm the feasibility of conducting intervention via telehealth. If feasibility and improvements are found, these results will lead to establishing a practical treatment alternative for families dealing with Prader-Willi syndrome.

Eligible participants will be children with PWS between 5-11 years of age. Individuals without internet capable computers will be excluded from participation; however equipment will be available for participating families who do not have necessary components (i.e. webcam). Details of eligible participants will be up available in the near future through PWSA (USA) Internet sites.

(Note: Wouldn't it be wonderful if we could provide help for families that live far from major treatment centers and spare all families dealing with PWS from some of the multiple trips they need to take to get the proper intervention? We see this pilot project as the 1st step in bringing this type of intervention to your home. It's the wave of the future! ~ *Janalee*)

WEIGHT & BEHAVIOUR MANAGEMENT

An excerpt from the article *WEIGHT & BEHAVIOR MANAGEMENT* www.pwsausa.org

There are no easy answers to any of the problems surrounding the management of this unique condition. The following "do and don't" suggestions have come from parents, professionals, and observations of persons with PWS. In addition, use good humour, kindness, affection, determination and respect, seasoned with flexibility and good judgment, and there will be a good amount of success.

DO

- Keep food inaccessible at all times. Persons with PWS cannot fight their compulsion to get at it. Put it away and lock the cupboards and refrigerator.
- Keep their lives structured. They need structure. Pre-plan changes.
- Praise and recognize good performances. A lot of mileage can be obtained with a few words, smiles and hugs.
- Listen to a person who needs to talk. The time it takes may alleviate or prevent unpleasantness later.
- Include the person with PWS in planning and programming. They feel a need for some control and will cooperate 100 percent if they feel it was their idea.
- Keep sight of the fact that the hand of a person with PWS is quicker than your eye.
- Enjoy your treats in private. It is very hard for the person with PWS to watch others enjoying the goodies he is denied.
- Use smaller plates and cups, spread the food out, and add extra non-fattening items such as carrots, dill pickles or diet jelly to make the amount on the plate look larger.
- Inform neighbours, relatives, teachers, babysitters, classmates, everyone with whom the child with PWS comes in contact - and then inform them again!
- Be consistent. They thrive on routines and knowing exactly what the guidelines are.
- Remember that logic and reason will not prevail when the person with PWS gets upset and/or "stuck" on an idea or position.

DON'T

- Use food as a reward or punishment except on a very limited basis.
- Assume if the person has lost weight, that the problem is now "cured".
- Nag. Once a behaviour has been dealt with, do not bring it up again.
  Discuss temper tantrums and then forget it.
- Argue. Make the statement, allow the person one more comment, warn that the discussion is over and stick to it. You will never win an argument.
- Tease, be sarcastic, or even use subtle humour. People with PWS do not respond well to such tactics.
- Ignore bad behaviour. Try interventions to prevent it.
- Lose your temper. Easier said than done, but do whatever it takes to keep your cool; nothing will be gained if you lose control.
- Promise anything that you cannot or will not do. Persons with PWS rarely accept change.
- Ignore poor table manners. They are capable of using utensils; they can slow down; they can stay until the meal is finished without additional food.
- Try to talk things out with a child with PWS. It does not work!
- Lose sight of the humorous aspects of all of this. Hang on to your sense of humour while figuring out "how to lock up the apple tree".
- Hesitate to ask for professional help.
- Forget that this is a life-threatening situation.

TIPS FOR CREATING A POSITIVE BEHAVIOURAL PLAN

WRITE IT, SHARE IT, POST IT, AND USE IT. From the website of PWSA (USA)

Positive behavioural support is an essential tool for all caregivers of a person with Prader-Willi syndrome. It is best to implement positive behavioural strategies early in a child's life – even before behavioural challenges emerge. It is an important pro-active and preventative step to become familiar with and implement positive behavioural strategies.
A positive behavioural support strategy uses a person's strength and interests to help motivate them to achieve non-preferred goals and activities. It is a way to do some behavioural problem solving in a fun, creative, and effective way.

Challenging behaviours are a feature of Prader-Willi syndrome (PWS). When responding to these behaviours, it is important to remember that negative consequences (which include responses such as shaming, threatening to take things away, etc.) are not typically effective in helping a person with PWS to manage their behaviour more appropriately. The cognitive and impulse control deficits caused by PWS inhibit the ability to understand what a negative consequence is trying to teach so it does not altar future behaviour. And most often it leads to a power struggle which rarely helps to improve a challenging behaviour. So what does work?

People with PWS are successful behaviourally when a positive behavioural strategy is developed and consistently employed. This is a tried and true strategy that is effective for people with PWS of all ages. A positive behavioural strategy uses incentives and rewards to move a person successfully through the day by mixing preferred and non-preferred activities.

For example: *If Tommy, who has had a problem getting up and ready for school in the morning, is able to accomplish this task in the designated half hour time period he will receive a sticker. If he earns 10 stickers during the week, he will be able to pick a movie he wants to watch.*
In this example, Tommy's desire to watch a movie of his choice is used to motivate him to successfully accomplish a non-preferred activity – getting ready for school on time.

How to get started?

- Create a list of challenging behaviours. These are the behaviours you want your plan to help change.
- Prioritize the list by identifying one or two behaviours to be addressed first. Behaviours that are most disruptive could be at the top of the list or you could start with behaviours that will improve more quickly so the person begins to earn rewards and feel successful more quickly which can increase ongoing commitment to the plan.
- Create a list of rewards or incentives that will motivate the person. When possible, invite the person with PWS to help you create this list. This creates a good opportunity also for you to begin to explain the purpose of the new plan you are creating in a positive way.
- Create the plan for the initial behaviours you want to target. As with the example above, create a strategy for each challenging behaviour that includes the preferred behaviour you want to encourage and how and when the reward will be applied to motivate the behavioural change you want to help create. Pay attention to how often the reward is applied. For some people a weekly reward works well but for

others the positive reinforcement might need to be daily or even hourly. Find what works for the person you are supporting!

Tip: Before beginning step three, do some research. You can find many helpful resources on positive behavioural strategies by searching online. If you are working with a counsellor, social worker, or school professional they also might be able to assist you with ideas. And don't forget to contact PWSA (USA) for helpful behavioural resources (video and written) for supporting people with PWS. The more you know the better your plan will be!
Once you have a written plan designed, share it with the person with PWS and others involved in their life so they understand the expectations of the plan and how it works, post it where you both can see it every day, and use it consistently. A good positive behavioural plan is an essential foundation for diminishing challenging behaviours experienced by a person with PWS.

AGGRESSIVE OR VIOLENT BEHAVIOUR IN PWS

AGGRESSIVE OR VIOLENT BEHAVIOUR IN PWS

On request received from PWSA (UK)

Children and adults with PWS are usually happy, friendly, sociable and loving individuals, so it can sometimes come as a shock if they exhibit violent or very aggressive behaviour, or when emotional outbursts (sometimes called "tantrums" or "melt-downs") begin to escalate into physical aggression.

We do not have any definite figures to say how common aggressive or violent behaviour is in PWS, especially as it can occur in phases, affecting individuals at different times and in different circumstances. We do know that it can occur in very young children and at any age thereafter.

UNDERSTANDING AGGRESSIVE OR VIOLENT BEHAVIOUR IN PWS

Aggressive or violent behaviour in any one, regardless of whether or not they have PWS, usually results when someone has reached the limits of their endurance of a situation. How much any one person can stand varies considerably between all individuals.

People with PWS are much more vulnerable to stress than the general population. Generally speaking, they have a low tolerance for anxiety and frustration. They also have poor control over their emotions, acting impulsively. Most find it difficult to deal with change. They have difficulties processing information, so that it takes longer for them to work out what is happening and why. The combination of these factors means that acting out is more likely to happen in PWS, and sometimes this result in aggression.

However, the onset of mental health problems such as psychoses and an escalation of anxiety and mood disorders can also be exacerbated by stress, and consideration must be given to this as a possibility, if the behaviour cannot easily be attributed to other triggers.

Certain medications may also give rise to more aggressive or violent behaviour. Testosterone as a hormone treatment for males is sometimes mentioned anecdotally as a cause, but research has so far not discovered a distinct relationship. There are case reports suggesting that oestrogen therapy for females can increase mood liability as well. The use of SSRI antidepressants and atypical neuroleptics has been linked with mood and

behavioural activation that can present with impulsive behaviour, aggression, self-injury, and property destruction.

TRIGGERS

These are some of the more general triggers, not only for aggressive or violent behaviour, but for emotional outbursts of any kind:

- Anxieties around food; presence of food in the room or nearby
- Frustration at not being able to do what he or she wants to do
- Communication or speech and language problems
- Feeling under pressure (whether or not the pressure is actually there – e.g. saying "she forced me to do it" when in actual fact the person was merely making a suggestion about what they the person with PWS might like to do).
- A "chaotic" environment with too much stimulus and/or too little structure
- In a low or unstable mood
- Unrealistic expectations on the part of others
- Real or perceived unfairness in a situation
- Change of any kind
- False expectations or disappointment
- Bullying, or physical or sexual abuse by others
- Harsh tone and punitive attitudes from carers
- Additional undiagnosed medical and learning disability conditions (e.g. autistic spectrum disorder)

However, there are many other individual triggers. Finding out what is the cause can be difficult, because of the propensity of people with PWS to shift blame or give another reason for an outburst than what was the real cause.

PREVENTING OR MINIMISING AGGRESSIVE AND VIOLENT BEHAVIOUR

Create the right environment

In view of all the above considerations, the environment for someone with PWS has to be "right"- but it is up to others to provide the environment in which they can thrive. It is most likely the environment, or other people in that environment, which are the cause of challenging behaviour when it only occurs in certain situations, e.g. at school, but not at home, or vice versa.

For many individuals with PWS, their behaviour will stabilize when in a structured, low-stress environment, where food access is controlled, boundaries are clear and a consistent approach is adopted by everyone involved. Everyone working with the individual should be aware of potential triggers and how to avoid them.

Recognise signs of emotional distress and act before the situation escalates

Each individual may have their own way of showing that they are becoming distressed: rubbing or shutting eyes, wringing or waving hands etc. Over time you will recognise what they are, and sometimes it is possible to take the person to a calm, safe space, acknowledge that you have recognised they are becoming upset, and address any issues they may have – particular if these are relatively minor.

TEACH COPING SKILLS AND BEHAVIOURAL RELAXATION TECHNIQUES

Coping skills
Coping skills should be embedded into the daily programme of activities for people with PWS. They should also be scripted and rehearsed with carers in residential care and other situations where the person is away from home. Then, when the carer perceives an escalation, they can cue the person to use one of the practiced techniques. In this way, they can draw on these before they begin to go into meltdown. These skills include:

- Deep breathing
- Stress relief through sensory balls
- Listening to music
- Learning how to communicate feelings appropriately
- Taking a break

Behaviour relaxation techniques
Learning simple breathing control exercises and how to tense and release different muscles groups throughout the body can help reduce feelings of anxiety arousal or calm an individual who is feeling annoyed. Again, these techniques must be scripted, rehearsed, and cued by carers and imbedded in the daily program.

Adopting a Relaxed Posture Encourage the person with PWS to sit comfortably in an arm chair, with arms should be placed palms down along the arms of the chair in a resting position. Encourage the person to rest their head back in the chair and put both feet up on a foot rest, and take slow deep breaths and close their eyes. (You may need to demonstrate how to do this and praise when it is done correctly)

Tensing and Releasing – Show the individual how to tense the following muscle groups in turn. Each tensed position should be held for a slow count of 10, and then relaxed so muscles go floppy like jelly. Repeat each position at least three times.

- Hold arms out and clench fists
- Pull shoulders up to ears
- Push both arms down on arm rests
- Strongman (tense biceps)
- Push both knees together
- Push head back hard into head rest
- Open mouth and eyes wide
- Close eyes and scrunch up face

Abdominal Breathing – Encourage the person with PWS to lay back in a comfortable chair with their feet up, hands on belly, with fingers interlinked. Show them how to breathe deeply into their belly, not their chest, so that their belly moves in and out. You'll know if they are doing this correctly as their interlinked fingers will separate slightly.

Breathing Elephants – Ask the person to take a very deep breath in and as they exhale, try and count as many elephants as they can until they run out of puff "one elephant, two elephants, three elephants…"etc. (They should not continue to breathe normally as he counts, the object of the exercise is to regulate breathing). This is a good, fun exercise which can be done as a competition with another person to see who can count most elephants before they run out of breath!

MANAGING AGGRESSIVE AND VIOLENT BEHAVIOUR

The person who is being attacked is not necessarily the cause of the aggression, which is often focussed onto the nearest available object or person.
Your immediate response and throughout should be calm and low key, ignoring screaming and shouting. Do not do or say anything which might make the situation worse. Be aware that people with PWS are very sensitive to tone of voice; even the slightest hint of irritation, cajoling, or frustration on your part will be picked up by them. Do not argue with a PWS person; this will only escalate or prolong the incident. Threats and bribery are ineffective. It is futile to attempt to reason with a PWS person in the middle of an outburst, e.g., saying "things aren't that bad" will not work - the thinking and problem solving part of their brain is "offline" at this time.

Make the environment as safe as you can, and direct other people away from the incident.

After an incident
The person with PWS will usually need time to recover their equilibrium. So provide a calm, low stimulating space, where they can go and lie or sit quietly. Many often go to sleep.

Wait until everything is calm again, and then, if possible, try to address the issue in a sensitive manner and find out what caused it. This is not always possible, however, because the individual may become upset again, feel under pressure or be unable to identify a cause. You may have to wait until the next day for the person to be calm enough to respond without the situation escalating again. What may seem a very minor issue can make a person with PWS very anxious or frustrated, so sometimes it is relatively easy to ensure that issue is addressed, but often there are more deep-seated concerns.

The most important thing after an outburst resolves is to get the person back into the program of the daily schedule as soon as possible.

Parents - Help for you
If your son or daughter is displaying aggressive or violent behaviour it can be very stressful. He or she may direct this behaviour towards yourself or other family members, or it may be directed at children and teachers at school or college, or staff in day centres or residential homes and supported living. Many describe their situation as "walking on eggshells" – never sure when the next meltdown will occur, or, if the phone rings, if it will be yet another request to collect your child from school or college, with a threat of, or actual, exclusion.

The important thing to remember is that PWS is a very complex disorder and that your parenting skills are not necessarily at fault. You may need help from experts (see below) who can carry out a thorough analysis of what is happening with your child and recommend management strategies or treatments.

On very rare occasions, this may mean moving the child or adult out of their present circumstances into something more appropriate (e.g. from mainstream to special school, or from supported living to residential care), or into a specialist treatment centres. Again, this is not an indication of failure on your part, and may well be in your son or daughter's best interests, providing them with an environment in which they are better able to cope.

Recognise when you are reaching a situation in which you personally are unable to cope:

- Feeling helpless or powerless
- Breaking down in tears
- Dreading phone calls

- Feeling frightened of the person with PWS
- Wanting to run away

Talk to your GP or call social services and ask for help. Remember too that the PWSA UK is here to offer a listening ear.

Help from experts

If you have tried everything, or the aggression is becoming a danger to others or the person themselves, ask your GP, community nurse, consultant or social services for a referral to psychiatric services. *When you go for the referral, make sure you take along information about PWS, including this leaflet, and this very helpful article for psychiatrists written by Janice L Forster and Linda M Gourash from the USA.*
http://www.pwcf.org/wp-content/uploads/2012/02/Psychiatry-Primer-for-PWS1.pdf

If the psychiatrist has little or no experience in working with someone with PWS, we can provide contact details of someone they can speak to for more information.

In some cases, medication may be prescribed. As a general rule, this should be at a lower dose than normal. © PWSA UK

A FRIENDLY REQUEST

The next newsletter will appear in November 2015. We invite you to share your story and tell us about your loved one with PWS. Parents, brothers, sisters or even a grandma or granddad are welcome to contribute to our newsletter.

STRENGTH IN BOUNDARIES

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

Personal boundaries are guidelines, rules or limits that a person creates to identify for themselves what are reasonable, safe and permissible ways for other people to behave around them and how they will respond when someone steps outside those limits. They are built out of a mix of beliefs, opinions, attitudes, past experiences and social learning (Wikipedia, the free encyclopedia)

People with PWS have a heightened anxiety and need others to set consistent and consistently enforced guidelines or boundaries for them to feel safe and respond appropriately to what is going on around them. Boundaries include positive behaviour strategies through the encouragement of desired behaviour rather than the punishment of undesired behaviour. Boundaries teach the person with PWS what is desired by you and society and how to communicate effectively. Anything taught must be simply stated, understandable and reinforced in a consistent manner to minimise anxiety.

Using boundaries is important for all people working with a person with PWS. They are tools to help reduce anxiety and poor reactions by the person with PWS to what is happening or being said to them. When boundaries are not put in place by all people involved with the person with PWS, it is confusing and problematic for the person with PWS. Lack of consistent use of boundaries can create long lasting difficulties for people with PWS. Boundaries keep us all “on the same page”.

Let's remember how people with PWS think...

1) For most people with PWS, there exists constant, life-long anxiety from choices to be made, the things they hear and see, the speed at which decisions have to be made, the constant drive to eat and in many cases, the desire to be "the same" as others. As well, most people with PWS are concrete thinkers! The words you use and the tone of voice you use are what the person with PWS will hear. Very few can "read between the lines" or interpret an underlying meaning. If you speak loudly or with even a slightly annoyed tone, they will "hear" that you are angry. This will increase their anxiety and most often result in a negative response! If you become anxious, their anxiety will also increase. Make every word count because the person with PWS will hear all the words but may not always understand the meaning of what is said. Using fewer words and a gentle voice keeps you calm and stops you saying words that can be misinterpreted or latched on to for an argument. Less is more

2) Too much choice is confusing for people with PWS. Limited, appropriate choice keeps everyone safe. We would not ask someone with PWS: "where would you like to go, today?" We might ask: "would you like to see a movie or go to the zoo, today?" ***Less is more***

3) Using visual guides is always helpful! Too many instructions are confusing as people with PWS have a delay in their information processing ability and often miss bits of what is said. Displaying pictures to describe a task of several steps makes the process easier to understand. For example: washing hands - a picture is easier to understand, learn from and remember for people with PWS. Remember that people with PWS have enhanced spatial patterning skills – they take in the whole pattern at a glance!

The earlier in life boundaries are established, the easier it is for the person with PWS. By the time adulthood is reached, many parents are exhausted by the constant PWS management they have had to practise. Some parents also feel that as the person is now an adult, things will change or they should have the ability to be responsible for their own behaviour, and boundaries that were consistently in place when they were children, can now be relaxed. We know this is not the case for people with PWS. They will always require consistent behavioural management strategies, including firm, clear boundaries, which will be constantly tested by the person with PWS. If they think there is any chance of breaking them down they will persist even more so. Once boundaries have been loosened or the person with PWS has experienced the freedom of minimal or no boundaries, their world gradually starts to fall apart. It is then so much harder to reinstate the boundaries that had previously kept them feeling "safe" and had reduced behavioural outbursts. As well as respecting the individual with PWS we must remember to always respect the characteristics of the syndrome that will not change. Expectations for the person with PWS must remain realistic.

Reinstating boundaries is hard, but not impossible! Generally, a catalyst will help provide a platform from which to introduce change – that is, by changing the non "bounded" environment back to one that is "bounded". Writing up new or reinstated boundaries in the form of a contract can be very helpful! Simple, clear explanations as to why the boundaries are necessary are essential and if possible, can be suggested by a doctor or other professional detached from the family.

Basic boundaries practised for children with PWS, for example, around the regularity and quantity of treat foods, will still apply to adults with PWS, however, the implementation of the boundaries may alter for the adult with PWS. Boundaries need to be reviewed frequently, to maintain their appropriateness to the individual and their effectiveness in each person's situation. Boundaries may need to be adapted to different stages in life and changed circumstances – for example, during the transition period from school to post school programs or from a "community" program to a work program. Be aware of changes in boundaries, especially those that sneak into the person's usual lifestyle!

Parents are the initial and most important boundary setters, as they know their child (young or adult) better than anyone else and they have been the key people in the lives of their child. When an adult with PWS starts attending programs independent of family it is often seen as an opportunity to develop his/her independence. Parents may, often reluctantly, step back from their role as boundary-keeper, as they allow caregivers to develop a working relationship with their person with PWS. This is when boundaries may be refined for the situation, but their necessity still remains.

A parent writes…

> When my daughter started attending a day program she was introduced to the practice of having an early morning coffee with the staff and again at tea time. This number of cups of coffee have gradually increased and become "holy ground" for her. In conjunction with this her use of artificial sweeteners has escalated dramatically, because she was in charge of the sweeteners. In hindsight I realise I should have been more on the alert! The process to set new boundaries was a very difficult one and caused challenging behaviour. I should have kept limits on her usual daily coffee intake and never have allowed her to start using artificial sweeteners.

Should the breaking of boundaries carry consequences?

Consequences emphasise the importance of boundaries, but do people with PWS learn from consequences? We all know that food will be eaten if accessible, no matter what, and that having "punishment" type consequences around the eating of unplanned food is not fair for people with PWS. However, having appropriate consequences for known, discussed and understood boundaries, can be quite powerful. The consequences must also be well understood and always carried through, should a boundary be broken. Remember, the power of consistency!

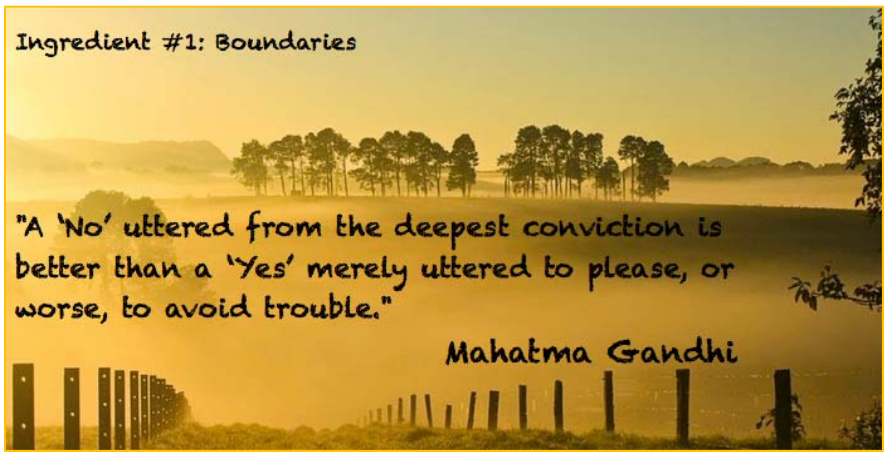
For example:
Peter loves bowling and usually goes every week. There is a cake shop on the way to bowling and on one occasion, when he was being taken to bowling by a casual staff member, he managed to get them to buy a fruit bun for him. His reason was that he had not eaten all of his breakfast and was feeling a little "weak". The staff member was rebuked for allowing this, as it was not Peter's schedule but he can be very convincing to unsuspecting new staff! Ever since this event, on occasions when Peter is in a bad mood, he will stop on his way to bowling and try to convince the staff member accompanying him, that he really needs a fruit bun. In most cases he is reminded of the "boundaries" but sometimes he will become very agitated and on rare occasions, a full blown temper tantrum will occur. His staff has now implemented the consequence of returning home immediately, should he becomes agitated concerning the cake shop. So he does not attend bowling that day. The frequency of missed bowling days is very low, as he is reminded of how much he enjoys bowling each night prior to his planned attendance.

Is it the boundary or the consequence that has improved Peter's behaviour near the cake shop? Possibly the answer is the combination of both.

Both example situations highlight how important it is for all family members and caregivers to be well informed about PWS and the need for people with PWS to have consistent boundaries.

The most effective boundaries are those that are seen, by the person with PWS, to be beneficial to him/her in some way, or to be acceptable to an intelligent person, such as himself/herself.

What keeps a boundary effective is the belief in its worth and the consistent practice of that boundary.



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.

TIP OF THE WEEK: MANAGING STRESS

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

Here are some tips for helping your child reduce their anxiety:

1. Teach coping skills. Deep breathing, visualizations and muscle relaxation are all excellent tools to decrease anxiety.
2. Praise bravery. Every time your child does something that previously made them anxious or scared, reinforce their bravery.
3. Point out their triggers. The more your child understands what causes their anxiety the more successful they will be in managing their own feelings and actions.
4. Stick to a routine. The more predictable the better. Unknowns are very difficult for our kids so limit them as much as possible.
5. Check yourself. If you are stressed or anxious your child will sense that and respond accordingly.
Stress and anxiety can be debilitating for people with PWS. Helping them to reduce feelings of fear and stress will allow for greater success in all areas of their life.

Patrice Carroll, Manager of PWS Services

IPWSO NEWS

PLEASE VISIT IPWSO's blog and become A FRIEND of IPWSO. Don't miss the important article on Constipation in individuals with Prader-Willi Syndrome. You are welcome to communicate directly with the Communications Director of IPWSO, Linda Thornton. She will welcome your comments. http://ipwso.blogspot.com

CARING FOR YOUR ADULT DAUGHTER OR SON AT HOME? NEED SOME HELP?

Please share these FAMCARE articles with others!
The following articles are available from the FAMCARE page on IPWSO'S website:

- 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 2014
- STORY-TELLING – June 2014
- "I WANT THE SAME" – March 2015
- STRENGTH IN BOUNDARIES – June 2015

Articles are also available from the chairperson PWSA (SA): chairperson@praderwilli.org.za

NEW RELEASE OF PWS FILMS

Earlier this year IPWSO and the Prader-Willi Syndrome Association of Ireland joined forces to make four educational films. Please share these and use them for educational purposes.

The following are available on YouTube:

- A New Diagnosis of Prader-Willi Syndrome: by Dr Susanne Cassidy
- Growth Hormone Therapy in Prader-Willi Syndrome: by Dr Charlotte Höybye
- Dietary Management in Prader-Willi Syndrome: Introduced by families
- Behaviour and Mental Health in Prader-Willi Syndrome: by Professor Tony Holland

SAVE THE DATE: 9TH IPWSO CONFERENCE

The International Prader-Willi Syndrome Organization (IPWSO) and Foundation for Prader-Willi Reseach, Canada are proud to announce that the 9th IPWSO Conference will take place in Toronto, Ontario Canada, July 20 – 24, 2016.
Mark the dates on your calendar and start making plans to attend.
REGISTRATION NOW OPEN!!

PLEASE RENEW YOUR MEMBERSHIP

Payment of your membership fee is now due.
Thank you to those members for prompt payment.

Please contribute to ***People with Strength.*** Whether you are a parent, medical practitioner, therapist or relation, please send your contribution, questions or suggestions to:
PWSA (SA), PO Box 2399, Brooklyn, 0075
or email: chairperson@praderwilli.org.za

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.

ACTIVITIES OF THE ASSOCIATION:

- The association provides support to parents and others who care for children and adults with PWS.
- The association publishes a newsletter, *People With Strength* to update its members and other interested persons regarding news and the latest developments in the field of the Prader-Willi syndrome.
- The association disseminates important educational material such as information provided by the *International Prader-Willi Syndrome Organisation*, (IPWSO) and other sources to its members and others involved.
- The association organizes workshops, seminars or conferences from time to time on the latest research and effective management of PWS.
- The association organizes an annual general meeting to deal with official matters at which occasion parents are also afforded the opportunity to socialise and share their ideas and experiences with other parents in similar situations

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
Visit our website: www.praderwilli.org.za

COST OF MEMBERSHIP

Registration fee R50.00 (once-off payment)
Annual membership fee R200. R220 for 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: Your SURNAME

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).