

CONFERENCE REVIEW

The 1st Asia Pacific PWS Conference was held in Wellington, 1-2 March 2008, at Te Papa. The challenge for the organisers was to set a programme to meet the intellectual needs of everyone, from parents to professionals, from clinicians to specialists. This was done by inviting three main guest speakers, Professors Merlin Butler (geneticist) from USA, Urs Eiholzer (clinician and GHT specialist) from Switzerland, and Tony Holland (psychiatrist) from UK. Their areas of expertise included: different genetic diagnoses, growth hormone treatment, professional clinical care of those with PW and their families, and the psychiatric care and behavioural management of those with PWS.

From these subjects stemmed all the other topics.

- From genetics came the [role of hormones in appetite](#), the [genetic model of starvation](#), evidence supporting an [autistic spectrum](#), [different diagnoses](#).
- From clinical care came medical awareness of [gastric necrosis](#), [gastric bypass](#), [and early death in PW](#), [sleep disorders](#), [scoliosis](#), [weight management](#), [dietary care](#), [diabetes](#), [exercise and development](#), [sex hormone therapy](#), [growth hormone treatment](#). the concept of a '[five finger model](#)' of overall care through to [educational](#) , the importance of understanding [sleep apnoea](#) and [sleep disorders](#), and [residential](#) needs.
- From psychiatric and behavioural concerns, came issues around [behavioural management](#), understanding the mind-set of people with disabilities, understanding [repetitive question and temper outbursts](#), [ethics](#) of food control, through to the [power of positive attitudes](#).

Having the diversity of Te Papa to explore, gave the conference some versatility and those not wanting to hear some of the topics had the attractions of the museum.

Workshops on day two provided some good interaction between speakers and audience and many parents took advantage of speaking directly to professionals whose brief visit to New Zealand made it all the more important to seize the opportunity.

Thanks to some very generous funding from sponsors, Pfizer (Australia and New Zealand), Ipsen (Australia), Oasis (Australia), Perry Foundation (New Zealand) and a very generous anonymous sponsor, we were also able to offer discounted registration for parents across the Tasman and close to home.

Socially, this event provided plenty of opportunities for families to mix and make new friends, for professionals to network with each other, and for the two groups to join as part of one large 'family' for the weekend.

What did we learn?

- ✓ That the genetic diagnosis of PWS is more complex than first realised; apart from the three main diagnoses of deletion, maternal disomy, and imprinting, there is, in the deletion, two subtypes. Scientists are now studying the differences in

these diagnoses and this will eventually have an impact on how to offer better management for PWS. *(Speaker: Merlin Butler, USA)*

- ✓ That there is genetic evidence to support the theory that PWS can cross into the autistic spectrum. This will have an impact on management and understanding of behaviours and lead to a better comprehension of those with PW. *(Speaker: Merlin Butler, USA)*
- ✓ That behaviour, when looked at carefully, can be understood to be a method of communication, survival, and protection. Learning to understand this 'language' will provide caregivers and parents with a set of unique skills. *(Speaker: John Ford, NZ)*
- ✓ That although people with a maternal disomy diagnosis are more prone to suffering psychotic episodes in later life, there are good medical interventions that will help treat this and keep it under control. *(Speaker: Tony Holland, UK)*
- ✓ That recognising PWS as a 'starvation' syndrome, rather than an eating disorder, will change the way we look at – and manage – the syndrome, providing us with yet a better understanding of why the drive to eat is expressed so strongly. *(Speaker: Tony Holland, UK)*
- ✓ The importance of dietary management will always be first and foremost in any caregiver's directory. It can be managed, but requires extreme vigilance from carers. Hand in hand with this goes exercise and wellness. Exercises and dietary guidelines – including such things as using low GI foods – are a major factor in management. The correct nutrition, learning how to read labels on foods and recognise such things as fat content, were brought home with some excellent visual aids. *(Speakers: Peter Davies, Janet Franklin, Georgina Loughnan, Australia)*
- ✓ What are the ethics of food control? How can we work and behave ethically as caregivers, denying open access to food on the one hand, and providing safety on the other, when the person with PWS is legally old enough to make up their own mind? *(Speaker: Tony Holland)*
- ✓ That it is still an unknown mechanism to hypophagia (the unrepressed desire to eat), but Ghrelin hormone which stimulates appetite, is high in PWS. Insulin inhibits ghrelin secretion – in obesity there is low ghrelin levels but high insulin levels. Perhaps this inhibits the ghrelin level. PYY (pancreatic poly-peptide) hormone, produced in pancreas, in response to food, is known to decrease appetite. Role unclear, but some studies show a dysfunction in PW. Can decrease appetite by 12% but only in females. Is this a future drug target? There seem to be some highly controversial studies done in this area. Dramatic weight loss studies on MIC1 'anorexia' hormone. Researchers think there is much more to be discovered through gut hormone study to solve the obesity problem. *(Speaker: Alex Viardot, Australia)*
- ✓ How do we manage Diabetes in PWS? Type 2 diabetes, adult onset, is more frequently found in PWS when the patient is obese. Type 1 diabetes is not so

common in PWS, but the PW population is just as at risk as the general population. Management of diet is a priority and oral therapy with regular glucose testing and insulin injections is needed, but in an obese patient, this can often be difficult. (*Speaker: Paul Hofman, NZ*)

- ✓ Comprehensive care of PWS: Five major options should be focussed on: (1) weight control and dietary management (2) Growth hormone treatment (3) Daily training programme – exercise and activity to strengthen muscle mass (4) Male sex hormone therapy – helps improve muscle mass in boys (5) Parent and family support. Keeping to a simple management routine is better than going through unnecessary medical examinations for the sake of them. (*Speaker: Urs Eiholzer*)
- ✓ Growth hormone treatment: still by far the best medical treatment for body composition in children and adults with PWS. The growth hormone deficiency in PWS is adjusted with treatment. Body composition is dramatically improved and the lifestyle of those treated takes on a completely new turn. Studies on GH in adults with PWS is still quite new, and although studies are being carried out in USA and Sweden, at first blush, it doesn't seem as though weight loss is a feature, although improvement in muscle tone has been noted. (*Speakers: Urs Eiholzer, Switzerland and Charlotte Höybye, Sweden*)
- ✓ Sleep apnoea is an area for increasing concern and we saw a video clip vividly describing the affects of this. Breathing masks (CPAP machines) are readily available and extremely effective in helping the person breath normally and gain a good night's sleep. (*Speaker: Brendon Yee, Australia*)
- ✓ Residential care has changed substantially over the years from institutions to houses where people living there should feel able to call it 'home' and staff intervention is supportive rather than directive. The challenge is always to do better, we heard how a home in Australia is run. Residential accommodation today can offer one-person supported living, group residential living, with a mix of differences, and PW-only homes. (*Speakers: Angus Capie, NZ, Anna Hughes, Australia*)

Where to from here?

One of the most important things that conferences can do is to encourage researchers to continue seeking for answers, enlighten the medical fraternity as to the most successful interventions, and equip parents with the knowledge and confidence to ask questions and get answers.

Breaking down information from the scientific level to a useful tool that parents, caregivers, teachers and family can use, is the next step. With this in mind, we hope to be able to offer you two-day seminars with speakers you want to hear more from, and in a forum where both speaker and listener can learn from each other. We are looking forward to putting this into action.

Audio CD copies of talks

These are still available. Contact David Hargreaves: totalrecall@xtra.co.nz

And finally, a word from our IPWSO President, Pam Eisen:

“I can’t help but be sad that I am not with you all at this time. You know how much this conference meant to me and how much I wanted to speak at this groundbreaking regional meeting. The knowledge of all the work put into planning the program and getting the top speakers is something I understand and appreciate. I believe your regional conference will become the gold standard for the future as our regions mature.”