People With Strength

Newsletter for parents by parents

Volume 17 Issue 4 **AN INVITATION** November 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

PRADER-WILLI SYNDROME ASSOCIATION (SA): AN INVITATION

PWSA (SA) is a support group for parents whose children are diagnosed with PWS. We invite health care professionals to join PWSA (SA). This includes everybody who is interested in the Prader-Willi syndrome and others who are involved in the caring of persons with PWS. There is a need of health care professionals who can support and guide parents. The ideal will be to have a multi-disciplinary clinic in every province where parents and caregivers can receive the necessary support and guidance.

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WHAT IS PRADER-WILLI SYNDROME?

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

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FROM THE CHAIRPERSON

Dear Readers

2014 was a special year. In this last issue for 2014 I'm sharing some information with you about this exceptional year. This newsletter also contains other interesting educational articles. Everybody should find something worthwhile.

The presence of Dr Janice Forster and the two lectures she delivered on 3 October in Cape Town were definitely a highlight. Promising feedback was received and it was indeed an educational and awareness opportunity. A wide range of professions was represented; inter alia, dietetics, physiotherapy, occupational and speech therapy, social work and the nursing profession. Staff members of House Jonathan, Adams Farm, Iris House, Woodside Special Care Centre, Bel Porto School, Agapé, Ocean View School and the Oasis Association were also interested. The Division of Human Genetics of the University of Cape Town was well represented by geneticists, students, post graduate students and genetic counsellors. There were also a paediatric neurologist and a registrar in neurodevelopment present. Ten families also attended the lectures.

PWSA (SA) expresses their thanks to Dr Karen Fieggen of the Division of Human Genetics of the University of Cape Town for helping with the logistics. Please see pictures elsewhere in this newsletter and some personal details of Dr Janice Forster.

A special edition of *People With Strength* was distributed at the lectures delivered by Dr Forster. Afterwards an electronic version was distributed to member and others. If you haven't received a copy, please contact the chairperson at: rikadup@mweb.co.za.

Meet Molelekeng Sethuntsa, a clinical psychologist at the Dr George Mukhari Academic Hospital in Ga-Rankuwa, Pretoria. Ms Sethuntsa was a guest at the annual general meeting held in August and she had the opportunity to discuss her proposed study and research. She is busy with her doctorate and wants to conduct a study to develop a therapeutic intervention that will be most effective for individuals diagnosed with PWS and their primary caregivers. Members will be contacted in March 2015 with an invitation to take part in this research project.

Welcome to Maryke van der Hoogt, a dietician and currently a member of PWSA (SA). Read more on Maryke and her interest is the Prader-Willi syndrome. Parents may contact her for an appointment.

Debra Sowter of Iris House provides information on a special centre for children with special needs in the Western Cape. Read more about this dedicated team and the holistic approach they follow.

PWSA (SA) invites health care professionals to join our association. This includes everybody who is interested in the Prader-Willi syndrome or others who are involved in the caring of persons with PWS. There is a need for health care professionals to support and guide parents. The ideal will be to have a multi-disciplinary clinic in every province where parents and caregivers can receive the necessary support and guidance.

Welcome to the Grix and Pillay families who have joined the association recently.

Dr Janice Forster mentioned the TRAIN concept in her lectures to diminish anxiety, insecurity and non-compliance in persons with PWS. Read *Implementing the Train* in this issue. The train with its trucks are visually placed on a straight line and it represents the activities for a day. The implementation of the TRAIN is explained step by step.

Information about gastric and intestinal problems regarding persons with PWS has been previously published in newsletters. Janalee Heinemann who is the Director of Research & Medical Affairs, PWSA (USA) gives some feedback on research in this regard. The oesophagus, stomach and colon are discussed. Note her warning when constipation occurs.

The end of a year is always typified by different parties and special events. When receiving an invitation a parent is always worried about all the food which will be available. Only the thought thereof brings anxiety. Patrice Carroll of Latham Centres gives some tips in this regard.

The annual general meeting was attended by only seven families. Some extended family also attended and they have found the meeting very informative. Thanks to Dr Engela Honey. Her presence is always a big support to the families. Look at some pictures in the photo gallery.

A well-earned summer holiday to you all! Enjoy your togetherness with family and friends.

Warm regards
Rika du Plooy

VAN DIE VOORSITTER

Liewe Lesers

2014 het gesorg vir ‘n spesiale hoogtepunt! In hierdie laaste uitgawe vir die jaar deel ek graag inligting oor die besondere geleentheid. Ander waardevolle inligting is ook in hierdie nuusbrief beskikbaar en ek vertrou dat daar iets vir almal sal wees.

Die twee lesings wat dr. Janice Forster op 3 Oktober in Kaapstad aangebied het, was beslis ‘n hoogtepunt. Goeie terugvoering is ontvang en hierdie was ‘n wonderlike leer-en bewusmakingsgeleentheid. ‘n Wye verskeidenheid beroepe was verteenwoordig, o.a. dieëtkunde, fisioterapie, arbeids-en spraakterapie, maatskaplike werk en die verplegingsberoep. Personeel van House Jonathan, Adams Farm, Iris House en Woodside Special Care Centre, Bel Porto Skool, Agapé, Ocean View School en die Oasis Association het ook belanggestel. Die Afdeling Mensgenetika van die Universiteit van Kaapstad was baie goed verteenwoordig deur genetici, studente, nagraadse studente en genetiese raadgewers. Verder was daar ‘n pediatriese neoroloog en ‘n kliniese assistant in neuro-ontwikkeling. Tien families het ook die geleentheid bygewoon.

PWSV (SA) is baie dank aan dr. Karen Fieggen van die Afdeling Mensgenetika van die Universiteit van Kaapstad verskuldig vir haar hulp met die logistieke reëlings. Iewers in die nuusbrief foto’s en iets meer persoonlik oor Janice Forster.

‘n Spesiale uitgawe van *People With Strength* is met dr. Forster se lesings uitgegee en is ook elektronies versprei. Enigeen wat nie hierdie spesiale uitgawe ontvang het nie, kontak gerus die voorsitter: rikadup@mweb.co.za

Ook in hierdie nuusbrief meer oor Molelekeng Sethuntsa, ‘n kliniese sielkundige by die Dr. George Mukhari Akademiese Hospitaal in Ga-Rankuwa, Pretoria. Me Sethuntsa het as gas die Algemene Jaarvergadering bygewoon waar sy die geleentheid gehad het om die lede oor haar studie in te lig. Molelekeng is besig met ‘n doktorale studie en sy rig haar navorsing op persone wat met PWS gediagnoseer is en hulle ouers. Vroeg 2015 sal ‘n skrywe deur PWSV (SA) aan lede gerig word oor die navorsing, asook ‘n uitnodiging om deel te wees van hierdie studie.

‘n Spesiale welkom aan Maryke van der Hoogt, ‘n dieëtkundige en nou lid van PWSV (SA). Maryke stel baie belang in die Prader-Willi-sindroom en ouers is welkom om haar te kontak vir ‘n afspraak.

Debra Sowter van Iris House verskaf inligting oor die versorgingsfasiliteit vir kinders met spesiale behoeftes. Hierdie goed opgeleide personeel het ‘n holistiese benadering met ondersteuning op ‘n persoonlike en professionele vlak.

PWSV (SA) nooi graag professionele persone om by die Vereniging aan te sluit. Dit sluit enigeen in wat ‘n belangstelling in die Prader-Willi-sindroom het of persone wat by ‘n persoon met die sindroom betrokke is. Daar is ‘n behoefte in Suid-Afrika aan professionele persone wat die nodige ondersteuning en leiding aan ouers kan gee. Die ideaal is natuurlik om ‘n multi-dissiplinêre kliniek in elke provinsie te hê, waar ouers die nodige ondersteuning en leiding kan ontvang.

Baie welkom ook aan die Grix en Pillay gesinne wat aangesluit het.

Dr. Janice Forster het o.a. in haar lesings die begrip van ‘n *TRAIN* gebruik om anstigheid, onsekerheid en koppigheid by die persoon met PWS te verminder. Lees die artikel *Implementing the TRAIN* wat sy hieroor geskryf het. Die trein met sy trokke word visueel op ‘n reguitlyn voorgestel en gee struktuur aan aktiwiteite deur die dag. Die gebruik van die *TRAIN* word stap vir stap verduidelik.

Inligting oor gastriese- en dermkanaalprobleme by die persoon met PWS het voorheen al in die nuusbrief verskyn. Janalee Heinemann, wat verantwoordelik is vir mediese sake en navorsing by PWSA (USA) gee kortliks terugvoer oor navorsing wat in hierdie verband gedoen is. Die slukderm, maag en kolon word bespreek. Let veral op na die waarskuwing wat sy gee oor wanneer hardlywigheid voorkom.

Die einde van die jaar word altyd gekenmerk deur afsluitingsfunksies, familiebyeenkomste, ens. Wanneer ‘n uitnodiging ontvang word, is die eerste ding waaraan ouers dink die oorvloed en verskeidenheid eetgoed - dit is ‘n bron wat angstigheid by ouers veroorsaak. Lees gerus wat Patrice Carroll van Latham Centers aanbeveel.

Die Algemene Jaarvergadering in Augustus is slegs deur sewe gesinne bygewoon. Wat bemoedigend was, is dat ‘n gesin deur familie vergesel was. Die familie het die geleentheid baie leersaam gevind. Ons het waardering vir Dr Engela Honey wat met haar teenwoordigheid steun aan ouers bied. ‘n Klompie foto’s in die foto gallery.

Aan almal ‘n welverdiende somervakansie. Mag die samesyn met familie en vriende ‘n geseënde tyd wees.

Opregte groete
Rika du Plooy.

HIGHLIGHT OF THE YEAR!

An overview of Prader-Willi syndrome: Food is the tip of the iceberg

By Dr Janice Forster

Thank you to Janice Forster for giving up some of her time in Cape Town to come and share her expertise and experience with South African health professionals and families. Janice and her husband were on a safari in Africa and on their return to the USA they had a stopover in Cape Town. (Read more under *From the Chairperson*)

Dr Karen Fieggen (left) and Dr Janice Forster (right). Dr Forster is a Child and Adolescent Psychiatrist and also co-founder of the *Pittsburgh Partnership, specialists in PWS.* Dr Karen Fieggen is a senior specialist and Head of Clinical Genetic Services at the Division Human Genetics at UCT.

BY THE WAY…….

Written by the chairperson, Rika du Plooy

Janice and I met for the first time in Cluj-Napoca, Romania attending the 6th International Prader-Willi Syndrome Conference in 2007. Janice said that she then knew that someday she would visit South Africa as part of her new and incredible, international, "extended" family of PWS.

Janice explained this longing to visit SA and I quote:
My father was a mechanical engineer for MESTA, a Machine Company in Pittsburgh PA (USA) who designed machines for the steel industry. He designed and then supervised the installation of a rolling mill for ISCOR in Johannesburg. My mother joined him for the last two years of his residence there and they had a flat in Vanderbijlpark. When I visited them, we took a trip across the Transvaal to the Eastern coast of SA through Durban, Port Elizabeth and Cape Town. This was my first trip out of the USA, and I have always thought of South Africa as my "second home". Needless to say, the Cape Town that I remembered changed dramatically! Now it is a beautiful, modern, vibrant city that welcomes change and diversity.

This was a very special trip for me, as I was able to walk where my parents did so long ago! Thank you so much for making the lectures possible. It was a wonderful, meaningful experience meeting your PWS professionals and families.

Welkom aan die volgende nuwe lede:

Quintus en Liezl Grix van Potchefstroom. Hulle dogtertjie, Larike is 18 maande oud.

Maryke van der Hoogt, 'n dieëtkundige van Pretoria met 'n belangstelling in die Prader-Willi-sindroom.

We also welcome Cedric and Caren Pillay of Brackenfell, Cape Town. Their daughter is nine and at the Paarl School.

Dr Engela Honey from the Department of Genetics at the University of Pretoria with Ms Molelekeng Sethuntsa a clinical psychologist. Ms Sethuntsa explained her proposed study to members at the AGM August 2014.

MEET MOLELEKENG METHUNTSA

Clinical Psychologist, Dr. George Mukhari Academic Hospital, Pretoria

I qualified as a Clinical Psychologist in 2009. I am currently working at the Dr. George Mukhari Academic Hospital in Ga-Rankuwa, Pretoria. Amongst other things I do psychological assessment and provide psychotherapy to in and outpatients. I am now registered for my doctorate with the Department of Psychology at the University of South Africa.

I am conducting a study to develop a therapeutic intervention that will be most effective for individuals diagnosed with PWS and their primary caregivers. In order to achieve this it is important to learn about family's individual experiences. This will be achieved through clinical interviews and implementation of different interventions. I hope to start with the interviews and interventions around March 2015. Families will be invited to participate through the PWSA (SA). Once the family gave their consent I will then start with the process of interviews and intervention.

The time frame will depend on the intervention that may be sort. The participants will be seen at Dr. George Mukhari Academic Hospital, University of Pretoria. For those families who are not able to access the venues, I will visit them in their homes. Interviews and intervention will be recorded in order to ensure that data is not lost. Their individual responses will be kept completely confidential.

I hope that results of the study will share light to psychologists and other health professionals as well as add to the existing literature. I also hope that the results will assist the South African government to provide comprehensive care for the individuals with PWS and their families. Contact: **082 957 2595**

MEET MARYKE VAN DER HOOGT

Dietician, Little Company of Mary Hospital, Medical Centre, West Wing Level 2, Pretoria

My name is Maryke. I am a dietician situated at Little Company of Mary Hospital in Pretoria. I qualified in 2010 and am currently doing my MSc in dietetics. I was privileged enough to meet my first patient with Prader-Willi syndrome in a government hospital in 2011. At the time I had no clue about the condition as at undergraduate level we don't learn a lot about the condition. This boy was around 12 years old and referred for weight loss. He was adorable and fell asleep in my chair, I remember thinking my session must be so boring that he decided rather to sleep than engage with me....!

Maryke van der Hoogt is a dietician and has an interest in Prader-Willi syndrome.

After that day I started reading up on Prader-Willi syndrome, not only the dietary side of things but also the emotional and behavioural aspects. Kids with Prader-Willi syndrome and their families need a bit of extra support and understanding when it comes to diet therapy, it's not just eat less and exercise more and you'll lose weight.

Working with children is my passion, other than children with Prader-Willi syndrome I do mainly endocrine conditions like type 1 and type 2 diabetes. I am a member of the PWSA (SA) and I am excited to be part of the Prader-Willi syndrome family. Contact: **072 369 4865**

ACKNOWLEDGEMENTS 2014

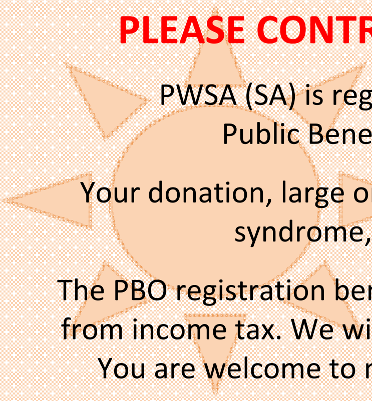
PWSA (SA) would like to acknowledge with gratitude the goodwill and support of:

Our volunteers who offer their time and dedication:

Elsa Volschenk for her involvement in assisting with the newsletter
Wilna Basson for taking care of the Library and educational material
Karin Clarke and Magdaleen Kloppers for the birthday cards
Johan and Elmaré Mostert for the annually preparation of the financial statements
Dawid Basson, the webmaster of the website and also responsible for Facebook.
Francis Morrison for the translation of *Van die Voorsitter*
Jan Els, chartered accountant for auditing the income and expense accounts
Dr Engela Honey, the medical advisor of the Association, who is always available

Members for your loyalty and support. Thank you for prompt payment of membership fees
All those who contributed to *People With Strength*
IPWSO for continuously forward information regarding important PWS issues
All other **PWS Associations** for assistance and sharing of knowledge
Our donors for your support to our Association
afrihost.com for hosting the Association's website

PLEASE CONTRIBUTE AND HELP TO MAKE A DIFFERENCE!

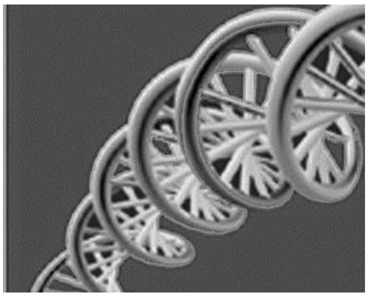


PWSA (SA) is registered as a non-profit organisation (No. 035-837 NPO)
Public Benefit Organisation (PBO Exemption no.930 016 853).

Your donation, large or small, provides vital support to individuals with Prader-Willi syndrome, their families and others in the supportive team.

The PBO registration benefits donors and all donations made to PWSA (SA) are exempt from income tax. We will issue an official certificate for donations of R100.00 or more. You are welcome to make a direct deposit. Please ensure that your surname and cell number are included as reference.

The Australian and New Zealand Prader-Willi Syndrome Associations invite you to Melbourne in April 2015, to join them at the 3rd Asia Pacific Prader-Willi Syndrome Conference. Email: conference@pws.org.au.



Pittsburgh Partnership

Specialists in Prader-Willi Syndrome

IMPLEMENTING THE TRAIN

By: Linda M. Gourash, MD and Janice L Forster, MD of the Pittsburgh Partnership

Specialists in Prader-Willi Syndrome

The TRAIN description

The TRAIN (Tool for Reducing Anxiety, Insecurity and Noncompliance) is an eco-environmental tool to manage symptoms of cognitive rigidity, perseveration, and stress sensitivity that are associated with the PWS behavioral phenotype. A visualized, linear schedule informs the PWS person what activities and tasks are expected to occur throughout the day. By arranging the daily schedule along a linear queue, the “engineer” or carer alternates the order of activities from non-preferred (resistance) to preferred (reward, i.e., mealtime). This flow of activities manages transitions to keep the person with PWS moving through their day, resulting in greater productivity, mastery and improved self-esteem.

Relevant background

Studies show that persons with PWS have certain cognitive characteristics that suggest that visual display of information can improve on-task behavior. Individuals with PWS have deficient working memory, the cognitive skill that allows all of us to hold multiple tasks in mind in order to execute multi-task objectives. In addition, individuals with PWS are mostly visual learners, so a visual display of sequential tasks helps the person know what to do next, reducing uncertainty and anxiety. Finally, individuals with PWS do not have a good sense of quantity, time or space, which leads to problems with arithmetic and perception of amount of time passing. However, they are able to understand calculations on a number line and a sense of time passing on a time line. Hence the linear, visualized schedule, defining the order of activities across the day, compensates for phenotypic deficiencies, and allows the person with PWS to focus on the activities at hand without preoccupation with what might happen next. Identifying when meals, snacks and preferred activities will occur during the day not only helps to define *psychological food security* but *everything security* as well. When people with PWS have food security, they can focus their minds on other adaptive tasks.

Designing the TRAIN

STEP 1

With a pencil and paper, write down the schedule for a typical day as it currently exists for your child with PWS, emphasizing the order of activities and an estimate of the amount of time devoted to each one. Make any comments that explain the amount of time expended for each activity, if relevant. This is an exercise. Not too many families can actually complete this schedule and keep it running consistently.

Note: If your schedule looks like this, and your child with PWS knows or refers to this schedule, you may not need the TRAIN!

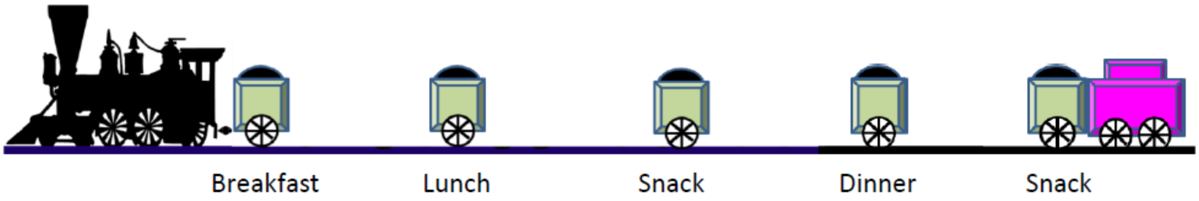
Daily activity:	**Time of day:**	**Estimated duration:**	**Comments:**
Getting up in the morning	6 am	1 hour	Doesn't awaken with alarm; never wants to get out of bed
Getting washed and dressed	7 am	1 hour	Dawdles; needs 1:1 coaching; argues about what to wear
Breakfast	8 am	15 minutes	
Gets on the school bus	8:30 am	15 minutes	Always slow
Gets off the bus	4 pm	5 minutes	
Snack	4:05 pm	10 minutes	
Walks the dog with mom	4:15 pm	30 minutes	
Videogames	4:45 pm	1 hour	
Dinner	6 pm	30 minutes	
Homework	7 pm	1 hour	Slow; needs 1:1 coaching
Showers and gets ready for bed	8 pm	1 hour	Dawdles; uses too much water and body wash
Bedtime snack	9 pm	15 minutes	
Bedtime	9:30 pm	1 ½ hours	Never wants to go to sleep!

STEP 2

Create the track for the linear schedule for the day, starting with the morning (near the engine) and ending with bedtime (near the caboose). It is helpful to have "train cars" that are either stylized to represent meals and activities (for younger children) or small white eraser boards to write on for meals and activities for older persons. Some parents/carers use a velcro strip to fasten the train cars.

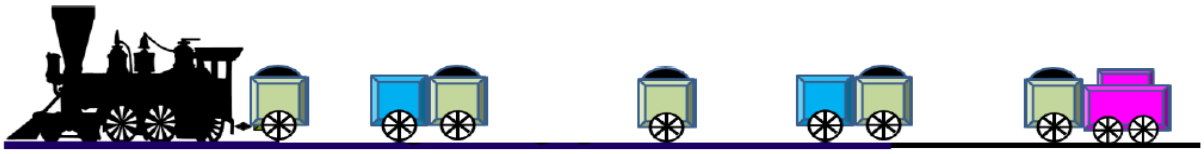
STEP 3

Place the mealtimes and snack on the track.



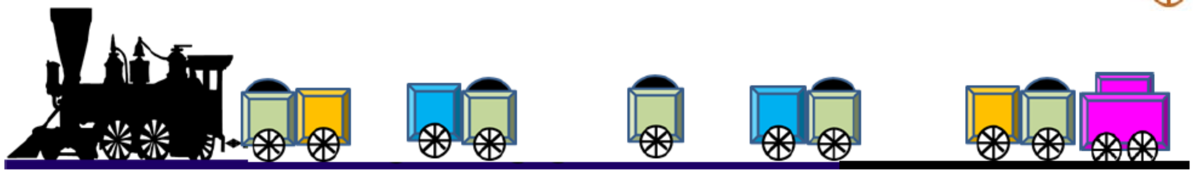
STEP 4

Place the exercise times on the track. Exercise is usually right before the meal or snack.



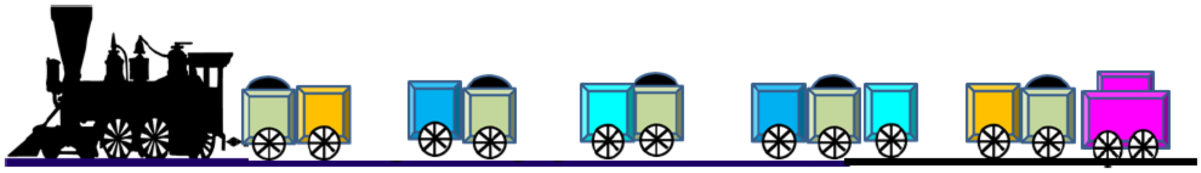
STEP 5

Place the exercise times on the track. Exercise is usually right before the meal or snack.



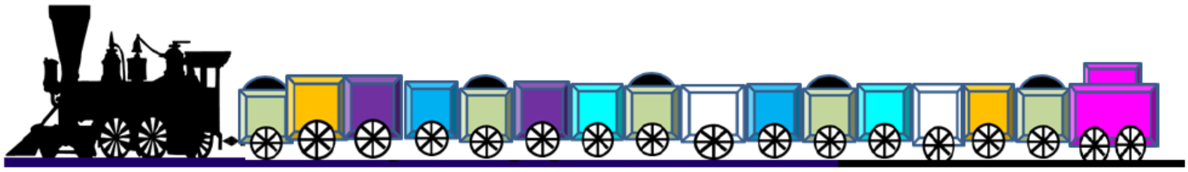
STEP 6

Place the opportunities for leisure time on the track; there can be more than one time for this in the day.



STEP 7

Place non-preferred activities, such as school work or chores (laundry), before the leisure time.



Remember, it is the order of the events in the day and not the clock time of day that is critical for the running of the TRAIN. The engineer determines the activities and the order of events in a given time frame. There can be several engineers of the TRAIN across the day; for example, one engineer for the morning routine, one engineer for school or work, and one engineer for the evening routine. The person with PWS should always know who the engineer is.

Find a place in the home, school or work environment to display the TRAIN. It is important to have the schedule in a prominent place, so the person with PWS can refer to it.

Implementing the TRAIN

Step 1. Explain the concept of the TRAIN to the person with PWS as follows, positive and upbeat: “We have an exciting new way to keep you “on track” with your daily schedule! It’s called the TRAIN. We know that you like to know what is happening in your day. So, just like a real train, all the events in the day are put on the schedule so that you can see what comes next and what you’ve already accomplished. This is just like the rail cars on a real train track.”

Step 2. Show the linear schedule of the TRAIN without the rail cars, and demonstrate in real time where they are in the schedule just now.

Just as you conceptualized your design of the TRAIN, you will implement the schedule for the person with PWS in the same way.

Step 3. Show where the meals and snacks appear on the linear schedule. Practice this for one week. If you are using a white board, you can actually write in the menu for that meal.

Step 4. Show where the exercise falls on the schedule. This should always be before a meal of a snack. Implement this for one week.

Step 5. Show where grooming and dressing occur on the schedule.

Step 6. Show where leisure “self-guided fun” activities fit into the schedule.

Step 7. Show where chores and school work occur on the schedule.

Establishing contingencies

For every preferred activity, there should be an alternative determined in advance. For example, if an outing is planned but not possible due to weather or transportation problems, a movie, game or extra leisure time can be substituted. The person with PWS should know this alternative in advance.

The Day Stops Here!

When a tantrum or shut down impedes the flow of the day, the schedule stops until the person’s behavior reorganizes. When the person is ready to get back on the TRAIN and resume their day, the order of activities on the schedule should be followed exactly, but the duration of time in the activity is decreased, and the expected outcome of activities is diminished. For example, exercise time might be decreased from 1 hour to ½ hour, TV time is similarly reduced, and instead of doing two loads of laundry, only one load is required. The meal time schedule may be delayed. If the tantrum or shut down lasts long enough to miss a meal, the person can still get their meal if they pull themselves together. But if hours go by and the meal cannot be saved, the person is offered a predetermined “no frills meal” with fewer components and condiments to equal the calories they lost. The objective is to catch up to the daily schedule by the next meal, if possible, and most certainly to complete the daily schedule before bedtime, because the next day should start anew.

Tantrums and shut downs should receive low attention (matter-of-fact demeanor; acting as if bored), but the safety of all should be assured. The PWS person in a shut down or tantrum should never be ignored, however. People with PWS have a poor sense of time passing. Incidents can last 5 minutes, 5 hours, or 5 days. While providing low attention and low affect, the carer is also supportive of the person in shut-down, reminding them that time is passing, and that they really want the person to get to experience their next preferred activity. The context of the event is an important consideration also. A person with PWS could be angry at one carer, but interact with another as if nothing happened. This characteristic can be used adaptively to get the person back on the TRAIN.

Explain *The Day Stops Here!* to the PWS person in the following way, in words that are appropriate for their developmental level: “We know that every now and then you have a [tantrum or meltdown].” *It is important to use the words that the person knows for these behaviors.* “When you have [a behavior], it stops the TRAIN; that means, everything that is supposed to happen is postponed, including meals and snacks, until you get your behavior organized. There is good news; we will hold your meal or snack for a while; but if your behavior goes on too long, we will give you a drink that has all the calories of a meal. So, together, we will do everything we can to get your behavior back on track. After your behavior gets back to normal, you can get back on the TRAIN and resume your daily schedule.”

Troubleshooting

Dawdling (due to deliberate slowness, getting lost in thought, or being unable to remember what to do next) and difficulty leaving one activity to begin another (shifting) are common manifestations of the cognitive style associated with PWS. When the next activity is a rewarding one (something the person enjoys doing), the transition is usually smooth and timely.

1. If there are times in the day when the person shuts down, melts down, or the transition is rough, change the sequence of activities by scheduling a *more preferred activity* at the time of the meltdown to improve the flow of the day.
2. If you have optimally managed the sequence of the activities in the day, increase the density of reward by scheduling two preferred activities in a row to get over the rough transition.
3. If you have increased the density of the reinforcing activities, and there are still problems, implement a behavior plan as follows: Tell the person that you know (and demonstrate on the schedule) that they seem to have trouble at this particular time of day. Tell them that whenever they make a successful transition over the rough spot, they will earn a ticket or token. This small reward usually represents bonus time for a preferred activity (e.g., screen time, collections, time with a family member or peer, etc.).

FROM TOP TO BOTTOM GI ISSUES PWS

By Janalee Heinemann, PWSA (USA) Director of Research & Medical Affairs

From: The Gathered View ~ Prader-Willi Syndrome Association (USA) July-August 2014

We are learning more about the fact that people with Prader-Willi syndrome are at risk throughout the G.I. (gastro-intestinal) tract system.

Esophagus:

We do know in our medical and study of death surveys that 39% of families reported choking episodes and 12/152 PWS deaths were choking deaths (not infants or toddlers but much older than the norm with an average age of 24). This does not include a very recent choking death reported. There is a swallowing study going on right now by Dr. Gross and Dr. Cherpes and sponsored by PWSA (USA) where preliminary results have shown that participants had a significant amount of oesophageal residue but they thought the food all went down, creating a risk factor for aspiration/choking.

Stomach:

Dr. Ann Scheimann reports that the PWS stomach is thin-walled and doesn't produce a lot of fluid. She and Dr. Klish are doing a G.I. study sponsored by PWSA (USA) which has shown that a significant number of people with PWS have slow stomach emptying/gastroparesis. We have been aware for some time about the risk of G.I. necrosis and perforation deaths in PWS, and now suspect the gastroparesis may be adding to this risk factor.

Colon:

A new Denmark study titled, "*A descriptive study of colorectal function in adults with PWS: high prevalence of constipation*," confirms what we have known all along, that our children and adults with PWS have a high prevalence of constipation. The G.I. transit time was over three days in 5/21 with PWS and none of the controls. 8/20 (40%) fulfilled the criteria for constipation. 12/20 (65%) had fecal mass in the rectum vs. 3/25 (12%) of the controls.

My personal concern, due to taking the medical calls for 17 years, is that even though we have known unofficially for a long time that constipation is not at all unusual with PWS, when a person with the syndrome goes to the ER with G.I. symptoms, often they take an x-ray and see that the person is constipated and think that is the only problem, thus often overlooking a more serious problem. Also, constipation may increase the risk for rectal picking behaviours.

Parents will sometimes ask, "Since growth hormone strengthens muscles, shouldn't it strengthen the motility of the entire G.I. tract?" Unfortunately, the answer is no. As we learn more about these G.I. issues, and document them through studies, hopefully we can find some resolutions for the safety of our children and adults with Prader-Willi syndrome.

LEARNING TO TYPE

Kate Beaver, Family Support Counsellor PWSA (USA)

***The Gathered View* ~ Prader-Willi Syndrome Association (USA), July-August 2013**

Working with schools and helping teachers to identify what causes our children anxiety in the school setting has been educational for our staff. We have learned that one thing the school can do to help decrease anxiety and to build long term skills is to teach children how to type and use a word processing program. Our children, who usually want to please teachers, become anxious when they are asked to practice writing or to write out spelling words or other subject matter. It seems that the children perseverate on the process of writing the words to look like they have been typed or to look like how the teacher wrote them. In doing so, our children lose sight of what they are learning and become anxious to recreate the word perfectly. Teaching typing to young children even in first grade helps lower anxiety and also gives them a long term skill. Writing will always be important, but if we can separate writing from the other learning, it's a win-win scenario.

TIP OF THE WEEK: SHOULD WE STAY OR SHOULD WE GO?

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

You will ask yourself this question dozens of times between now and the end of December. The holiday season brings parties, gatherings, and special events that stray from your normal routine. In other words, they bring anxiety. Some family members will insist that you come to their parties, while others will ask you not to attend with your child. Both scenarios are equally as stressful and will leave you feeling guilty regardless of your decision; but it doesn't have to be that way. Here are some things that you can do to make the holiday season what it should be, festive and full of wonder:

1. Be realistic. You know your child better than anyone. If you know that your child cannot handle a large party, then do not set them up to fail. Plan a smaller get together where your child can be included or arrange for your child to attend the larger party for a short amount of time.

2. Use social stories to prepare your child for what to expect. More exposure to change does teach flexibility in the long run. Just be sure you have the necessary supports to manage
   your child if he or she becomes overwhelmed during the event.

3. Give yourself permission to stay home. If you know that attending a party will be too much, then don't go. If your family is discouraging you from bringing your child, opt to

plan a smaller event where they can see for themselves how amazing your son or daughter is

4. Most importantly, plan time to enjoy yourself and your family during the holidays. If that means keeping it low key, then do so. Whatever you plan, just remember that the holidays are a time to be with family and friends in whatever way feels most comfortable for you and yours.

Patrice Carroll
Manager of PWS Services

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).

PHOTO GALLERY

The AGM was held on Sunday 24 August 2014 at the Vriendekring Bowls Club, Pretoria. It was a successful get together, although few families attended.

It was a lovely sunny day and the children enjoyed the ball games.

Extended family joined the Senne family. Metsiatsile with her grandmother (left) and aunts.

Every year Willemien and Marise enjoy the card games. They are joined by Soné Wilken, a volunteer.

The siblings were part of the fun. Elisna Jacobs a volunteer, assisted.

The following pictures were taken at the Red Cross War Memorial Children's Hospital, Cape Town on Friday 3 October 2014. Dr Janice Forster delivered two lectures which were very well received.

A wide variety of health professionals as well as parents wait for the lecture to start.

Janice explains the key role of the hypothalamus in the pathology related to Prader-Willi syndrome.

Families got the opportunity to get to know each other. From left to right: David Clarke, Caren and Cedric Pillay, and Karin Clark.

Kirsten and Terence Youlding Hall (Gauteng) with Dr Janice Forster as well as David and Karin Clarke (Cape Town) on the right.

IPWSO NEWS

SAVE THE DATE: 9TH IPWSO CONFERENCE:

Building Global Community **- July 20 – 24, 2016, hosted by FPWR Canada**

Together we are "Building Global Community" to eliminate the challenges of Prader-Willi syndrome. This theme symbolizes the importance of bringing all members of the PWS community together to unite under one global mission. 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 at the beautiful Sheraton Centre Toronto located in heart of downtown Toronto.

This Conference will enable parents, caregivers, scientists and professionals to join together to share expertise and collaborate best practices in order to improve the quality of life of all individuals with Prader-Willi syndrome. Organizers expect more than 600 delegates from around the world and our Canadian culture will embrace all nationalities and provide a rich and inclusive environment.

There will be four key program streams: Scientific (July 20-22), Caregivers, People Living with PWS, and Parents (July 22-24). The Scientific Program Call for Abstracts will be announced through Co-Chairs, Dr Theresa Strong, Dr Rachel Wevrick and Dr Tony Holland.
Mark the dates on your calendar and start making plans to attend.
Our online registration form and additional Conference details will be released in 2015.

RESEARCH SUPPORT FOR MEDICAL PROFESSIONALS

The latest scientific papers on PWS, collated by the International PWS Organisation Scientific Advisory Board, which is headed by PWSA UK President, Prof Tony Holland, have been posted on IPWSO's website: www.ipwso.org

IRIS HOUSE CHILDREN'S HOSPICE

Written by Debra Sowter

Debra attended the lectures delivered by Dr Janice Forster

Iris House Children's Hospice is a living Children's Hospice providing loving care for special needs children with life threatening illness or life limiting conditions in the Western Cape. When we say living we mean that the majority of our children suffer from conditions that are non curable but are not necessarily terminal. Iris House is a happy place where we celebrate the unique abilities of each of our children.

The staff at Iris House promote the concept that every life is a gift, and valuable.

We understand that the emotional and financial burden of raising a child with special needs is enormous and we focus on holistic family care that factor in each and every member of the family. Our aim is to enhance the quality of life for the special needs child, offering a high quality free service with both personal and professional support. In partnership with the child and family, each child's individual needs will be met; those of their family and significant carers will be recognised and respected. We aim to uplift communities by training and employing carers to provide community based respite care.

We are committed to extending our community based care model into more areas of the Western Cape and in the long term the whole of South Africa. Community based care is the cornerstone of our organisation, however we have a larger vision. We aim to provide end to end respite care and support for our families. We have secured a building which we hope to open in early 2015 and there we will create a home away from the home environment where children can attend play groups and therapy sessions to enable them to be the best they can be.

To find out more about our services please e-mail debra@iris-house.org or alternatively please call the office at 021-559 0800. Visit our website www.iris-house.org and our face book page to take note of our latest family events.

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

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