

Report on the 7th International PWS Conference 19 – 23 May 2010

By Linda Thornton, National Director PWSA (NZ)

The 7th International Conference in Taiwan is now over, but will not be forgotten. It was attended by five of us from the New Zealand PWS Association: Cindy, Karen, Emma, Melanie, and me. IPWSO paid for my costs while the PWSA NZ paid for the others. I was co-organiser for the General (Parent/Professional) Conference with Janice Forster (USA). The conference was attended by over 240 people (including 26 with PWS), from 33 countries, mainly from Taiwan and Japan, but good contingents from the States, Australia, and good representation from most European countries. We even had a doctor (who has a daughter with PWS) from Cuba – something that took a huge amount of organisation as the Cuban government is reluctant to endorse external travel.

The Conference comprised 1.5 days scientific, 1.5 days caregivers' programme, and 1.5 days general conference. There was also a programme for children and siblings. Despite some initial misgivings, it was well-organised, and held in the Taipei County Hall which was a 10-15 minute walk from the hotel where we stayed. There was also an underground connection, with an added walk. The humidity was around 90-100% and the outside temperature was 29C. It took a little getting used to.

Scientific highlights: The huge strides in genetic discovery over the years were particularly marked. Much of the scientific research reported this year has been carefully built on the first research papers presented back in 1992. Back then research was being discussed on UPD (uni-parental disomy), growth hormone trials had just started, but quite often diagnosis was still largely clinical and "early" diagnosis was around years 1-3, and only then was chromosome testing done. Sleep disorders were just being discussed and there was some suggestion that patients be treated as narcoleptic. Profiles on intelligence, cognitive and behavioural characteristics were starting to appear, but psychosis had not yet been tagged to the UPD genetic subtype. Prozac was the nominated drug for the "reduction of PWS behaviours" but the concept of understanding just why the behaviours were occurring had not come into play. Experiments were being carried out to see whether there was any pharmacological way of controlling the appetite, but were inconclusive. Most patients or groups of people with PWS who were studied, lived in institutions, were largely overweight, and had behavioural challenges.

Scientists have come a long way in their genetic understanding of PWS, the various subtypes and what differences may be seen in each; growth hormone is generally universally accepted as the chief beneficial medication for children (and now adults) with PWS, behavioural characteristics have been divided into those with a genetic deletion or UPD, or Imprinting, and psychotropic drugs to deal with these are now common practice.

Two issues interested me: the first is research being carried out in Australia for a nasal spray of oxytocin to help control behavioural outbursts; the second is a recognition that the hormone, ghrelin. (which is a hormone in the gut that controls our appetite and which was a hot topic some years ago when it was discovered that those with PWS had a larger amount of ghrelin than normal, thus it was hoped by controlling the amount of ghrelin, there would be control over the appetite... but this was not to be) is now purported to act as a 'memory stimulant' which increases a person's desire for food, rather than controlling the appetite. (French research)

A full scientific review is being prepared by Dr Ellie Smith for Australasia. You can read this report by clicking [here](#). There is also a report from Janalee Heinemann (USA) which you can read [here](#).

However, it is in the General Conference and the new additional Caregivers' conference that the most progress has been made.

Caregivers' Conference:

Built on the first two Caregivers' Conferences, this third conference was a presentation of the workings around the Best Practice Guidelines and introduced the audience to the methodology. In presenting this, it was easy to see how much better understood the person with PWS is today, than s/he was some years back. I think the introduction of Human Rights has played an important role in identifying the person separately from the

syndrome so that people with PW are now recognised as individuals and not just put into one PW category where one size has to fit all. There has been a huge change in the management around PWS and people are no longer put on psychotropic drugs just to calm them down and make life easier for the service provider. The whole concept of individual programmes has been accepted (if not wholly implemented) and the discussion around challenging behaviours now looks at the cause, rather than the treatment.

However, it was still obvious that New Zealand does things differently. In some regards this is something to be proud of, inasmuch as we take very seriously the individuality of the person, but in other ways, those with PWS suffer (and fail) because of the insistence of freedom of choice, independent living, and the non-establishment of homes specifically designed for the syndrome.

I am hopeful that our meetings with the Ministry of Health will correct not only this, but also the inter-NASC transfer problem that is now being enforced by the Ministry.

Copies of all the workshops over the two years will be provided by IPWSO at a cost. However, as a facilitator at one of these conferences and having attended all three, I have a copy of these which I am currently putting into a format which is more easily understood and useable. I hope that our workgroup will be able to add to this so that the Best Practice Guidelines becomes a New Zealand model and accepted by the NASC agencies across the board. This will influence service providers to provide across-the-board best practice management of PWS.

General conference:

Karen gave a particularly good talk on why an early diagnosis makes a difference. She introduced (via Powerpoint) her family and Cameron, explained how difficult it was to accept such a diagnosis and what it meant to her family, and then pointed out that having a platform to work from has made all the difference for Cameron. Her talk was clear, cohesive, with a touch of humour, and really set the feeling for all the parents present that although PWS is the most challenging of syndromes, you can be positive and with the right support, successful. We were all immensely proud of her!

The General conference tends to bring the main points from scientific research to a level where parents can take the information and apply it. So that presentations on nutritional needs in PWS, by Peter Davies (Aus) and fitness in PWS by Georgina Loughnan (Aus) were very well received, and in spite of everyone knowing the importance of these two issues, the way they were presented gave us all a good grounding once again. Peter Davies' book on Exercise and Physical Activity in PWS is now available from the office, free of charge to paid up members, and \$8 otherwise. These will also be available at training sessions.

Maintaining good health in PWS is a topic that Susanne Blichfeldt (Denmark) has presented over the years and this is now finely-tuned to include all ages. It is essential health management knowledge specifically for PWS and I am thinking of turning this into a separate publication, with Susanne's permission and co-authorship. Hopefully this can be sponsored by Pfizer.

Puberty, Osteoporosis, and Sex Hormone Therapy: again, this is a topic that has been presented by Kate Steinbeck (Aus) at previous conferences and she does this in a manner which is clear, concise, and readily accepted by parents. This is something that also needs to be included in any publication about good health in PWS. It is now commonly accepted that sex hormone therapy for boys and girls helps with bone density and social acceptance. Patches, injections, subcutaneous slow release are methods used.

Education and Transition into the Community: presented by BJ Goff (USA). BJ and I collaborated on the booklet PWS in the Classroom, but her additional thoughts on transition into the community filled the gap between school and residential care. BJ is doing a large survey on sexuality in PWS and this would also be important in the Health & Wellness booklet.

Food Security: Presented by Janice Forster (USA) took the hard line of there being absolutely no chance or expectation of food for a person with PWS, which takes away the anxiety and determination to food-seek. I think this is a difficult subject to get across to parents, and probably more designed for residential caregivers. But the message is quite clear.

Although there was much reinforcement of the basic management procedures in the General Conference, I was disappointed in the talk by two young behavioural students from Cambridge University, "Predictability and Prevention of Tantrums". I don't think I was alone in this. This topic was introduced at the last IPWSO conference in Cluj and we thought, given there were three years inbetween conferences, there might be some further good research on this topic, but I didn't find this at all. Their work was all done by observation and "set-ups" some of which were videoed. I don't doubt that every parent in the room could have said "I could have told you that". The point that tantrums could be "predicted" if certain circumstances were present, seems obvious to parents and rather unhelpful. What parents need to know is what actually has caused the rise in anxiety and what they can do about lessening the effects, or preventing them. There was little on the prevention of tantrums.

Social activities

We had a wonderful Welcome Dinner, with lots of formality to begin with, with talks from various dignitaries, and formal welcomes to each country and lots of recognition for various individuals. The food was delicious, and the entertainment was lovely with a drumming number by some children with PWS and their siblings (really good), songs from an adult choir (each person had a rare disorder) which was very moving, and a stilt-walker who "walked" and balanced on a tall ladder, doing lots of acrobatics. Under the limiting circumstances of a stage with fairly low roof, he did fantastically well.

On Saturday night we had a most interesting banquet/formal dinner, up in the hills. It took about an hour to drive there by bus after having spent an hour or so wandering around the Lin Family gardens and seeing some early style architecture. The banquet was very formal with many speeches, awards and presentations (I was honoured with one of these). The food was an occasion in itself in true Taiwanese style which included whole chicken (feet, head & beak – delicacies), beautiful whole fish (including eyes, another delicacy), pig cortex (spinal cord) which looked like soft, fat noodles (and tasted similar), cow intestine in broth (fairly tasteless), sea slug (tasted fresh, chewy and slightly salty) and various other dishes. Wine is not a common drink, but a bottle of very rough red was on offer, along with brightly coloured cordial. The banquet was a lot of fun, with much singing from some very special choirs, all with rare disorders. (Cindy and I wondered whether we could get our own PWS Choir together! Particularly as it was so good for the breathing, as well as social contact.)

On Sunday night the IPWSO board was invited to a dinner at a downtown hotel. This was many, many courses, fortunately of small portions, and very delicious. It was a nice wind-down from the conference and time to thank our hosts.

Next International Conference: This will be held in Cambridge, UK, and will be of a very high calibre as far as the scientific conference is concerned as much of the international research on PWS comes from this university. Dates are 11-14 July 2013. Accommodation will be on-site at the university, or at adjacent hotels.

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