People With Strength

Newsletter for parents by parents

Volume 15 Issue 3 **SUPPORT WORLDWIDE** November 2012

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

PWSA (SA) is a member of IPWSO. IPWSO cares for people with PWS worldwide and currently 100 countries are part of IPWSO. *Detail on page 17*

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FROM THE CHAIRPERSON

Dear Readers

This is the last newsletter for 2012. Thank you to everybody who rendered their support in different ways on a voluntary basis. Meet our volunteers - their photos appear elsewhere in this newsletter.

Make use of this opportunity to update your knowledge or to refresh your memory on the Prader-Willi syndrome before the summer holiday arrives. Our readers have divergent interests in PWS and I hope that you will find valuable information that is applicable to your situation. Articles on genetics, for instance, are usually not read because they are too long and complicated. But don't skip Suzanne Cassidy's article on the different genetic subtypes found in PWS. It is indeed interesting and readable. Much is still to be learnt about genetics and PWS.

Another article deals with the drug called **Qsymia**. Much research is nowadays being done on remedies which will have a positive effect on the appetites of persons with PWS. I also refer to an advertisement in our newsletter of June 2012. It is about the 2nd International Conference on Hyperphagia in Louisiana, USA, which was held in October 2012. Well-known specialists on hunger, obesity and the uncontrollable drive to eat were present. Janalee Heinemann from the USA is very positive about the outcome of this conference and we are looking forward to the feedback which she will give us shortly.

Matters regarding the life expectancy of persons with PWS are often under discussion. Read about a study done with 12 persons with PWS – all of them over 50 years of age. Interesting facts and guidelines are given. Parents and caregivers, who are involved with adult persons with PWS, will definitely find this article very useful.

The two articles received from IPWSO contain information for law enforcement personnel – people every parent would like to avoid in an official capacity. But what if it happens? The information was compiled by PWSA (USA) and it gives a clear background and reasons why a person with PWS can commit a crime, such as theft. Useful hints are given for law enforcement personnel when they are faced with such a crisis situation. This article is also electronically available on request. Contact the chairperson of PWSA (SA) or visit www.pwsausa.org.

A questionnaire was sent electronically to parents with adult children with PWS at the beginning of August 2012. The FamCare Committee under leadership of Georgina Loughnan of Sydney, Australia, received 81 completed questionnaires, including three from South Africa. Many thanks to these three families who responded to the questionnaire. The aim of this questionnaire was to gather practical information to help parents with adult children in exercising effective control over all surroundings where the young adult may venture. The information will also help them to avoid obesity and difficult behaviour. Where an adult with PWS is still staying with his or her parents it will always be the family's responsibility to

create the best possible environment for this child. Interesting, parents find the occurrence of skin picking the most difficult to handle. Famcare will first of all address this aspect. All information is first submitted to a professional committee and afterwards to IPWSO. Parents, who still wish to complete this questionnaire, can contact FamCare. More information about FamCare can be found under IPWSO News in this issue of the newsletter.

The Huisgenoot/You of 18 October 2012 carry an article on Ruan Burger who was diagnosed with PWS before he was four years old. At present he is 16 years old. This article creates an awareness of PWS among the general public. Arising from this article we have already received five enquiries. We appreciate the fact that these family members were prepared to expose themselves by reacted in a very positive manner to the request of Huisgenoot.

Do look at our potpourri of information. Sources are given where obtainable. To those readers who don't have access to the internet, the PWSA (SA) will gladly supply the necessary information.

Please take note of the following:

- Notices which appear under IPWSO News
- The IPWSO 8th INTERNATIONAL CONFERENCE which will be held in Cambridge in July 2013
- IPWSO Medical Alert Booklets are still available
- Two DVDs which are available at no charge
- Photos taken at the AGM
- Feedback on Huis Henri
- A request from Elize Robarts
- This newsletter is also available electronically in A4 format

Summer has arrived! The rain in Pretoria is most welcome. Everything is green. The jacaranda blossoms add an unusual colour to the city. I feel energised by all the lovely things. We can be truly grateful.

You earn a break. Enjoy the festive season with family and friends.

Sincere greetings
Rika du Plooy.

VAN DIE VOORSITTER

Liewe Lesers

Hierdie is die laaste nuusbrief vir 2012. Hiermee baie dankie aan almal wat die komitee ondersteun het en aan hulle wat 'n vrywillige diens gelewer het. Foto's is geplaas - maak gerus met hierdie mense kennis.

Gebruik hierdie geleentheid om voor die somervakansie nog interessante inligting oor die Prader-Willi-sindroom te bekom. Die lesers het uiteenlopende belange by PWS en ek vertrou dat daar in hierdie nuusbrief vir elkeen iets waardevols sal wees. Ons is dalk geneig om

artikels oor genetika nie te lees nie, omdat dit dikwels lank en ingewikkeld is. Moenie die artikel deur Suzanne Cassidy oor die verskillende genetiese subtipes wat by die Prader-Willi-sindroom voorkom, oor te slaan nie. Die inhoud is interessant, verstaanbaar uiteengesit en soos sy te reg opmerk, is daar nog baie om oor die genetika van PWS te leer.

'n Ander brokkie interessante inligting gaan oor die middel **Qsymia.** Navorsing word ernstig gerig op 'n middel wat 'n positiewe uitwerking op die aptyt van die persoon met PWS kan hê. Hierby aansluitend - in die Junie 2012 uitgawe is 'n advertensie geplaas van die *2nd International Conference on Hyperphagia,* in Louisiana, VSA, wat in Oktober 2012 gehou is. Bekende kundiges op die gebied van honger, obesiteit en onversadigbare eetlus was byeen. Janalee Heinemann (VSA) voel baie positief oor die uitkoms en ons sien uit na die die terugvoer wat binnekort gegee sal word.

Die vraag oor die lewensverwagting van persone met PWS word dikwels gevra. Lees gerus die artikel wat handel oor 'n studie wat gedoen is met 12 persone met PWS wat 50 jaar en ouer was. Interessante feite en riglyne word gegee. Ouers en versorgers wat by volwasse persone met PWS betrokke is, kan beslis hierby baat vind.

Twee artikels is vanaf IPWSO ontvang wat gerig is op inligting aan wetstoepassers.
Geen ouer wil in hierdie situasie beland nie, maar dit kan wel gebeur. Die inligting is opgestel deur PWSA (USA) en gee 'n duidelike agtergrond en redes waarom die kind/volwassene met PWS by diefstal betrokke kan raak. Waardevolle wenke aan die wetstoepasser oor hoe om in 'n krisissituasie op te tree word gegee. Hierdie artikels is op aanvraag eletronies beskikbaar. Kontak gerus die voorsitter van PWSV (SA) of besoek www.pwsausa.org.

Begin Augustus 2012 is 'n vraelys aan ouers wat volwasse kinders met PWS in die huis het, per epos gestuur. Die FamCare komitee onderleiding van Georgina Loughnan van Sydney, Australië, het 81 vraelyste terug ontvang, waaronder drie uit Suid-Afrika. Baie dankie vir die betrokkenheid van hierdie drie gesinne. Die doel van die vraelyste is om praktiese inligting saam te stel om ouers van volwassenes wêreldwyd, te help om alle omgewings waar die volwassene dit mag waag, doeltreffend te beheer. Die ondersteuning is daarop gerig om oorgewig en moeilike gedrag te voorkom. Waar 'n volwassene in 'n ouerhuis is, sal dit altyd die familie se verantwoordelikheid wees om die bes moontlike omgewing te skep. Dit is interessant om daarop te let dat "skin picking" die aspek is wat vir ouers die moeilikste is om te hanteer. Die komitee gaan hierdie aspek eerste aanpak. Alle inligting word eers aan 'n professionele komitee voorgelê en dan aan IPWSO. Ouers wat steeds graag die vraelys wil invul, kan dit vanaf Famcare aanvra. Meer inligting oor FamCare en kontakbesonderhede is onder *IPWSO News.*

In die Huisgenoot/You van 18 Oktober 2012 was daar 'n artikel oor Ruan Burger wat voor die ouderdom van vier jaar met PWS gediagnoseer is. Hy is nou 16 jaar. Die artikel dra by tot groter bewuswording onder die algemene publiek. Daar is na aanleiding van die artikel reeds vyf navrae ontvang. Ons het waardering vir Ruan se familie, wat ten spyte van die blootstelling, tog positief op Huisgenoot se versoek gereageer het.

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

Neem ook asseblief kennis van die volgende:

- Inligting wat onder IPWSO Nuus verskyn
- die *IPWSO 8th INTERNATIONAL CONFERENCE* wat Julie 2013 in Cambridge gehou word
- *IPWSO Medical Alert Booklets* is steeds beskikbaar
- Twee DVD's wat gratis beskikbaar is
- In die foto-album 'n paar foto's van die AJV
- Terugvoer oor Huis Henri
- Versoek van Elize Robarts
- Hierdie nuusbrief is ook elektronies in A4-formaat beskikbaar.

Die somer is hier! In Pretoria het ons heerlik reën gehad, dit is groen en die jakarandas blom! Die mooi het beslis in my ook 'n nuwe energie gebring en daar is soveel om voor dankbaar te wees.

Aan almal 'n welverdiende somervakansie. Mag die kerstyd vir almal 'n geseënde tyd wees.

Opregte groete
Rika du Plooy.

We are excited to announce that we have launched a Facebook page.
Please visit us at www.facebook.com/pwsasa
Ons is opgewonde om aan te kondig dat ons 'n Facebook blad geskep het.
Besoek ons gerus by www.facebook.com/pwsasa

Elize Robarts from Bloemfontein has a 27 year son with PWS. She would like to have contact with other parents in a similar situation. Elize works during the day. The family is very fond of their pets, likes fishing and Elize participates in angling competitions.
Please email Elize at accounts@goodfrank.co.za

BIRTHDAY CARDS…

We are happy to announce that **Karin Clarke** volunteered to do the English cards. **Magdaleen Kloppers** will continue with the cards to Afrikaans speaking children. It does happen that birthday cards don't reach their destination. Please send a notification via SMS to the appropriate lady when your child receives his/her birthday card. **Magdaleen: 083 663 7234** **Karin: 083 681 6842**

Please help ……who made this deposit?
A cheque deposit was made on 3 August 2012 in Eastgate, Gauteng for the amount of R200.00 - no reference. It might be for membership or a donation?
The treasurer would like to issue a receipt.

MEET OUR VOLUNTEERS

Volunteers are STARS!

People, all over our country contribute their time and knowledge. Thank you for your support.

Johan and Elmaré Mostert - Financial statements

Johan and Elmaré with their two daughters.

Dawid Basson -Webmaster and responsible for Facebook

Dawid, Corneli and their three sons

Karin Clarke - birthday cards (English)

David, Karin and the kids as pirates at a party.

Magdaleen Kloppers - birthday cards (Afrikaans)

Magdaleen is since 1994 a member of PWSA (SA)

Francis Morrison: Translation

Francis is a freelance translator

Elsa Volschenk - Layout of newsletter

Elsa has been assisting PWS since November 2011

Wilna Basson - Library and educational material

Wilna served for many years as secretary of PWSA (SA)

DIFFERENCES BETWEEN THE GENETIC SUBTYPES OF PRADER-WILLI SYNDROME

Dr Suzanne Cassidy
Permission is granted by PWSA (USA) and *The Gathered View* to publish this article.
The Gathered View (ISSN 1077-9965), September-October 2012

Why do different genetic changes cause PWS?

Dr Cassidy is currently President of the International Prader-Willi Syndrome Organisation (IPWSO). She also has served for many years on the PWSA (USA) Scientific Advisory Board.

PWS is always due to the same thing: deficiency in the expression of certain genes on chromosome 15. Specifically, the deficient expression is from genes that are normally expressed from the member of the chromosome 15 pair that was inherited from the father, because the copy of these genes on the chromosome 15 inherited from the mother is normally not expressed ("imprinted"). This lack of expression on the maternally-inherited copy is the result of a chemical reaction called DNA methylation. So, in the general population, the genes relevant to PWS are only expressed (like reading a blueprint) from the chromosome 15 inherited from the father, and in PWS, that copy is missing (when there is deletion or uniparental disomy); or else the chromosome 15 inherited from the father is erroneously switched off ("imprinted") like the genes on the chromosome 15 inherited from the mother (an imprinting defect).

A deletion is a missing piece of a chromosome. In PWS, the missing piece is at the top of the long arm (called q) of chromosome 15, in a region called 15q11.2-q13. There are three common points along the chromosome in which the break occurs, two at one end with four genes between them and one at the other end. The type of deletion missing the larger piece of genetic material is sometimes called a Type 1 deletion, and the smaller deletion is sometimes called Type 2. A few deletions are larger or smaller than the common deletions, but most genetic testing for PWS does not identify those differences.

Maternal uniparental disomy (also called UPD) is a situation in which both of the members of the chromosome 15 were inherited from the mother (and the genes relevant to PWS are therefore not expressed from either copy). The chromosome 15 that should have been inherited from the father—the one with the expressed genes for PWS—is missing. This situation is somewhat more common among children with PWS born to women over age 35 years.

An imprinting defect is a condition in which there is one chromosome 15 from the mother and one from the father, but the one from the father behaves as though it were inherited from a mother, at least as far as the genes for PWS are concerned. Those genes relevant to PWS are switched off in both the chromosome 15 inherited from the mother (as is normal) and the one inherited from the father (which is not usual). Most cases of imprinting defect are random and of unknown cause, but a small percent are due to a small deletion in the i1mprinting center which is responsible for switching genes nearby off and on (applying or erasing DNA methylation). In some families that have a child with PWS due to an imprinting center deletion, there is a significant risk to have another affected child. Therefore, in

families planning future children, further genetic testing and genetic counselling are important.
A few people who have PWS are found to have a **"translocation**", which is a rearrangement of two chromosomes such that part of one is stuck onto part of another. Most of the time that this happens in someone with PWS, the translocation involves one chromosome 15 and there is a deletion on the rearranged chromosome. In that case, the individual in effect is like people with PWS due to a deletion. Occasionally, a translocation can cause uniparental disomy 15, and the individual in effect is like people with PWS due to UPD. Sometimes with a translocation, there is an effect from another missing piece of chromosome in addition to it causing PWS.

What are the differences in the effects of the different genetic causes of PWS?
Researchers, by studying many people with each genetic type of PWS (primarily the most common deletion and uniparental disomy types) have identified a few differences between the groups. Most of these differences are in the frequency or severity of findings in the two groups. People with an imprinting defect appear to be most similar to those with UPD.

Before mentioning these differences, it is very important to explain that these are GROUP differences, not individual differences. *There is no feature that is exclusively found in one of the three genetic categories.* And it is also important to recognize that even within a genetic type there is a lot of variation among people affected with PWS, so there is a lot of overlap in the genetic groups. For example, even though the people with UPD as a group have a slightly higher mean intelligence quotient (IQ) than people with deletion as a group (8 points, with an IQ of 100 being the average in the general population), there is a very wide range of IQ within each genetic group (about 40-90). So in an individual, knowing the genetic type is not very helpful in knowing the future abilities or problems of the individual.
In general, and as a group, people with a deletion are more likely to have fair colouring (hair, eyes, skin), whereas those with UPD more closely resemble their parents. People with a deletion are more likely than those with UPD to have the characteristic facial appearance of PWS. And they are more likely to be skilled with jigsaw puzzles.
In general, and as a group, people with UPD are more likely to be born late. As noted above, they have a slightly higher average IQ than the group with deletion. In addition, they have somewhat milder behaviour problems (temper tantrums, stubbornness, repetition and controlling behaviour). However, it appears that people with UPD as a group have a significantly higher incidence of autistic characteristics and of psychiatric disorders, including psychosis, than those with deletion.
In addition to these differences, there are some studies suggesting somewhat worse behavioural problems in those with a Type 1 (slightly larger) deletion versus those with a Type 2 deletion. There are also some studies that have not shown these differences.
It is important once again to stress that knowing the genetic type does not predict the manifestations of PWS in an individual. It is primarily important for genetic counselling purposes, as far as we know today.

Why are there clinical differences between the different genetic types?
The genetic or biological basis for these differences has not been determined. In people with a deletion, a number of genes are missing, and some of those genes are normally expressed from both members of a chromosome pair. In people with UPD or an imprinting defect, the genes are not actually missing, but are just not being expressed (turned into messenger

molecules, regulators or proteins). This presence of some genes that, as far as we know, are not the ones causing PWS but are also in the chromosome segment included in the deletion, may have an impact on the clinical manifestations in an individual. There is still much to learn about the genetics of PWS!

WEIGHT LOSS DRUG APPROVED BY FDA

By Janalee Heinemann, M.S.W. PWSA (USA) Director of Research & Medical Affairs. Permission is granted by PWSA (USA) and *The Gathered View* to publish this article. *The Gathered View* (ISSN 1077-9965), September-October 2012

A new weight loss drug, **Qsymia**, has been approved by the U.S. Food & Drug Administration (FDA). Weight loss was more with this drug than with two others recently reviewed by the FDA with one approved (Lorcaserin/Belviq) that was not nearly as remarkable regarding weight loss. Qsymia is a combination of two older drugs that have long been known to help with weight loss: phentermine (the safer half of the old fen-phen drug that was banned) and topirimate – a drug that was studied back in April 2000 under a PWSA (USA) grant, *"Open-Label Pilot Study of Topirimate in Adults with Prader-Willi Syndrome."*

Topirimate is an anti-consultant drug that makes people feel more satiated after eating, which is why we sponsored the study by Nathan A. Shapira, M.D., Ph.D. The study showed that Topirimate did not significantly change the calories consumed, Body Mass Index, or decreased self-reported appetite in PWS; there were no significant changes in compulsions. Surprisingly though, Topirimate treatment resulted in a clinically significant improvement in the self-injury (i.e., skin-picking) characteristic of PWS.

The researchers of Qsymia state that it targets multiple brain signals that drive people to overeat. We cannot say at this time if it will be effective with PWS. Please know that we will keep you informed of any new outcomes on this and other obesity drugs that might be helpful with PWS. As with any new medication, if you wish to try it before it has been proven to be effective in PWS, it is most useful to do so as part of a clinical trial.

Currently, I have been working informally and under strict agreements of confidentiality with two pharmaceutical companies which are working on potential products that might impact on PWS. We are also working with the Foundation for Prader-Willi Reaserch on financially supporting the Best Idea Grants post our 2nd International Hyperphagia Conference (go to www.hyperphagia.org) with 2012 One Small Step funds. We never forget that the #1 deadly enemy of PWS is the appetite.

AGING IN PRADER-WILLI SYNDROME: TWELVE PERSONS OVER THE AGE OF 50 YEARS

Summary of research article by Sinnema M, Schrander-Stumpfel CTrm, Maaskant MA, Boer H, Curfs LMG. 2012. Am J Med Genet Part A 158A: 1326 – 1336.

This study looked at 12 individuals with PWS aged 50 and older. Individuals with PWS aged 18 – 49 years were used as a control group.

Age range 50 – 66 years

- 5 males, 7 females
- 4 with deletion, 8 with mUPD (maternal disomy)
- 11 in community or residential facilities, one living at home with elderly mother
- Mean age at moving to residential facility – 19.4 years
- Mean BMI in persons with deletion was significantly higher than persons with mUPD – three of the latter had a BMI under 25.
- Mean maximum BMI was 36.5, with a range of 23.6 – 44.4
- 3 people smoked (cigarettes, pipe and cigars)

Health Issues

- One woman died, aged 65, shortly after data collection, due to lung problems
- Half had diabetes mellitus, mean age of diagnosis 41.6 years
- No one in this group had received sex or growth hormone therapy
- No one had epilepsy or cancer

Physical health problems	Total prevalence (N)
Hypertension	3 out of 12
Stroke	3/12
Diabetes	6/12
Pneumonia	3/12
Excessive daytime sleepiness	8/12
Constipation	5/12
Reflux	2/12
Aenemia of unknown origin	2/12
Kidney problems (congenital)	1/12
Osteoporosis	2/12
History of any fracture	6/12
Primary amenorrhea (no menstruation at any time in life)	1/7
Scoliosis	5/12
Foot problems	10/12
Hip problems	2/12
Oedema	9/12
Erysipelas (skin infection)	6/12
Varices (varicose veins)	3/12

Comparison with younger people with PWS

- Functioning, behaviour and care dependency revealed worse functioning in the older than the younger groups.
- Scores in the older group were significantly lower on the following items:- personal hygiene, dressing, eating, being ambulant, mobility, grooming, memory, orientation, sleeping difficulties, physical complaints, hearing, vision, dependency on medical care and care dependency (*note that deterioration in some of these areas would also be evident in the over 50 population without PWS)*

- Older individuals scored higher on bizarre speech, gorging food, masturbating or exposing oneself in public, and hallucinating.
- Scores on lack of self-confidence or poor self-esteem were, however, statistically significantly lower than those for people under 50.
- No significant difference in eating behaviour between over and under 50s.
- No reduction in behaviour problems as people got older, as opposed to findings in some previous studies. More behaviour problems were noted in those with mUPD, which may overlap with the psychiatric disturbances that this sub-group displays.

Psychiatric illness

- No-one with deletion (4 people)had a psychiatric illness
- 7 out of 8 mUPD had a history of psychiatric illness:
- Bipolar disorder with psychotic symptoms – 3
- Psychotic illness – 2
- Depressive illness without psychotic symptoms – 1
- Bipolar disorder – 1
- All with mUPD used psychotropic medication.
- One woman presented with symptoms highly suggestive of dementia

Health checks

- The study underlines the need for regular health checks for adults with PWS. In particular, re:
    - Cardiovascular disease
    - Diabetes
    - Dermatological problems
    - Orthopaedic problems
    - Sleep problems
    - Osteoporosis
    - All usual age-appropriate screenings should be carried out (eg hearing, eyes, cancer etc, with possible exception of cervical smear tests for women with no history of sexual activity.
- Sleep problems and osteoporosis are likely to be under-reported and deserve special attention.
- Diagnosis of pneumonia is frequently delayed in older adults with PWS because of absence of fever.
- No indication that sensory impairment or cancer is more prevalent in PWS than in the general population (bearing in mind that this was a very small study)

Functional decline

- From 40 years onwards, individuals showed decreased energy levels and lessening of mobility (*as it evident in most of the general population*)
- Special medical surveillance should be available to those aged 40 and older to ensure significant medical, behavioural and social issues are not overlooked.

Other points

- There were relatively more individuals in this study in the moderate-severe learning disability range. These individuals may have required earlier intervention by being placed

in structured residential settings at an earlier age, which in turn could have contributed to their longevity because of better weight management and prevention of serious medical complications.

- The researchers remarked on the predominance of those with mUPD type of PWS in this age group (*Across all age groups, mUPD is estimated to account for about 25% of the entire PWS population)*. They comment that this may have implications for the survival rates of different genetic sub-types, but stress that more studies are needed to check these findings.
- The researchers commented, "Undiagnosed psychopathology or physical morbidity can have an atypical presentation in people with PWS …" and emphasise that support in residential settings should be adjusted to fit the higher levels of care dependency, the different needs in the day care programme, and diminishing mobility.
- Greater likelihood of bereavement for these people, as parents die. Recognition that this may cause behavioural disturbances and emotional distress.
- The researchers hypothesize that there may be premature aging in PWS, especially where no sex or growth hormone is given. They state that aging in PWS starts at 50 or younger.
- With better management, the number of people with PWS aged over 50 is likely to increase.

* * * * *

In the PWSA UK

We are currently aware of 30 individuals aged 50 or older: 27 in England, 2 in Scotland, 1 in Eire. Of these 30, three people are aged 60 or older. The oldest is aged 66.

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

The child of this parent has Prader-Willi syndrome (PWS). Because of the unique behavioural and emotional characteristics related to this rare syndrome it is imperative that you read the following information in order to better understand the situation you've encountered. PWS is a genetic disorder that causes the brain to function in a way that is not typical of other children. This brain dysfunction can cause problems that often lead to the involvement of law enforcement because:

(1) The brain of a child with PWS sends a message of constant hunger due to an inability to register a feeling of "fullness" after eating regardless of the amount of food ingested. As a result, sneaking or stealing food is common and due to this intense drive to eat, a child with PWS cannot always control themselves around food. The reality is that any person who felt the same way would react in a similar manner. No medication, to date, is effective in curbing the insatiable appetite of a person with PWS. And because food is everywhere in our society, it is impossible for a parent to always avoid situations where their child with PWS might have an opportunity to "steal" food so theft can occur.

(2) Over consumption of food can be medically dangerous and even life threatening for a child with PWS. For this reason parents and caregivers are encouraged to practice strict food security in their household. This food security practice can include locking refrigerators,

cabinets, and any other place food is kept. It is very important to understand that such measures are not an indication of child abuse. These precautions are taken to safeguard a child with PWS and are not signs of a "bad" or punitive parent. Such food security practices are encouraged by PWSA (USA) and other medical and professional PWS experts.

(3) The same part of the brain that regulates appetite (the hypothalamus) also regulates temper and emotional response. As a result, it is not uncommon for a child with PWS to experience significant temper tantrums and behavioural outbursts. Such outbursts are a feature of PWS. If the child you encounter is experiencing a tantrum or emotional meltdown, we recommend allowing the child space to work their way out of the tantrum if they do not pose a danger to self or others. Responses such as over talking and apply punitive approaches will most likely result in an escalation of behaviour. In addition, due to respiratory features of PWS, injury and even death can occur if improper physical restraint is applied. Often the parent(s) of a child with PWS can help you with the best suggestions for how to respond to their child when they are upset. Typically it is important to remain calm and use clear and simple directions with the child.

(4) A small portion of children with PWS exhibit "running" or elopement behaviour often for no identifiable reason (although in some cases it is to pursue food). For this reason, some parents will need to utilize motion detectors and/or locks to keep their child from leaving home without supervision. Again, this is a practice put into place for the safety of a child – not as a punitive or abusive action.

(5) Skin picking and some forms of self abuse are also common so it is not unusual for a child with PWS to exhibit sores and scars in various stages of healing. At first glance this can look like the signs of possible caregiver or parental abuse. And, while we encourage all potential situations of abuse to be appropriately addressed, it is important to take this information into account when investigating potential abuse involving a child with PWS.

(6) Some children with PWS will also call 911 when upset with their caregiver. It is important to remember that children with PWS are most often very sweet and loving children who, with the help of their parents, are positive and helpful members of their community. Occasionally, however, due to their disability they will experience behavioural problems such as we've described. We hope this information will help you place these behaviours in the appropriate context so that you are able to offer the support and help they need when encountering law enforcement.

Many helpful resources are available on the website PWSA (USA) at www.pwsausa.org.
9/4/2012

The two articles on Law Enforcement were received from IPWSO with the request that delegates should distribute it in their countries.
Our appreciation to PWSA (USA) for their willingness to share this important information.

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

The child of this parent has Prader-Willi syndrome (PWS). Because of the unique behavioural and emotional characteristics related to this rare syndrome it is imperative that you read the following information in order to better understand the situation you've encountered. PWS is a genetic disorder that causes the brain to function in a way that is not typical of other children. This brain dysfunction can cause problems that often lead to the involvement of law enforcement because:

1. The brain of a person with PWS sends a message of constant hunger due to an inability to register a feeling of "fullness" after eating regardless of the amount of food ingested. As a result, sneaking or stealing food is common and due to this intense drive to eat, a person with PWS cannot always control themselves around food. The reality is that any person who felt the same way would react in a similar manner. No medication, to date, is effective in curbing the insatiable appetite of a person with PWS. Food can be dangerous and even life threatening for people with PWS so parents and caregivers do all that they can to prevent exposure to food that is not secured. But, because food is everywhere in our society, it is impossible for a person with PWS to always avoid situations where they might have an opportunity to "steal" or take food so theft of food can occur. In addition, people with PWS may also steal non-food items (i.e. money) usually in an attempt to buy or trade for food. When theft occurs, PWSA (USA) encourages you to work cooperatively with the parent(s) or caregiver to resolve the situation through implementing new positive behavioural and environmental supports. Especially because when a person with PWS engages in theft of any kind it is a manifestation of their disability rather than an indication of wilful criminal intent.

2. The same part of the brain that regulates appetite (the hypothalamus) also regulates temper and emotional response. As a result, it is not uncommon for a person with PWS to experience significant temper tantrums and behavioural outbursts. Such outbursts are a feature of PWS. We encourage you in response to allow the person with PWS the space to work their way out of a tantrum if they do not pose a threat to self or others. Responses such as over talking, threats or other punitive approaches will most often cause an escalation of behaviour. In addition, due to respiratory features of PWS, injury and even death can occur if improper physical restraint is applied.

3. Over consumption of food can be medically dangerous and even life threatening for a person with PWS. For this reason parents and caregivers are encouraged to practice strict food security in their household. This food security practice can include locking refrigerators, cabinets, and any other place food is kept. It is very important to understand that such measures are not an indication of abuse. These precautions are taken to safeguard a person with PWS and are not signs of a "bad" or punitive parent. Such food security practices are encouraged by PWSA (USA) and other medical and professional PWS experts.

4. Since behavioural instability can be a feature of PWS there are other unusual behaviours that can be the cause of police attention including:

Calling 911 when upset with a caregiver. This can include accusations of abuse or some type of mistreatment by a parent or caregiver.
Destruction of property when upset – most often when denied food.
Elopement or "running away" behaviour often for no identifiable reason - although in some cases it is to pursue food.
These behaviours, while actively discouraged by parents and caregivers, should also be treated as a manifestation of the person's disability rather than a criminal act. Appropriate responses include working with the parent(s) and or caregivers to develop positive behavioural solutions in each situation to discourage repeat incidents in the future.
PWSA (USA) is happy to serve as a resource in these situations to make behavioural suggestions based on best practices when caring for a person with PWS.

5. Skin picking and some forms of self abuse are also common so it is not unusual for a person with PWS to exhibit sores and scars in various stages of healing. At first glance these can look like the signs of possible caregiver or parental abuse. And, while we encourage all potential situations of abuse to be appropriately addressed, it is important to take this information into account when investigating potential abuse involving a person with PWS.

Tips for Working with a Person with PWS in Crisis
It is important to remember that people with PWS are most often very sweet and loving people who, with the help of their parents and other caregivers, are positive and helpful members of their community. Occasionally, however, due to their disability they will experience behavioural problems such as we've described. We hope this information will help you place these behaviours in the appropriate context so that you are able to offer the support and help they need when encountering law enforcement.

While each person with PWS is unique, the following tips will help to create a more successful response to a person with PWS who is in need of police assistance:

- Clearly identify who you are and your role as a helper to the person with PWS.
- If a caregiver or parent is present, before directly addressing the person with PWS, ask the caregiver/parent if they have suggestions for the most effective way for you to approach the person.
- Utilizing a calm voice and non-anxious presence at all times will help to de-escalate a person with PWS who is upset.
- Avoid any threats of jail, punishment, or confrontational tactics. Such negative approaches typically are not effective for people with PWS and actually can significantly worsen the situation.
- Please remember a person with PWS needs your help in understanding what is happening and why you are involved.
- Unless under the most extreme circumstances avoid physical contact or using restraints of any kind. People with PWS will often react negatively if they feel backed into a corner and this can result in physical and verbal aggression and other destructive behaviours.
- PWS is fundamentally a disability that severely impairs a person's ability to self-regulate critical functions such as appetite, emotion, and temper response. It can also create cognitive impairments as well. Therefore your role as a professional is critical in helping a person with PWS manage their emotional or behavioural crisis with minimal harm to self and others.

Many helpful resources are available on the website PWSA (USA) www.pwsausa.org.

A POTPOURRI OF INTERESTING INFORMATION

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

TWO BOOKLETS FROM THE UNIVERSITY OF QUEENSLAND, AUSTRALIA.

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

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

NEW BOOK FOR PARENTS OF BABIES AND CHILDREN WITH PWS

Miracle In Potential, by Australian author Joanne Griggs, is an inspirational story and early intervention resource. This sensitively-written 267-page soft cover book outlines how Joanne and husband Adam created the Multiple Initiative Approach (MIA) to overcome their daughter Mia's global development delay which was due to Prader-Willi syndrome. With a foreword by Associate Professor Dinah Reddihough, Director of Developmental Medicine, The Royal Children's Hospital Victoria, Australia, this "intervention lifestyle" program is based on the idea that everyday life and resources can be used as intervention, when targeted to bring out your child's potential.

From the time Mia was four months old, the family creatively constructed an approach to combat the difficulties related to low muscle tone, global developmental delay, vision impairment, obsessive compulsive behaviours and other behavioural problems associated with food. The strategies and processes they developed are individualized to your family/child's needs and are easy to follow.
www.miaresearchfoundation.com or to order info@pwsusa.org

BEST PRACTICE RECOMMENDATIONS

Guidelines for Residential and Environmental Structure in New Zealand by Linda Thornton, National Director, PWSA New Zealand, July 2010.

This document is developed from the IPWSO Best Practised Guidelines.
The document has 158 pages and the file is 3MB. This document not only provides a high level of skilled management, but also assists to recognise the individual needs of each person with PWS.
Available via email from the chairperson PWSA (SA): chairperson@praderwilli.org.za

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

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

NEW! ELEMENTARY SCHOOL POWER POINT - DALE AND DOTTIE COOPER

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

INFORMATION SHEET: SELF-INJURIOUS BEHAVIOUR

Self-injurious behaviour was defined by Murphy and Wilson (1985, p. 15) as:
'Any behaviour, initiated by the individual, which directly results in physical harm to that individual. Physical harm includes bruising, lacerations, bleeding, bone fractures and breakages, and other tissue damage.'
From: The Challenging Behaviour Foundation – making a difference to the lives of people with severe learning disabilities. www.challengingbehaviour.org.uk

IPWSO NEWS

PLEASE VISIT IPWSO's blog and become a FRIEND of IPWSO. Read the different stories as well as all about the activities of IPWSO around the world.
http://ipwso.blogspot.com

NEW! FAMCARE - A NEW INITIATIVE FROM (IPWSO) FOR THOSE CARING FOR AN ADULT WITH PWS AT HOME

FamCare is an international committee that has been formed for the purpose of supporting families who have their adult son/daughter/relative with PWS living in the home with them. Georgina Loughnan, an IPWSO Board member, is heading the project. She is supported by a committee made up of parents of adults with PWS and experienced professionals. The committee is also most fortunate to have a highly skilled team of advisors.

Parents or other family members, who are caring for adults with PWS living with them in the family home, are asked to complete this survey, as honestly as possible, to help the committee to determine the needs of parents and the manner in which FamCare can be most supportive. Answers will remain confidential.

The questionnaire is available from Georgina Loughnan at famcare.pws@gmail.com. The committee will be most grateful if parents would take the time to complete the survey and return it by email to famcare.pws@gmail.com.

(The questionnaire was already sent to members of PWSA (SA) with adults with PWS living in the home. Please ask for the questionnaire and support this worthy initiative. PWSA (SA) chairperson)

A REQUEST: PARENT SUGGESTIONS REQUIRED FOR SKIN PICKING ARTICLE

FamCare is writing an article on skin picking and would like to add suggestions from parents for parents of strategies/practices that have been successful in either ceasing the behaviour or avoiding it. Do you know of any that have worked well for your son/daughter? I look forward to hearing many positive replies.

Regards, Georgina Loughnan

The Metabolism & Obesity Services & the Prader-Willi Syndrome Clinic in Australia.

Contact Georgina at georgie@email.cs.nsw.gov.au

FamCare committee:

Georgina Loughnan (clinician)	Australia	georgie@email.cs.nsw.gov.au
Kate Beaver (crisis ounsellor)	USA	kbeaver@pwsausa.org
Fanny Cortes (geneticist)	Chile	fcortes@clinicalascondes.cl
Evan Farrar (crisis counsellor)	USA	cic3@pwsausa.org
Sabeen Iqbal (parent)	USA	sabeeniqbal5@yahoo.com
Damien Jones (caregiver manager)	Australia	djones@interactiondisablity.com
Rika du Plooy (parent)	South Africa	rikadup@mweb.co.za
Lesley Robertson (parent)	Australia	lesdonrobertson@tpg.com.au
Linda Thornton (parent)	New Zealand	linda.thornton@xtra.co.nz

REPORT FROM THE EUROPEAN SOCIETY FOR PAEDIATRIC ENDOCRINOLOGY CONFERENCE HELD IN LEIPZIG (GERMANY), SEPTEMBER 2012.

Excerpted from a report written by Janalee Heinemann, MSW, Director of Research & Medical Affairs, PWSA (USA) and Vice President, IPWSO.

The International Prader-Willi Syndrome Organisation (IPWSO) was again at the ESPE conference in Germany and as usual it was a big success. The awareness and education booth was visited by 532 doctors from 70 different countries. These doctors will share the information on PWS with their staff and PWS families. Tunisia and Kosovo joined the IPWSO family which increases IPWSO's member countries to 100. The most significant change which was noted was the dramatic increase of interest from physicians from Arabic countries. China and Argentina also shown interest and visited the booth for information. Attendees appreciated the fact that the *Medical Alert booklet* was available in many languages.

Thank you IPWSO for spreading awareness internationally. Well done!

The following information was distributed to medical people at the ESPE conference.
Please email the chairperson if you are interested: chairperson@praderwilli.org.za

Medical Considerations in Prader-Willi Syndrome
Chapter 5, from *The Management of Prader-Wili Syndrome* (3rd Edition) Urs Eiholzer and Phillip D.K. Lee
Growth Hormone and Prader-Willi Syndrome (second edition)
A Reference for families and care providers. Published by the PWSA (USA).
Recommendations for the Diagnosis and Management of Prader-Willi Syndrome
A. P. Goldstone, A. J. Holland, B. P. Hauffa, A. C. Hokken-Koelega, and M. Tauber, on behalf of speakers and contributors at the Second Expert Meeting of the Comprehensive Care of Patients with PWS. J Clin Endocrinol Metab, November 2008, 93(11): 4183-4197
DVD: Food, Behaviour and Beyond
A valuable tool in teaching caregivers, teachers and other professionals to have a better understanding of the person with PWS.

IPWSO 8th INTERNATIONAL CONFERENCE 2013 - 18 - 21 July, 2013.
The conference will be held at the Fitzwilliam College, Cambridge, United Kingdom.
Registration is planned to be open by January 2013.
Contact Jackie Waters at jwaters@pwsa.co.uk in order to receive further details.

You are invited to contribute to People with Strength

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

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

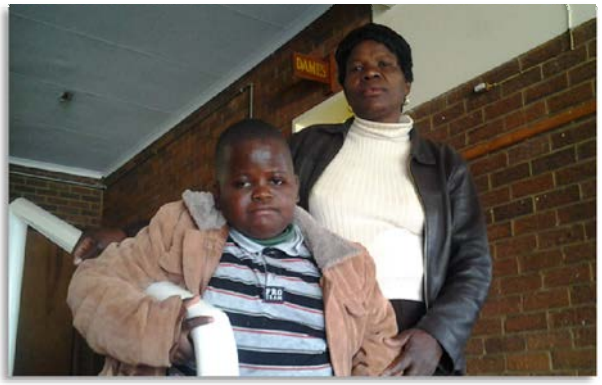
PHOTO GALLERY

The AGM was held on Sunday 19 August 2012 in Pretoria.

Camille Castelyn and Meegan Lategan, students at the University of Pretoria see that the children enjoy themselves

Marise, Gary and Willemien are fond of card games and keep themselves busy

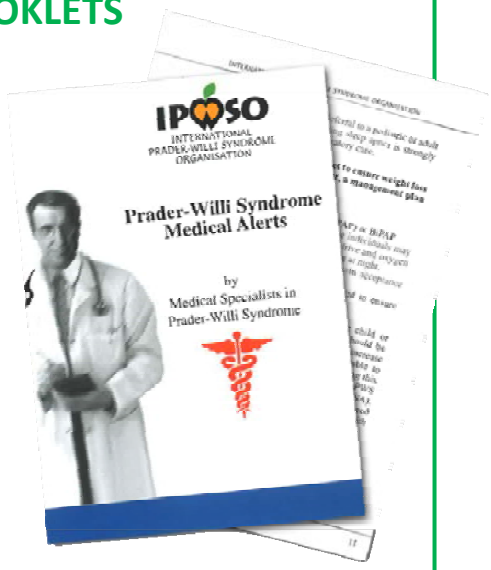
Tea time: Grace Putuka, Gary Edwards and Welhemina Senne



Grace Putuka and Clement attended the AGM for the first time

PLEASE HELP: IPWSO MEDICAL ALERT BOOKLETS

Readers are requested to assist the committee to distribute the IPWSO MEDICAL ALERT booklets to professionals who you come into contact with.
The booklets are specifically printed in A5 format and the contact details of the PWSA (SA) are included as well as the diagnostic testing procedures for PWS in South Africa.



Please inform Janet Legemaate (Vice-chairperson) if you are willing to assist and how many copies you would need. Thank you to those members who already responded to a previous request!
Please contact Janet Legemaate at:
Tel: 031 767 4493 or 082 737 6144 legemaate@absamail.co.za

HUIS HENRI

Feedback by Botha Warnich, chairman of the Henri Warnich Foundation
www.henriwarnichfoundation.co.za

Op die front van die Henri Warnich Stigting gaan dit heel goed en ons is baie opgewonde oor al die projekte wat vir 2013 voorlê! Ons het deur die loop van 2012 meestal gefokus rondom bewusmaking van die Prader-Willi-sindroom en het onderandere 'n praatjie op RSG gehad en is ook deur Huisgenoot genader om 'n artikel te doen oor die sindroom wat in die 18 Oktober 2012 uitgawe geplaas was.vOns hou op 24 November 2012 'n "Wit Olifant" veiling as ons laaste fondsinsamelingsprojek vir 2012. Die veiling gaan in die Strand plaasvind en as daar dalk enige lesers in die nabye omgewing is, wat graag iets wil skenk vir die veiling, sal ons dit met graagte by hulle gaan optel.

Ek wil dan ook net namens die Henri Warnich Stigting van hierdie geleentheid gebruik maak, om vir almal te bedank wat ons so passievol deur die loop van 2012 ondersteun het. Julle hulp en ondersteuning word opreg waardeer.
'n Baie geseënde Kersfees ook aan almal en ons sien baie uit daarna om julle volgende jaar op te hoogte te hou van al ons 2013 projekte!

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Everything is going well at the Henri Warnich Foundation. We are looking forward to all the projects planned for 2013.
During 2012 we had focus primarily on making people aware of the Prader-Willi syndrome. We gave a talk on the radio (RSG) and Huisgenoot/You approached us for an article on PWS which appeared in the issue of 18 October 2012.
Our “White Elephant” auction will take place on 24 November 2012 at the Strand. This will conclude our fund-raising projects for 2012. If there are readers who are nearby and wish to donate something to the auction, we will gladly collect such items at your homes.

On behalf of the Henri Warnich Foundation we wish to thank everybody who supported us during 2012. We appreciate your support. We are looking forward to all our 2013 activities and we will keep you updated.

A merry Christmas to you all.

Regards,
Botha Warnich

THE PRADER-WILLI SYNDROME ASSOCIATION OF SOUTH AFRICA

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

WOULD YOU LIKE TO JOIN THE PWSA (SA)?

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

COST OF MEMBERSHIP

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

BANK DETAILS OF SAVINGS ACCOUNT

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

WE ARE MEMBERS OF

- The International Prader-Willi Syndrome Organisation (IPWSO) www.ipwso.org
- The South African Inherited Disorders Association (SAIDA) www.saida.org.za
- WESTERN CAPE FORUM for Intellectual Disability (WCFID) www.wcfid.co.za
- The South African Association for the Scientific Study of Mental Handicap (SAASSMH) email: saassmh@telkomsa.net

MAAK 'N VERSKIL MET 'N SKENKING!

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

WITH YOUR HELP WE CAN MAKE A DIFFERENCE!

PWSA (SA) is registered as a non-profit organisation (No. 035-837 NPO) as well as a public benefit organisation (PBO Exemption no.930 016 853).
The PBO registration benefits donors and all donations made to PWSA (SA) are exempt from income tax. We will issue an official certificate for donations of R100.00 or more.
You are welcome to make a direct deposit. Please ensure that your surname and cell number are included as reference.

ACKNOWLEDGEMENTS 2012

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

- **Members** for prompt payment of 2012 membership fees
- **afrihost.com** for hosting the Association's website
- **Dawid Basson** the webmaster of the website and also responsible for Facebook. His advice on other matters is sincerely appreciated.
- **Johan and Elmaré Mostert** for the annually preparation of the financial statements
- **Jan Els**, chartered accountant for auditing the income and expense accounts
- **Elsa Volschenk** for her involvement in assisting with the newsletter
- **Karin Clarke and Magdaleen Kloppers** for the birthday cards
- **Dr Engela Honey**, the medical advisor of the Association, who is always available
- **Wilna Basson** for taking care of the Library and educational material
- **IPWSO** for continuously forward information regarding important PWS issues
- **Coral Strydom** for the educational information she brought all the way from the ESPE Conference in Leipzig, Germany.
- **Mike Dovey** of McCarthy Ford and Mazda-Silver Lakes for the generous donation towards PWSA (SA)
- **Keith Kuhn** for his continuous support of the HOME fund under the initiative of Linda Stoffberg
- **THE CO-WORKERS AND ALL THOSE WHO CONTRIBUTE TO *PEOPLE WITH STRENGTH***