



People With Strength

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Prader-Willi Syndrome Association of South Africa

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Newsletter for parents by parents

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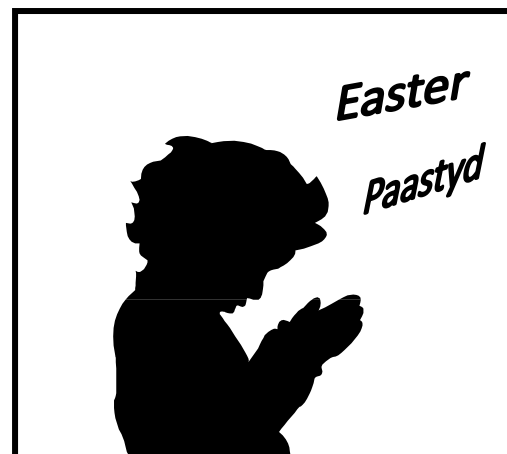
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2011

*Jaar van bewusmaking van die Prader-Willi-sindroom / syndrome
Understanding through awareness...*

What is Prader-Willi syndrome?

Genetic disorder
Low muscle tone
Short stature
Obesity
Increased appetite

Van die voorsitter...



Liewe Lesers

Die jaar stap aan en ek vertrou dat 2011 vir almal goed begin het. In baie lande was daar erge natuurrampe en ons gedagtes gaan veral uit na die inwoners van Nieu-Seeland en Japan wat deur aardbewings getref is. Ek het met Linda Thornton kontak gehad en volgens haar is geen families met PWS betrokke in Christchurch getref nie. Almal is egter geskok en getraumatiseer en dit gaan 'n tyd neem om na normaal toe terug te keer. Eiko Shoji, die vise-president van die PWSV (Japan) berig dat hulle van een familie nie rekenskap kan gee nie. Ons is almal terdeë bewus van die ontstellende omstandighede in Japan - die PWS families en almal in nood, kan verseker wees van ons meelewing.

Die hoofdoelstelling van die Vereniging vir 2011 is om uitvoering te gee aan 'n groter bewustheid vir die Prader-Willi-sindroom. Dit was een van die motiverings wat aan die Willowton Groep voorgelê is, toe daar vir die R100 000 aansoek gedoen is. Vroeg in die jaar het geleentheid hulle voorgedoen en die eerste stap om uitvoering aan hierdie doelstelling te gee is 'n volkleur inligtingsblad wat in die Maartuitgawe van die tydskrif *EZEMPILO, Health Matters* gaan verskyn. Die tydskrif word maandeliks gratis versprei en is die inisiatief van die Departement van Gesondheid in KwaZulu-Natal. Wat dit uitsonderlik maak is dat daar vir die eertse keer inligting oor die sindroom in 'n swart taal, nl. Zoeloe gaan verskyn. Inligting oor die sindroom word ook vroeg April 2011 aan kongresgangers (persone wat by kinders met leergestremdhede betrokke is) by 'n SAALED kongres in Kaapstad verskaf. 'n Verdere oogmerk is om aan al die finalejaar mediese studente aan die onderskeie universiteite, elk 'n *IPWSO Medical Alert Booklet* te gee. Daar is nog idees in die pyplyn, maar lesers word uitgenooi om ook hulle voorstelle en idees te stuur, sodat inligting oor die sindroom wyd in ons mooi land versprei kan word.

Hierdie is die eertse nuusbrief van 2011 en die doel van ons nuusbrief is dan ook om lesers bloot te stel aan ontwikkeling in navorsing sodat die leser self inligting kan navors, om dit dan op hulle eie omstandighede toe te pas. Ek verwys hier na die twee artikels oor die bynieroetereikendheid (waarvan kortisol die belangrikste is) wat by persone met PWS kan voorkom. Al die inligting kan vir ouers oorweldigend en verwarrend wees. Bring gerus die inligting onder die aandag van die huisarts en moenie skroom om hulp en leiding van 'n kundige te vra nie.

Carnitien, Co10Q en Omega vetsure sal vir party lesers bekend wees en weer vir ander vreemd. Meer hieroor in drie artikels wat deur dr. J Miller van die Universiteit van Florida, VSA verskaf is en soos met baie ander inligting is dit op Amerikaanse omstandighede gerig. Die voordele van hierdie middels is nog nie wetenskaplik bewys nie, maar daar is ouers wat positief daarvoor berig. Wanneer die gebruik van aanvullings oorweeg word, is dit baie belangrik dat dit eers met die pediater of

huisarts bespreek word. Elders in die nuusbriëf is die kontakbesonderhede van prof. Nola Dippenaar wat ook bereid is om op navrae te reageer.

Ontmoet Luke Legemaate wat op twee jaar en agt maande met PWS gediagnoseer is. Janet, Luke se ma en ook die ondervoorsitter van die Vereniging, skets die pad wat hulle tot nou toe met Luke geloop het. Ons kan leer uit die sukses van Janet se ingesteldheid om ander op te lei en streng voorskrifte te hê oor die hantering van Luke. Met die vriendelike vergunning van Natasha Martin, 'n dieetkundige, plaas ons 'n gebalanseerde eetplan wat vir Luke, nou vyf jaar uitgewerk is. Dit is 'n voorbeeld van 'n eetplan en gee waardevolle riglyne, maar neem asseblief kennis van die boodskap dat 'n kilojoule-beperkte eetplan altyd onder die leiding van 'n geregistreerde dieetkundige moet geskied.

Die nuusbriëf bevat nie net feitlike inligting nie, maar ook die ervaring en gevoelens van 'n ma toe haar baba met PWS gediagnoseer is. Lees gerus hoe die hartseer, swaar en ervaring van die onvolmaakte wêreld vir haar ander betekenis gekry het, hoe sy nou anders na die lewe kyk en haarself en ander beter verstaan.

Oor die jare heen is daar baie geskryf oor die gevare van versoeters. Lees gerus die artikel oor Aspartaam wat van die webwerf van NICUS gekry is. By persone met PWS loop ons die gevaar dat hulle uit hulle ingesteldheid van *meer is beter* maklik té veel versoeters (pilletjies) kan gebruik. Ouers moet wat dit betref ook maar in beheer bly. Is daar huishoudings waar die gebruik van versoeters by persone met PWS 'n twispunt is? Hoe hanteer julle dit?

Verder is daar brokkies nuus en inligting wat die aandag van die leser vra en teken asseblief nou al die datum van die 2011 Algemene Jaarvergadering aan.

Baie dankie aan Wilna Basson wat vir die **PRET**-bladsye, die saamstel en versending van die nuusbriëf verantwoordelik is. Dit is altyd 'n groot taak wat afgehandel moet word.

Die seisoen is besig om te draai en goeie wense aan almal totdat die volgende nuusbriëf in Juniemaand verskyn!

Herfsgroete

Rika du Plooy.

Skakel of epos gerus vir Janet Legemaate om gedagtes en vrae oor die hantering van 'n kleuter met PWS uit te ruil.

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From the chairperson...

Dear Readers

The year marches on! I trust that 2011 had a very good beginning for you all. Many countries experienced huge natural disasters. We especially think of the people in New Zealand and Japan which were battered by earthquakes. I spoke to Linda Thornton and luckily, according to her, no families in Christchurch with PWS dependents were caught up in the earthquakes. Everybody was nevertheless shocked and traumatised and it will take time for things to return to normal. Eiko Shoji, the vice-president of the PWSA (Japan) reports that they still can't make contact with one PWS family living near the heavily damaged area. All PWS families and all those affected can count on our sympathy.

The main aim of our Association for 2011 is to foster a greater awareness of Prader-Willi syndrome. This was one of the motivations submitted to the Willowton Group when we applied for the donation of R100 000. We have already had an opportunity at the beginning of the year to promote Prader-Willi syndrome. We arranged for a full colour page article PWS in the March edition of *EZEMPILO, Health Matters*. This monthly magazine is published by the Department of Health in KwaZulu-Natal and is distributed free of charge (100 000 copies). A first in this magazine is the fact that the information about PWS will also appear in a black language that is isiZulu. Information about PWS will also be supplied to professionals involved with children with disabilities, attending the SAALED Congress in Cape Town early in April. We also want to give copies of the *Medical Alert Booklet* of IPWSO to all final year medical students at the different universities. We are dwelling on other ideas as well, but we are also dependent on ideas from our readers. Do send us your suggestions and help to introduce PWS throughout the country.

This is the first newsletter for 2011 and one of the aims of the newsletter is also to introduce our readers to development and research in the field of PWS. We want each reader to do his or her own research and to apply the resultant knowledge to his or her own situation. In this regard I refer you to the two articles on central adrenal insufficiency (mainly cortisol) which may occur in persons with PWS. All the information supplied may be mind-boggling to parents. Guidance by an experienced person is definitely needed. Do not be afraid to ask for help and assistance.

Carnitine, CoQ10 and Omega supplements may be well-known to some readers but very much alien to others. Please read the articles on these substances written by Dr J. Miller of the University of Florida in the USA. Some of the information in these articles is only applicable to USA circumstances. The benefits in the use of these substances have not yet been scientifically proven; however, some parents with children with PWS have indicated that they have experienced positive results. Take note that the use of all supplements must be discussed with your doctor or paediatrician before you administer them. Elsewhere in the newsletter are the contact details of Prof Nola Dippenaar who is prepared to answer questions in this regard.

Our profile page in this newsletter is on Luke Legemaate who was diagnosed with PWS at the age of two years and eight months. Janet, Luke's mother, who is also the vice-chairperson of our association, gives an account of their experiences with Luke. Her mission is to train others about PWS. The family adheres to strict rules regarding the handling of Luke. Natasha Martin, a dietician, gave us permission to publish the balanced meal plan she has compiled for Luke (now five years old). This is a fine example of a meal plan and it also gives useful guidelines. A kilojoule restricted meal plan must also be discussed with a registered dietician before administering it.

Apart from mere facts, we also publish the experience and feelings of a mother when her child was diagnosed with PWS. Read about her sorrows and difficult times. But her experiences with her child gave her a new lease on life. She sees everything in a new light and understood herself and others better.

Much is written about the dangers of sweeteners. Read the article on Aspartame which was taken from the website by NICUS. Persons with PWS have the tendency to use more sweeteners (tablets or sticks) than necessary. Parents must be aware of it and take charge of this aspect to. Is the use of sweeteners a matter of dispute in your family? How do you handle it?

You will find other useful information in this issue. Please also take note of the date for the Annual General Meeting for 2011.

As always, our heartfelt thanks go to Wilna Basson who is responsible for the FUN pages and also for compiling and dispatching of the newsletter. It is indeed a formidable task.

Summertime is nearing its end. Best wishes to everybody. We'll meet again in our next newsletter which will appear in June!

Autumn Greetings

Rika du Plooy.

Phone or e-mail Janet Legemaate if you want to share ideas regarding the handling of a preschooler with PWS.

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Luke's Story

by Janet Legemaate

Brian and I are parents to two wonderful boys, Matthew (13) and Luke (5). Luke has PWS and was born almost 8 years after Matthew. Throughout my pregnancy we had prayed for a normal birth and normal child, no issues. We did not ask for an "Einstein" or "Sport's Star", just a normal baby. You may ask why the focus on this...well Matthew was born with a congenital heart defect and honestly we had been through the mill with him. At the time of Luke's birth, Matthew had already had five open heart surgeries, we have spent weeks outside of ICUs, and Matthew has consequential health and learning issues.



Luke and Rascal

Luke was born after an emergency Caesar, as fetal movement was severely diminished and my blood pressure was very elevated. We remember the doctors saying 'Wow, there's a lot of amniotic fluid' and then the pediatrician battled to get a response from Luke. With an initial Apgar of 3/10 and later of 6/10, concern was already raised as soon as Luke was born. He was taken to NICU and remained there for two weeks, coming home on 6 January.

By the time he came home, a long list of tests had already been done, all trying to establish why he was "floppy", non-responsive, failing to suck etc. A CT scan revealed cerebral atrophy throughout the brain and the doctor told us that at best Luke would have cerebral palsy but probably severely brain damaged. A friend had told me about a child in her class in Johannesburg with PWS, so we asked for the test to be done. The FISH test for PWS came back negative. In our ignorance we did not know there are three types of PWS and the FISH test only detects the deletion type. (The majority of PWS cases are the deletion type. The other test, which is a genetic test called Methylation, detects all three types, but of course is much more expensive.) So back to the drawing board it was.

We, my mom and I, in consultation with a speech therapist, occupational therapist, physiotherapist and Baby Gym specialist, began 8 therapy sessions a day. We had also contacted the Institute for Achievement of Human Potential in Philadelphia who advised us that the brain can be "recreated" with stimulation. So stimulate we did, physically, sensory and mentally....ALL THE TIME.

Although Luke's milestones were delayed he was walking totally on his own at 16 months and stopped falling regularly at 18 months. We realized he was picking up weight at about two years and of course, he would eat anything and loved his food. We thought, yes, he is getting better! How little we knew. He was not food seeking though.

We continued with our therapy sessions and continued doing research, eventually finding out that there was a second test for PWS, which we immediately had done. We received the result of this test when Luke was 2 years and 8 months old. We cried with frustration, but we knew Luke needed us, as the earlier you manage the circumstances and environment, the less you have to fix!

Luke had his adenoids and tonsils out before beginning growth hormone therapy at 2 years and 11 months. Looking back, we can see a big difference in his growth and development, especially if we look back to pictures and just look at his hands and feet which were really small and are now beautifully proportioned. So for Luke growth hormone therapy has certainly assisted his development in so many areas.

Luke was home schooled for two years after a disastrous attempt at play school. However, with our move to Durban in November 2010 we were able to find a lovely pre-school for him where they are prepared to make the small adjustments to cater for him and he is doing very well. He has settled into the routine and the staff are fabulous with him.

I spent a good couple of hours with the staff before Luke began school, effectively "teaching them" about PWS and this has certainly paid off. They are well prepared and as per my instructions, do not give an inch! They have made every effort to ensure Luke's safety and that he receives all the encouragement he needs. The school itself is very structured so this has helped considerably.

Luke is now 5 years old and has brought boundless amounts of joy to our lives and also added huge stresses. He has the greatest sense of humour, is not yet food seeking but has begun to tell us he is hungry more often, is very compliant and is a happy child. Even going shopping is of little issue as of yet as the rule is just "No treats!" and this is NEVER broken.

We are constantly questioning and researching how we can give our boys what they need in the future. Of huge concern is the lack of PWS focused residential care for our children in South Africa, because let's face it, we will not be around forever and we already know how difficult times can be when we do not have the support we sometimes need. Brian and I are looking at facilities in KZN (and if necessary we will develop one ourselves!) and are trusting that we will be able to do something before Luke requires it.

Dealing with two boys with medical, as well as educational, issues is difficult to say the least but we press forward. Matthew is an amazing older brother and has immense empathy and understanding, perhaps because of his own health issues.

One thing we are sure of is that we would not be where we are with our boys today without our faith and trust in God to see us through. Even despite this, at times it has been difficult and we have asked “why both boys?”. Ultimately, we know there is a plan in all of this. Sometimes we ask “Why doesn’t God heal our child of PWS?” Maybe we should be asking, “Does God have a destiny for a child with PWS?” If he didn’t, he wouldn’t be God!§

A Life Less Perfect

by Lisa Peters

Before my son Nicholas was born, my life was perfect.

I ran in an invisible race with neighbours and friends. A race to see who had the greenest lawn, the smartest kids, the whitest teeth. I was a member of an elite group, devoted to raising elite children. We spent our time at bbq's and soccer games tallying our points in our quest to grab that glittering gold ring of perfection.



5 years

As we admired our children, and our lawns, we never stopped to realize that on our faces we wore rose-colored glasses and in our hearts we felt an emptiness that searched for a deeper meaning to our lives.

On January 18, 2002, like a thin layer of glass, my perfect life came shattering down by the purest sound of six horrifying words...“Your son has Prader-Willi syndrome.” And suddenly I could not breathe.

I sobbed for my poor, weak, little child.

I sobbed for myself.

I sobbed for the perfect life we would never have together.

There were no flowers, no cards, no congratulatory notes from family and friends...my son entered the world in silence. No smiles, no laughter, no fanfare. No one welcomed him. Everyone was sad.

Where in a perfect life would this little child fit? It was as if his very existence threatened to tarnish this utopian world we had created. My tiny son was a giant monster of truth that threatened to expose the meaninglessness of a life built out of playing cards. And all who lived in these fragile card houses could not understand how to celebrate the birth of this little child.

My son lay limp upon his bed. A yellow feeding tube was taped to his cheek and travelled up his nose and into his stomach. Taped to his tiny skull another tube pumped antibiotics into his fragile veins. Around his floppy body a brace made of thick straps and stiff velcro held his weakened hips in place. Feeding machines and IV poles surrounded him like quiet metal soldiers standing at attention. Everywhere

alarms sounded...a constant reminder that this was hell and we now lived in it. Around me in the NICU, I saw only despair... parents with children struggling to live.

Like my newly born infant, I was abruptly and cruelly removed from the warmth of my womb-like perfect life. I was thrust head-first into a cold and terrifying new imperfect world.

This was my new home. I felt sick.

Every movement I made felt unnatural and awkward. My mind was frozen. My body moved like a robot. I did not want to look around me, for everywhere I looked, I saw pain. I felt like a soldier on the battlefield, frozen by the ghastly sight of the slain bloody carcasses at his feet. And yet, like this soldier in a war he did not create, I too could not escape my fate.

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The rose colored glasses I once blindly wore were smashed into smithereens. My eyes, unaccustomed to this new light, could not stop crying.

In his sad and traumatic entrance into this world, my imperfect son had given me a sad, beautiful and precious gift, the gift of sight. The ability to see the world not as I wanted it, but as it truly was. I saw the pain and sadness...the frailty of life.

When he finally came home from the hospital, I had to hold him carefully or his head would flop quickly forward. And when it did, he couldn't breathe. I felt helpless.

I questioned God, what had I done to my son? I felt guilty.

When I took him to restaurants or the mall, people would ask questions, "Why does your son have a tube in his nose?" I felt embarrassed.

When I handed my fragile son to the doctor at Children's Hospital for eye surgery, I felt scared.

So many endless days brought so many endless hurtful, hard emotions. I felt so very tired.

And when my tired body seemed like it could bear no more...my floppy little child began to get stronger. And as he did, I began to feel a lost emotion...happiness.

After almost a year, Nicholas finally held up his head. That tiny little infant who struggled to breathe was now able to see the world. I felt joy.

When his g-tube was removed, and the words "failure to thrive" were removed from his chart, there were tears. No more questions to answer. I felt relieved.

When he pushed away his metal walker and took steps for the first time, I wept.

Slowly, I began to realize that these tortuous feelings, these hardships were somehow very important for me to experience. For it was these extremes, these awful, uncontrollable feelings and hardships, that gave my life new meaning. I understood myself a little better. I understood others a lot better. And although these emotions left me feeling fragile and vulnerable, I couldn't help but wonder if this is God's intention?

As I became accustomed to my new sight, I saw we were surrounded here on earth by many hidden angels. Intelligent, kind human beings who were devoting their lives to curing and healing the sick. Why hadn't I seen them before? Why hadn't I appreciated them? And once again this imperfect child had enabled me to see. I could see the special souls that travelled among us here on earth. Selfless, gifted souls devoted simply to the healing of others. These guardians would now be a part of our lives forever. This horrifying new life of ours now seemed a little less scary.

There were other angels here on earth now visible to me for the first time...special parents of children with Prader-Willi syndrome whose paths were as treacherous as ours. And with these special few we shared our advice, our tears, our love...and a unique brotherhood was forged...for we could truly understand each other! I listened to the words of these parents I had never met. I listened to these strangers with quiet hearts and somehow my hardened spirit was now a little lighter. Why? I do not know for I was still sad. But somehow by releasing tears, and listening to others, I began to accept and understand this challenging life.

I began to accept that my son is not like others in this world. I began to accept that this was not a curse but a blessing. To me he was unusually happy, loving and kind. I was amazed by his keen perception of human beings and his unique ability to make even the grumpiest of grouches smile brightly. He lives to dance and laugh and love. He has a warm heart and a gentle spirit. And although he is my child, he has also been my teacher.

Each of us are blessed with special gifts and although his gifts are hidden, buried beneath a weakened body, his gifts are no less special. I do not have a son who can run very fast, I have a son with the precious gifts of empathy and human compassion. I now realize that my life with Nicholas will not be like the lives of so many others...ordinary. It is an extraordinary life. A life filled with high highs and low lows. I would not trade one day of feeling that terrible pain because I know now the terrible happiness that is on the other side waiting for me. What I have learned is to appreciate both. For it is these feelings, this blending of the good and the bad, that somehow seem to bring me closer to understanding my purpose here on earth.

This awareness, this blending of heart and spirit, has helped me to embrace my son and enjoy this journey we are sharing together. It is a sad, sweet, beautiful trip. It is a life less perfect. It is a life more meaningful.§

EXAMPLE of MEAL PLAN

A Meal Plan for a five year old boy, diagnosed with PWS.
His weight is 23kg and he receives daily growth hormone therapy

Recommended daily intake: **900 calories or 3 767 kJ**

Permission was granted by Natasha Martins, Paediatric Dietician, for the publishing of this information.

Dietary Prescription: 5 year old boy with PWS, weight 23kg.

Energy: max 900 calories per day or 3 767 kJ per day.

Daily Exchange List: Starch: 4 servings per day. Vegetables: 3 servings per day. Fruit: 2 servings per day. Meat (Lean): 3 servings per day. Dairy: 2 servings per day. Fat: I haven't included any fat exchanges as the above exchanges provide adequate calories.

Please feel free to contact Natasha Martins for any uncertainties about the meal plan and food exchanges. Tel: 031 764 4277 or Email: paediatric.dietician@gmail.com

Please note that a calorie restricted diet must always be followed under the direct supervision of a registered dietician. A child's growth, weight and height, and nutritional requirements need to be assessed before implementation of a meal plan.

SUMMARY OF FOOD EXCHANGES

STARCH	
Pronutro	25g
Oats	½ cup = 100g
Pasta	½ cup = 100g
Bread	1 slice
Proviso's	3
Beans	½ cup = 100g
Rice	1/3 cup = 50g
Popcorn	1cup
Pita (15cm)	½ pita = 90g
Rye vita	2
Melba toast	4 slices
Cream crackers	3
Rice cake (10cm)	2
Snack bread	3
STARCHY VEGETABLES	
Corn / peas	½ cup
Corn on the cob (6 inches)	1
Baked beans	½ cup = 100g
Sweet potato	½ cup = 100g
Potato	1/2 cup = 100g = 1 small
FAT FREE DAIRY	
Fat free milk	1 cup
In-Shape fat free yoghurt	100g
LEAN MEAT	
Lean beef	30g

Lean chicken	30g
Lean pork	30g
Fish: All fresh, frozen fish	30g
Tuna (canned in water)	¼ cup
Fat free cottage cheese	¼ cup
Egg whites	3 whites
FAT	
Olive oil	1 tsp
Sunflower oil	1 tsp
Margarine	1 tsp
FRUIT	
Apple, unpeeled, small	1 = 120g
Banana, small	1 = 120g
Blueberries	¾ cup
Cherries, fresh	12 = 90g
Grapes, small	17 = 90g
Mango, small	½ fruit (160g) or ½ cup
Nectarine, small	1 = 150g
Orange, small	1 = 180g
Peach, medium	1 = 180g
Pear. Large	½ = 120g
Raisins	2 Tbsp
Strawberries	1 ¼ cup
Apple / grape / orange / pineapple juice	½ cup
VEGETABLES	
Cooked vegetables	½ cup
Raw vegetables	1 cup
FREE FOODS (< 20calories per serving)	
Carbonated water	
Coffee or tea (no sugar or milk)	
Drink mixes, sugar free	
Cabbage	1 cup
Celery	
Cucumber	
Mushrooms	
Spinach	
Lettuce	
Seasonings	

Please note: 1 Calorie = 4,186 kilojoules (kJ)

cal X 4.186 = kJ

e.g. 900 x 4.186 = 3 767kJ

§

EXAMPLE OF MEAL PLAN

7 Day meal plan for a Prader-Willi Syndrome 5 year toddler

Courtesy of Natasha Martins, Paediatric Dietician

	DAY 1	DAY 2	DAY 3	DAY 4	DAY 5	DAY 6	DAY 7
BREAKFAST 1 STARCH ½ DAIRY 1 FRUIT	25g Pronutro ½ cup skim milk 1 small banana	1 Slice bread 100g InShape fat free yoghurt 1 small apple	½ cup oats ½ cup skim milk 2 Tbsp raisins	25g Pronutro ½ cup skim milk 1 small banana	1 Slice bread 100g InShape fat free yoghurt 1 small apple	½ cup oats ½ cup skim milk 2 Tbsp raisins	25g Pronutro ½ cup skim milk 1 small banana
SNACK ½ STARCH 1 FRUIT	½ cup popcorn 1 small apple	20g pretzels ½ cup fruit salad	1 rice cake 1 small banana	1.5 Provita's 1 small apple	1.5 cream crackers ½ cup mango	2 Melba toast 1 small orange	1.5 cracker bread 2 small plums
LUNCH 1 STARCH 1 MEAT 1 VEG ½ DAIRY	½ cup pasta 30g lean meat ½ cup cooked vegetables 100g InShape fat free yoghurt	100g potato 2 Tbsp parmesan ½ cup cooked vegetables ½ cup skim milk	1 slice white bread 30g lean meat 1 cup raw vegetables 100g InShape fat free yoghurt	½ cup baked beans 30g lean meat ½ cup cooked vegetables ½ cup skim milk	90g Pita 30g lean meat ½ cup cooked vegetables 100g InShape fat free yoghurt	1 slice white bread 2 Tbsp parmesan 1 cup raw vegetables ½ cup skim milk	½ cup pasta 30g lean meat ½ cup cooked vegetables 100g InShape fat free yoghurt
SNACK ½ STARCH 1 MEAT	1 rice cake ¼ cup fat free cottage cheese FREE: Cucumber ¼ cup	1.5 Provita's 30g lean meat FREE: Lettuce ¼ cup	2 Melba toast ¼ cup fat free cottage cheese FREE: Celery ¼ cup	1 rice cake 30g lean meat FREE: Cucumber ¼ cup	1.5 Provita's ¼ cup fat free cottage cheese	2 Melba toast 30g lean meat FREE: Celery ¼ cup	1.5 Cream crackers 40g ¼ cup fat free cottage cheese 55g FREE: Cucumber ¼cup
SUPPER 1 STARCH 1 MEAT 2 VEG	1/3 cup rice 30g lean beef 1 cup cooked vegetables	½ cup pasta 30g lean meat 1 cup cooked vegetables	100g sweet potato 30g lean meat 1 cup cooked vegetables	1/3 cup rice 30g lean beef 1 cup cooked vegetables	100g potato 2 Tbsp parmesan 1 cup cooked vegetables	½ cup pasta 30g lean chicken 1 cup cooked vegetables	½ cup baked bean 30g lean meat 1 cup cooked vegetables
SNACK = 1 DAIRY 100g InShape fat free yoghurt or 1 cup fat free milk							

Adrenal Insufficiency during Acute Illness in People with PWS: New Research suggest treatment to Minimise the Risks of Sudden Death.

*Dr Nicholas Finer, Clinical Director, Wellcome Clinical Research Facility,
Addenbrook's Hospital, Cambridge
Courtesy of Wavelength, February, 2009, p 17*

When people with PWS become ill, they may show few signs of illness and in particular often do not develop a fever, don't vomit and may not report pain or discomfort, and this may explain why they are at risk of sudden death. Important findings from Dutch researchers* have shed light on a possible explanation for this, and suggest treatment that could reduce these risks. Normally during illness the adrenal glands (small glands that sit on top of the kidneys) increase the amount of steroid hormones they secrete into the blood stream. These hormones (the main one is cortisol) help the body combat infection and stress. The adrenal glands are controlled by the pituitary gland that lies at the bottom of the skull, and which produces a hormone called ACTH that signals the adrenal glands to produce cortisol; the pituitary gland itself is regulated by a part of the brain called the hypothalamus. It is known that a shortage of cortisol is very dangerous and so people who do not have functioning adrenal glands (a condition known as Addison's disease) or who take large doses of steroid medications for asthma or other conditions that results in adrenal suppression must take extra cortisol if they fall ill. The same is true for people who are on replacement cortisol for pituitary disease.

The Dutch investigators studied 25 children with PWS and looked at the ability of their hypothalamus to stimulate the pituitary to stimulate the adrenals to produce cortisol. The children had normal cortisol levels during the day, but nearly two thirds of them failed to increase their cortisol levels in response to a drug called metyrapone. Metyrapone blocks cortisol synthesis, and so should cause an acute increased demand for ACTH production, a situation mimicking stress. Patients with an insufficient ACTH response during the metyrapone test are therefore considered as having adrenal insufficiency during stressful conditions. A particularly vulnerable time for children with PWS could be during the early morning when cortisol levels are normally at their lowest and any failure to increase levels could be critical.

The authors of this research suggest that in illnesses that cause 'moderate stress' (e.g. influenza, upper respiratory tract infection, ear infection) children (and probably adults too) with PWS should take extra cortisol (given in the form of hydrocortisone tablets 50mg four times daily) until they are over the acute illness. For more serious illnesses hydrocortisone should probably be given by injection and in higher doses; this would mean hospital admission in practice. This advice accords with the 'Sick Day Rules' given to patients known to have adrenal insufficiency for other reasons and would seem a sensible and safe precaution, although the benefits of this are not proven and are based on these early findings from this study. Parents or people with PWS may wish to carry this information with them as it is unlikely to be known by non-specialist doctors.§

**Roderick F. A. de Lind van Wijngaarden and colleagues. J Clin Endocrinol Metab, May 2008, 93(5):1649–1654*

Central Adrenal Insufficiency in Individuals with Prader-Willi Syndrome

Jennifer L Miller, MD Pediatric Endocrinology University of Florida, USA

Courtesy of the website of PWSA (USA) www.pwsausa.org

A recent article in the Journal of Clinical Endocrinology and Metabolism by de Lind van Wijngaarden et al indicated that there may be a high frequency of central adrenal insufficiency (CAI) in individuals with Prader-Willi syndrome. Morning salivary cortisol levels and cortisol profiles were normal in all the children studied, leading the authors of the study to conclude that CAI in individuals with PWS only becomes apparent during stress. Therefore, the presence or absence of CAI cannot be determined by measuring an 8 AM cortisol level – the individual must be tested while stressed (e.g. with febrile illness) or using a stimulation test.

Given this information, we recommend that all individuals with PWS should be screened for the presence of CAI. The two ways to test for CAI are (1) to measure a cortisol and ACTH level while the child is sick or (2) to perform a stimulation test which will evaluate the hypothalamic-pituitary-adrenal axis. Because some children with PWS do not have fevers when ill, it can be difficult for parents and physicians to know when the child is sick enough to put the body under significant stress to accurately assess the presence of CAI. Thus, a stimulation test may be the best way to detect adrenal insufficiency. The metyrapone stimulation test was used in the above-mentioned research study, but this test is not usually done in the United States. A low-dose ACTH stimulation test (1 mcg ACTH) has ~ 95% sensitivity for diagnosing impaired adrenal function, but may miss mild CAI; a glucagon stimulation test has equal sensitivity for diagnosing CAI but may pick up more subtle abnormalities of the hypothalamic-pituitary-adrenal axis, and an insulin-tolerance test is the gold standard for evaluating for the presence of CAI.

Please present this information to your endocrinologist and discuss testing for CAI with him/her. It is important to discuss the fact that the presence or absence of CAI in an individual with PWS cannot be determined by measuring an 8 AM cortisol level – the individual must be tested under a stressful condition (e.g. illness) or using a stimulation test.§

Please be aware:

There are reports and discussion in the medical literature about adrenal hypofunction or insufficiency in PWS. Single measures of cortisol levels will not be helpful and adrenal challenge tests may be warranted. Please consult an endocrinologist for his/her input and advice before starting growth hormone treatment. For more information on adrenal insufficiency, please see two articles on this topic in the newsletter. It is also important to bring these findings under the attention of your child's paediatrician or medical practitioner.

CARNITINE AND PRADER-WILLI SYNDROME (PWS)

Courtesy of Jennifer L Miller, MD Paediatric Endocrinology, University of Florida, USA

Carnitine is a natural antioxidant that improves cellular energy metabolism. Low carnitine levels may occur due to metabolic diseases or can be due to inadequate nutritional intake of carnitine-containing foods [Stephnes et al, 2007]. Carnitine supplementation has not been studied in individuals with PWS, but in other conditions has been shown to improve hypotonia, ataxia, activity levels, and alertness. Infants and young children with Down syndrome, who have muscle hypotonia and delayed growth similar to that seen in individuals with PWS, have significantly lower carnitine levels than unaffected children of the same age, and supplementation with L-carnitine results in significant increases in visual memory and attention in this population [Seven et al, 2001]. A subset of individuals with PWS has been found to have low serum carnitine levels [Miller et al, 2010].

Carnitine is available in a variety of formulations, including L-carnitine (available over the counter or as a prescription) and carnitine fumarate. Although no formal studies of carnitine supplementation have been done, we have anecdotally heard that some individuals have improvements in gross or fine motor skills and alertness with carnitine supplementation. We recommend that you ask your physician to check a serum carnitine profile to document whether or not carnitine deficiency is present. If the carnitine levels are low on this profile, we recommend that the child have a urinary acylcarnitine profile measured. If the serum carnitine levels are low, we recommend a trial of supplementation with carnitine. Even if the serum carnitine levels are within the normal range, some individuals still benefit from carnitine supplementation, and so we recommend a trial of this medication for individuals with PWS.

Because prescription-grade products are FDA regulated, whereas over the counter supplements are not, we prefer that children with PWS try the prescription L-carnitine.

The dose is typically 50 mg/kg/day divided twice a day. However, an evening dose of carnitine keeps some children awake at night, so if this occurs, then once a day dosing is preferable.

Start carnitine once other medication doses are stabilized, so that you are not changing several medications or doses at the same time – this will allow you to

Please note:

*The articles, **Carnitine in PWS, CoQ10 and PWS and Fish Oil Supplement and PWS** are from Dr Jennifer Miller, a paediatric endocrinologist at Florida University, USA. The articles are directed at the USA public. If you are interested in trying these supplements with your children or family members please take the information to your health care professional to discuss it. Some of the products, as discussed in the articles, are not available in South Africa. However, if you contact Professor Nola Dippenaar on www.healthinsight.co.za or Mobile (+27) 82 9000 970 or e-mail nola@healthinsight.co.za, she will be able to point you in the right direction.*

determine if there are benefits from the supplement. We recommend a 1 month trial of L-carnitine to determine if there are benefits to the supplement for your child or not. If you discontinue the carnitine and see negative changes in your child, then restart it. We recommend trying carnitine if the child is taking less than 20 oz of formula or breast milk per day as an infant or once the child has transitioned from formula to cow's milk, soy milk, or almond milk. Formula is fortified with carnitine, so provided your child is taking an adequate amount of formula, the carnitine levels should be adequate. A list of carnitine containing foods is available from any dietician.

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COENZYME Q10 (CoQ10) & PRADER-WILLI SYNDROME (PWS)

Courtesy of Jennifer L Miller, MD Paediatric Endocrinology, University of Florida, USA

Coenzyme Q10 (or "CoQ10") is a naturally occurring vitamin-like substance in the body. CoQ10 is essential in energy production in all living cells, especially in the muscle. If deficient in CoQ10, an individual may feel less energetic, have reduced muscle function and have a decreased metabolic rate. CoQ10 also acts as an antioxidant in the blood and all cell membranes.

Individuals with Prader-Willi syndrome may have decreased levels of CoQ10. A blood test can determine if an individual's CoQ10 level in the blood is lower than normal. A muscle biopsy would be the best way to determine cellular CoQ10 level, but at this point we do not feel muscle biopsies are warranted.

When CoQ10 levels are low, supplementing with CoQ10 may help increase energy level, muscle function and metabolism. Some parents also see an increase in activity and attentiveness after supplementing their PWS child with CoQ10. However, while some parents feel that their child demonstrates improvements with CoQ10, others feel that it has no effect. Also, parents should be clear that CoQ10 supplementation is not a substitute for growth hormone treatment which has definitely been shown to have multiple benefits for individuals with PWS by many well designed research studies.

The information we have on the effectiveness of CoQ10 is all "anecdotal" data (i.e., parents commenting on their personal experience with their child). There have not been any controlled scientific research studies about the effects of CoQ10 in individuals with PWS. At this time, there are no KNOWN adverse side effects of taking CoQ10 if taken in an appropriate dose.

The recommended starting supplemental dose of CoQ10 varies according to different groups. It is anywhere from 1 - 30 mg per kg per day for infants, and no more than **180 – 200 mg** per day for older children. We have typically found that 60 – 100 mg per day in the older children and adults is sufficient to raise the blood level to the normal range in those individuals who were initially found to have low levels. If families are contemplating supplementation with CoQ10 we would recommend testing a blood level before and after starting CoQ10. Several specialty labs offer testing. One such lab is Horizon Molecular Medicine in Atlanta, Georgia. They can be

reached at 678-225-0222 or info@horizonmedicine.com. Other labs include Metametrix in Norcross, Georgia and Quest Diagnostics in California.

Dissolved CoQ10 in soft gel capsules (which contain vitamin E and other lipids to help the body absorb the CoQ10) are better absorbed than dry powder CoQ10 tablets or capsules. The CoQ10 soft gels typically come in 60 or 100 mg doses. Therefore, 1-3 capsules per day will need to be taken by older children and adults.

CoQ10 can be purchased over-the-counter at most pharmacies. There is also a liquid preparation (“LiQ-10”) available through International Nutrition for children who cannot swallow pills.

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FISH OIL SUPPLEMENT AND PRADER-WILLI SYNDROME (PWS)

Courtesy of Jennifer L Miller, MD Paediatric Endocrinology, University of Florida, USA

WHAT IS FISH OIL?

Fish oil, also known as omega 3 fatty acids, is recommended for a healthy diet because it contains eicosapentaenoic acid (EPA) and docosahexaenoic acid (DHA). These are precursors that reduce inflammation throughout the body. EPA and DHA also work together for development and function of the brain and eyes. The synapses in the brain are rich in DHA suggesting that it is involved in signal transmission along neurons, it makes up some gray matter in the brain, and is also found in the retina. Additionally, there is a compound in DHA, called “Resolvin” that helps reduce inflammation.

FOODS THAT CONTAIN OMEGA 3 FATTY ACIDS:

Omega 3 fatty acids are primarily found in organ meats and cold water or fatty fish (ie albacore tuna, cod liver, halibut, salmon, sardines – contain about ~1 gram per 3.5 oz of fish). Since Americans eat less fish in their diet, some could benefit from supplementation in their diet.

PRADER-WILLI SYNDROME AND FISH OIL:

Individuals with PWS may have very caloric and fat restricted diets. Because of this, the diet may be low in healthy omega 3 fatty acids. This is highly important for children due to the developing brain. Recently, infant and toddler formulas have been supplemented with similar amounts of fatty acids found in breast milk. Therefore, starting fish oil supplementation generally begins after 1 year of age when a child’s diet is primarily whole milk and solid foods or if the diet is low in fat. As adults, supplementation may be beneficial for:

- Inflammation: Can reduce joint tenderness
- Hyperlipidemia: Lowers blood TG levels
- Aging: Evidence is inadequate to conclude that it protects cognitive function with aging and dementia.
- ADHD: One review stated that it “appears to alleviate ADHD-related symptoms in at least some children, and one study of OCD children also

found benefits for academic achievement. Larger trials are now needed to confirm these findings.”

We typically recommend starting the fish oil when the child is over 1 year of age and on milk rather than formula or breast milk or if they are 6-12 months of age and taking less than 20 oz per day of formula or breast milk, because both formula and breast milk contain the essential fatty acids found in fish oil supplements. Coromega makes a fish oil for kids - available in Lemon Lime and Orange. I have heard some of our parents say the kids like it. I have attached a link to the Coromega website. Calories are 20 kcals/packet, 850mg fish oil. The company is pretty reputable. The dose is 1 packet per day. The website for Coromega is: www.coromega.com/coromega/html/product_childsqueeze.html. Coromega is made with an egg-base so if your child has any food allergies, this is not recommended. However, many families like the Nordic Naturals brand of fish oil and they have an infant formulation (Coromega says that it is only for ages 4 and up – we give it earlier than that, but some of the parents are uncomfortable giving something that is only approved for over 4 years of age when their child is an infant). The website for Nordic Naturals is: www.nordicnaturals.com/. Coromega has a product that is a mixture of fish oil and CoQ10 which a lot of parents like because that way they only have to give the one packet and not both fish oil and CoQ10 (the less meds they have to give, the better). It is called “Healthy Heart” and is the same price as the regular coromega. Also, both Nordic Naturals and Coromega will send parents samples to try before they order a big supply so they can make sure the kids will tolerate the product – parents just have to call the company and ask.

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ASPARTAME: SWEET OR BITTER?

Aspartame is one of the most widely used artificial sweeteners and can be found in products like Canderel, Equal, Nutrasweet, and Pick and Pay Sweetener as well as in diet cooldrinks and other “diet products”. The main purpose of artificial sweeteners, like aspartame, is to provide sweetness to foods and beverages without adding to their energy content or increasing blood glucose concentration. These products are widely used in the diabetic and slimming markets. They are incorporated in the diets of many people, but they do not contribute meaningfully to their diets, since they are consumed in small amounts.

Aspartame is 200 times as sweet as sugar (sucrose), enhances and intensifies flavours, particularly citrus and other fruits, and has no after taste. Aspartame provides 4kCal/g in energy; however, because of the intense sweetness of aspartame, the amount of energy derived from it is negligible.

Composition of aspartame: Aspartame is made up of components found naturally in common foods. Upon digestion, it is completely metabolized to two amino acids (building blocks of protein) and methanol (approximately 50% phenylalanine, 40% aspartic acid and 10% methanol). These amino acids are derived from dietary sources (such as meats, milk, fruit and vegetables) and are consumed in much larger quantities as part of a normal balanced diet.

Phenylalanine is one of the ten amino acids that are considered to be essential in the human diet, since it cannot be synthesized in the body, and therefore must be present in sufficient amounts in the daily diet to prevent deficiencies. The estimated requirement for phenylalanine is 14mg/kg/day for adults. The average adult diet can provide anything from 80g – 140g of protein (plant and animal sources) per day. This results in an approximate phenylalanine intake of 3 – 5g per day. A person weighing 60kg and eating a typical well-balanced, moderate to high protein diet is therefore consuming between 50mg and 84mg of phenylalanine per kg of body weight daily.

Aspartic acid is considered as a non-essential amino acid, because it can be synthesized in the body from other amino acids. Essential and nonessential amino acids are utilised by the body to build proteins, promote growth and maintain bodily functions. When aspartame is digested, the body handles these amino acids in the same way as those in foods that we eat every day.

Is consumption of aspartame associated with ill health or “health problems”?

It is imperative that we, individually and collectively, guard against any adverse effects that may arise from the consumption of substances, or indeed from the excessive use of nutrients themselves, that find their way into our daily diet by accident or by design. It is also very unfortunate that from time to time unsubstantiated reports based on no or anecdotal evidence is published in the media creating fear and confusion among consumers. In the case of aspartame, such anecdotal and ill-founded reports include:

- **Epileptic seizures:** Extensive studies have failed to demonstrate an association between epileptic seizures in children and adults and the ingestion of aspartame even in large dosages of 50mg/kg.
- **Multiple Sclerosis:** The senior medical adviser of the Multiple Sclerosis Foundation in the USA, having reviewed the available scientific evidence, has categorically denied any association between the consumption of aspartame and the disease. Also, the disease has existed long before aspartame came onto the market.
- **Hyperactivity and attention deficit disorder in children:** Several studies in children have shown no relationship between aggressive, hyperactive or cognitive function problems and aspartame.
- **Depression, headache or behavioral functioning in adults:** Large daily doses (600mg/day) of aspartame had no effect on the brain’s psychological- and physiological functions, or behavioral functioning in healthy adults. However, one study (the results of which have not been confirmed) in a small number of subjects reported a worsening in depression when depressed patients did consume aspartame.

- **Hypersensitive- and allergic reactions:** There is no scientific basis for an association between aspartame and allergic reactions or hypersensitivity reactions. Subjects who believed themselves to be allergic to aspartame did not have reproducible reactions to it. It should however be borne in mind that individuals who have a history of allergy should be particularly careful *at all times*, when they introduce any new foods or substances in their diet.
- **Brain tumors:** There is no known association between an increased incidence of brain tumors and the intake of aspartame. The Food and Drug Administration (FDA) of the USA “stands behind its original approval decision (*on the safety of aspartame*), but the Agency remains ready to act, if credible scientific evidence (*to the contrary*) is presented to it – as would be the case for any product approved by the FDA” (*FDA official statement on aspartame*). In October 2000, the French Consumer Affairs Ministry asked the French Food Safety Agency (AFSSA) to form an expert committee to study any possible link between exposure to aspartame and the occurrence of brain tumors. On the basis of a comprehensive analysis of the current scientific data, the expert committee concluded that there is no relationship between aspartame consumption and brain tumors in humans or animals.

On balance, therefore, there is an almost total lack of credible scientific evidence that proves that aspartame is not safe for human consumption. Extensive product research has been done and the controlling bodies all over the world have approved the product. Until such time that there is evidence of health risks associated with the consumption of aspartame, it can be considered as safe.

According to the World Health Organization, the estimated amount of aspartame that can be ingested daily over a lifetime without appreciable risk for a healthy adult is 0-40 mg/kg/day (it is considered that a 60kg adult consuming 2400 mg aspartame per day for life, which is equal to 12 cans of diet soft drink, will have no adverse effects). Daily intake of aspartame in the United States is approximately 3mg/kg/day, Canada 6 mg/kg/day, UK 2 mg/kg/day and Germany 3 mg/kg/day (data not available in South Africa.) For a person weighing 60kg this translates to an intake of approximately 180-360 mg per day.

Health myths about aspartame uncovered:

- **Does aspartame contain methanol?** Aspartame itself does not contain methanol. A very small amount of methanol (10% by weight) is formed when aspartame is digested. The amount of methanol therefore produced from aspartame is very small and, in general, it is less than the amounts found in many fruits and vegetables. The body converts the methanol to formaldehyde, which is instantly converted to formic acid. Formic acid is then quickly eliminated by the body in the form of carbon dioxide and water.

- **Is aspartame safe during pregnancy?** There have been no reports of adverse fetal or maternal effects from the use of aspartame, either in laboratory animals or humans. Nevertheless and as a general rule, it is always safer to avoid an excessive intake of any nutrient or substance during pregnancy.

- **Is aspartame safe in phenylketonuria?** People with phenylketonuria (PKU) should not consume aspartame (or any other sources of phenylalanine) because of its

phenylalanine content. PKU is a rare hereditary metabolic disorder. Such individuals have elevated blood levels of phenylalanine, since their body has insufficient enzymes to metabolise phenylalanine. In these individuals, therefore phenylalanine intake is always restricted as part of their treatment.

- **Is aspartame safe in diabetes?** Sweeteners can be considered as a convenient alternative to sugar in the diabetic diet, since it allows them to enjoy sweetened foods without affecting their control of diabetes. The position of the American Diabetes Association is that aspartame is safe and can be included in a diabetic meal plan. There are no reports to indicate that the moderate use of aspartame by these individuals is not safe. Care should be taken regarding commercially available “diabetic” products where sugar is substituted for artificial sweeteners, as the fat content might still be high so to provide a more palatable product.

- **Will aspartame increase one’s appetite or induce carbohydrate cravings?** Available data indicate that participation in a weight management program, based on a well balanced diet, which included aspartame, resulted in long-term maintenance of reduced body weight. Aspartame was not associated with an increase in hunger at any time.

GUIDELINES FOR USING ASPARTAME:

- As part of a healthy balanced diet, moderate amounts of aspartame can provide a little sweetness without added energy.
- When aspartame is heated for long periods, loss of sweetness may occur. Rather add the sweetener at the end of the cooking process when the food is removed from the heat.
- Always use a variety of artificial sweeteners and do not use a particular one for long periods. Read the labels of products to identify the sweetener that has been used.
- People diagnosed with the metabolic disorder, phenylketonuria, must restrict their intake of phenylalanine from all dietary sources as their treatment requires them to do.
- As a general rule and as a precautionary measure, individuals with a history of allergy should be careful when introducing any new foods or substances in their diet.
- Individuals who suffer from depression should consult their doctors before they introduce aspartame or alter its current intake in their diets. §

The information explosion in the science of nutrition very often creates the impression that available information is contradictory. Consequently, it is no longer easy to distinguish between fact, misinformation and fiction. The Nutrition Information Centre of the University of Stellenbosch (NICUS) was established to act as a reliable and independent source of nutrition information. For further, personal and more detailed information, please contact NICUS or a dietician registered with the Health Professions Council of South Africa. References from the scientific literature used to compile this document are available on request.

NICUS

(Nutrition Information Centre of the University of Stellenbosch)

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PROBLEM CORNER

Q

I have just discovered that there is a procedure for obese people which will bring about weight loss called “gastric banding”. Do you or the PWSA (SA) have any information on whether this procedure is recommended for people with PWS?

A

Gastric banding is one of the surgical weight loss procedures (bariatric procedures) used in obese individuals. Due to the complexity of the dietary pattern in persons with Prader-Willi syndrome and the disappointing results with energy restriction and strict supervision to access to food parents look for other options. Small case series have reported success in the short term but an analytic review done in the USA didn't show enough long term benefit to subject individuals to the potential risk of surgical intervention. An alternative approach would be supervised low- energy diets with vitamin /mineral supplementation, restricted access to food and a daily exercise regime.¹

1. *Scheimann AO, Butler MG, Gourash L, et al. 2008 Critical analysis of bariatric procedures in Prader-Willi syndrome. Journal of Pediatric Gastroenterology and Nutrition 46:80-83*

We thank Dr Engela Honey for answering the question on gastric banding.
Dr Honey is a paediatrician at the Department of Human Genetics, University of Pretoria and medical advisor of PWSA(SA)



AGM & SOCIAL 2011

The next AGM will be held on
Sunday, 21 August 2011.
More detailed information to follow.

Die volgende AJV word op
Sondag 21 Augustus 2011 gehou.
Meer inligting sal later gestuur word.

Ipwso News

Message from IPWSO in connection with the earthquake and tsunami in Japan.

The Vice- President of IPWSO, Janalee Heinemann received the following message from our friends in Japan:

Dear Janalee,

Thank you very much for your concern. I still cannot make contact with one PWS family living near the heavily damaged area; I'm so worried about them. I hear there are still many small aftershocks even in Tokyo. Japanese people now fear for the leakage of radioactive substances from a nuclear power plant.

May I ask you a favour? I know our friends around the world are worried about the Japanese members, but so far I've managed to write only to Linda, Susie and Shuan-Pei. So, could you tell the IPWSO members all over the world (not literally all, but at least some important Board members) that I'm all right and one family is still missing, and that their prayers are needed, if it's not a burden to you?

I think Japan will suffer from serious economic and emotional depression at least for the next few months, but Japanese people really appreciate and are encouraged by lots of every kind of support from abroad.

Thank you very much for sending your thoughts and prayers for us.

Kindest regards

Eiko Shoji

Vice-president, PWSA Japan

IPWSO 8th INTERNATIONAL CONFERENCE 2013

The PWSA (UK), in partnership with the University of Cambridge Intellectual and Developmental Disabilities Research Group, (headed by the President of PWSA (UK), Prof Tony Holland) warmly invite you to the 8th International PWS Conference.

The conference will be held at the Fitzwilliam College, Cambridge, UK, from 18 - 21 July, 2013. Please visit www.pwsa.co.uk

If you would like to add your thoughts to our IPWSO blog:

<http://ipwso.blogspot.com> please feel free to do so. It is times like these that hearing from other people around the world make you feel less alone. Our thoughts and prayers from the IPWSO Board are with all our Japanese friends at this time...Linda Thornton

Brokkies

<p><u>Prader - Willi Syndrome Association of South Africa:</u> Entry fee R50. Annual membership fee R200. R220 members outside RSA. You are welcome to make a direct deposit into the savings account. Please ensure that your surname is included in the reference on the deposit slip or forward the deposit slip to the treasurer. Fax 086 551 5980</p>	<p>DVD <u>We're not that strange - Life with PWS in 2008</u> The Dutch Prader-Willi/Angelman association has recently produced an up-to-date informational documentary on PWS. The DVD shows the everyday life of 4 people, in the age of 2 to 32 years, with PWS in their own environment. In addition, we can see interviews with professionals and caretakers. The film is in Dutch, but thanks to a gift we were able to provide English subtitles as well, enabling this document to go out to a broader audience. Order a free copy from bassons@iafrica.com</p>
<p>BANK DETAILS of SAVINGS ACCOUNT PRADER-WILLI SYNDROME ASSOCIATION (SA) ABSA BROOKLYN, PRETORIA Branch number 632005 Acc. no. 113641800 Reference: Your SURNAME/cell number</p> <p>MEMBERSHIP FEES ARE DUE 1 APRIL 2011</p>	<p>Opinions expressed in <i>People With Strength</i> are those of the authors or editors and do not necessarily reflect the views of the management committee of the PWSA (SA). <i>People with Strength</i> welcomes articles, letters, personal stories and photographs, and news of interest to those concerned with Prader-Willi syndrome. Please send your contribution to PWSA (SA), PO Box 2399, Brooklyn, 0075. Fax 086 551 5980</p>
<p><i>PWSA(SA) is a registered non-profit organisation.</i> Nr 035-837-NPO PBO Exemption No 930 016 853</p>	<p>We are registered with: The Southern African Inherited Disorders Association (SAIDA) www.saida.org.za The International Prader-Willi Syndrome Organisation (IPWSO) www.ipwso.org WESTERN CAPE FORUM for Intellectual Disability (WCFID) www.wcfid.co.za</p>
<p><u>Wat is die Prader-Willi Sindroom-Vereniging van Suid-Afrika?</u> In Maart 1990 het 'n groepie toegewyde ouers hierdie vereniging gestig. Die hoofdoelwitte daarvan is die volgende:</p> <ul style="list-style-type: none"> ❑ Om aan ouers en versorgers van persone met PWS ondersteuning te bied ❑ Om kennis oor te dra en 'n bewustheid van die sindroom by die publiek sowel as lede van mediese en paramediese beroepe te kweek ❑ Om die vlak van versorging wat aan persone met PWS gebied word, te verbeter ❑ Publiseer en versprei "People With Strength" (nuusblad van PWSA(SA) vir ouers deur ouers) ❑ Hou jaarliks 'n sosiale byeenkoms om ouers en belangstellendes met kennis toe te rus en te ondersteun ❑ Verwys lede na kundige mediese dokters 	<p><i>PWSA (SA) acknowledge with gratitude the goodwill and support of:</i></p> <ul style="list-style-type: none"> ✓ <i>afrihost.com</i> for hosting the Association's website ✓ <i>Dawid Basson</i> the webmaster of the website. His advice on other matters is sincerely appreciated. ✓ <i>THE CO-WORKERS AND ALL THOSE WHO CONTRIBUTE TO "PEOPLE WITH STRENGTH" OUR SINCERE THANKS AND APPRECIATION</i> ✓ <i>Janet Drysdale & Magdaleen Kloppers</i> for the birthday cards ✓ <i>IPWSO</i> for another very informative December 2010 Wavelength and updated Medical Alert Booklet. Also available on www.ipwso.org ✓ <i>Incredible Door of Vereeniging</i> for the generous donation towards PWSA (SA) ✓ <i>Dr Engela Honey</i>, the medical advisor of the Association, who is always available

Brokkies

WOULD YOU LIKE TO MAKE A DONATION?

With your help we can make a difference!

PWSA (SA) is registered as a non-profit organisation (No. 035-837 NPO) as well as a public benefit organisation (PBO Exemption no.930 016 853). The PBO registration benefits donors and all donations made to PWSA (SA) are exempt from income tax. We will issue an official certificate for donations of R100.00 or more.

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

MAAK 'n VERSKIL MET 'n SKENKING!

Die PWSV (SA) is geregistreer as 'n nie-winsgewende organisasie (Nr. 035-837 NPO) ook as 'n openbare weldaadsorganisaie (PBO Exemption no.930 016 853).

Hierdie registrasie hou voordele in vir die donateur en donasies wat aan PWSV (SA) gemaak word is aftrekbaar van die donateur se belasbare inkomste. 'n Amptelike sertifikaat sal vir bedrae groter as R100.00 uitgereik word. Maak gerus 'n direkte inbetaling. Sluit asseblief 'n **selnommer** in as verwysing.

Would you like to join the PWSA (SA)?

Contact:

Chairperson: chairperson@praderwilli.org.za

Tel: 012 344 0241

or

Secretary: secretary@praderwilli.org.za

fax 086 551 5980

or

www.praderwilli.org.za

DVD

Help our children and help others to understand PWS better!

DVD: FOOD, BEHAVIOUR AND BEYOND

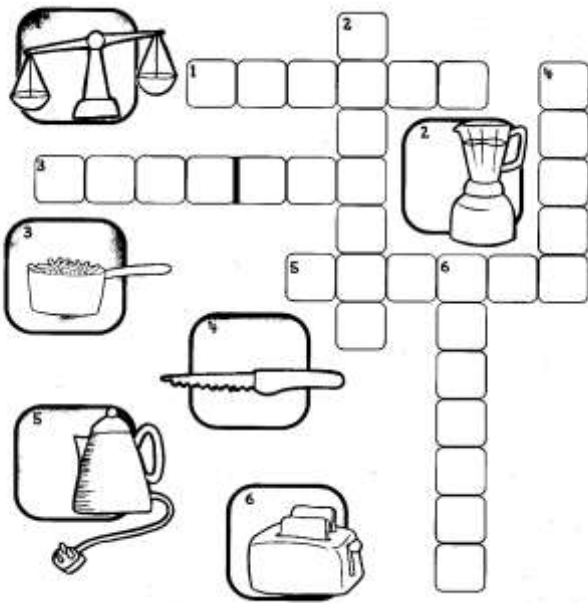
This comprehensive DVD which is a joint project of PWSA (USA) and IPWSO is an essential teaching and learning tool for parents and others involved with PWS. Having years of hands on intensive experience, Dr. Linda Gourash and Dr. Janice Forster shares very important TIPS for the everyday management of Prader-Willi Syndrome. Appetite and emotional aspects are addressed. The difference between food control and food security are explained. Typical and maladaptive behaviours, and to medicate or not, are also discussed.

Members, parents and other interested people are welcome to order above mentioned DVD (free of charge) from the PWSA (SA). It is a valuable tool in teaching caregivers, teachers and other professionals to have a better understanding of the person with PWS. If you are interested and want to make use of this opportunity, please contact the secretary bassons@iafrica.com Fax 086 551 5980

From *A Live less Perfect*. www.pwsausa.org

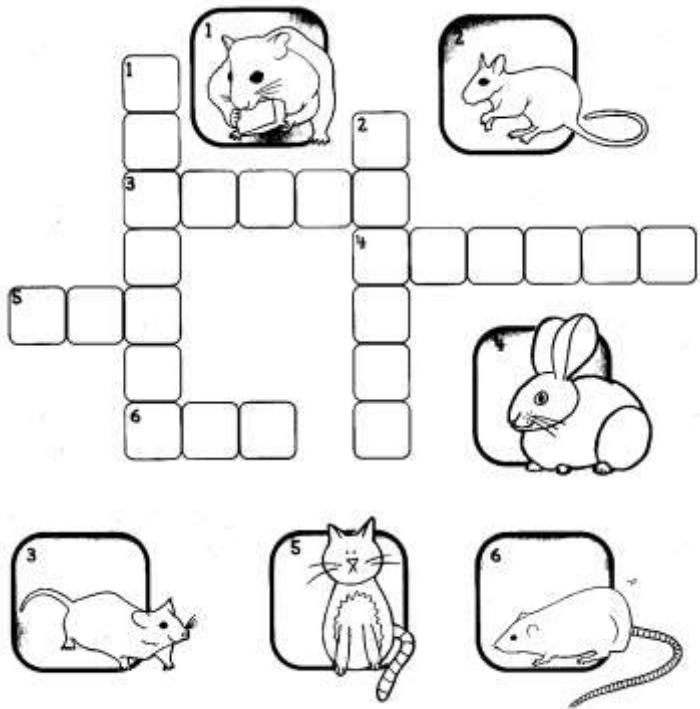
"I began to accept that my son is not like others in this world. I began to accept that this was not a curse but a blessing. To me he was unusually happy, loving and kind. I was amazed by his keen perception of human beings and his unique ability to make even the grumpiest of grouches smile brightly. He lives to dance and laugh and love. He has a warm heart and a gentle spirit. And although he is my child, he has also been my teacher".

PWS Fun Page



Do you know the items in a kitchen ?

Do you know the pets in the picture?



Answers kitchen
 1. SCALES 2. BLENDER 3. CHIP PAN 4. KNIFE 5. KETTLE 6. TOASTER
 Answers pets
 1. HAMSTER 2. GERBIL 3. MOUSE 4. RABBIT 5. CAT 6. RAT

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