

Autistic-like behaviours in PWS

There has been quite a lot of speculation that PWS can be placed somewhere on the autistic-spectrum for some behaviours. Many research papers presented at PWS conferences describe autistic-like behaviours in PWS, for example at the IPWSO Conference (June 2007) a paper was presented describing “The Relationship Between Temper Outbursts, Repetitive Behaviour and a Preference for Predictability in Children with Prader-Willi Syndrome”¹ It was most interesting to learn that “a high percentage of carers (83.7%) reported that *temper outbursts increased following unpredictability, but not necessarily after any of the other common repetitive behaviours.*”

What is it about the repetitive behaviours that might decrease the need for temper outbursts? The answer might lie in the autistic spectrum.

But before we look for that, let's look at another study also presented at the 2007 IPWSO conference which took a scientific look at whether PWS is actually part of the autistic spectrums. “Autistic Symptomatology, PWS and its Derivations for Treatment”². This study looked at the PWS diagnostic approaches that are also found in autism in order to make a therapeutic connection. The results reported problems in PWS which represent three major clusters also found in autism:

1. **Socialisation** (alteration of social development, especially interpersonal development). These range from loneliness to excessive sociability with strangers.
2. **Verbal and non-verbal communication**
3. **Restrictive and repetitive parameters of conduct** (rigid aspects and limited interest)

At the 2008 Asia/Pacific PWS Regional Conference, a paper presented by Merlin Butler described “PWS Evidence Supporting an Autistic Spectrum” and it would appear that there is a genetic connection.

Other studies have looked at the genetic subtypes of PWS to see whether one might be more prone to having autistic-like behaviours than another with a prediction that those with the uni-parental disomy (UPD) diagnosis would have more autistic-like behaviours. “As predicted, UPD cases were reported as exhibiting significantly more autistic symptomatology. They also were born to older mothers. Deletion cases were reportedly more skilled at jigsaw puzzles. The results lend further support to the notion that abnormality in the expression of maternal imprinted 15q11–13 genes may confer a susceptibility to ASD. They also suggest that there may be cognitive differences between the groups in processing visuo-spatial information.”³

¹ Kate A. Woodcock, Chris Oliver and Glyn W. Humphreys, School of Psychology, University of Birmingham, UK.

² L. Rosell-Raga, V. Venegas-Venegas (Valencian Assoc PWS, Spain), F. Mulas-Delgado (University Hospital “La Fe” of Valencia, Valencian Institute of Pediatric Neurology, INVANEP), M. Peiro (Infantile Endocrinology Service of the University Hospital “La Fe” of Valencia

³ Published in European Child & Adolescent Psychiatry, Volume 13, Number 1, February 2004, pp. 42-50(9)
 Authors: Veltman, Marijcke¹; Thompson, Russell¹; Roberts, Sian²; Thomas, N.2; Whittington, Joyce¹; Bolton, Patrick¹ ¹: Developmental Psychiatry Section, University of Cambridge, Douglas House, 18b Trumpington Road, Cambridge, CB2 2AH, UK, ²: Wessex Regional Genetics Laboratory, Salisbury, UK,

From my own personal observations throughout New Zealand, I have seen children with the imprinting diagnosis to show more autistic-like behaviours than the other two subtypes. I have also seen ADHD being presented in the imprinting subtype.

Autism 101 website <http://www.laurel.mapcl.org/Autism101.html> lists Prader-Willi Syndrome as one of a large group of disorders associated with autism. But, it's all very well having research to tell you that your child has autistic behaviours, but *what does that mean?* Is there a real difference? Having watched, listened, read, and heard from parents about behaviours that seem different from the usual PWS ones, I became interested to know exactly what is behind the person in the research.

I attended a day-long seminar run by Tony Attwood, in Wellington. He is a specialist in this field and has a close connection in his family with his sister-in-law having Asperger's syndrome. During this seminar I was quietly surprised by the number of issues that were raised with which I could clearly associate PW. Then there was the reasoning behind the behaviours which, to me, was more important than the research. Being able to understand behaviour as a form of communication is essential to any management plan that might be needed.

I also began to understand why repetitive behaviours might play a part in the lower number of temper outbursts.

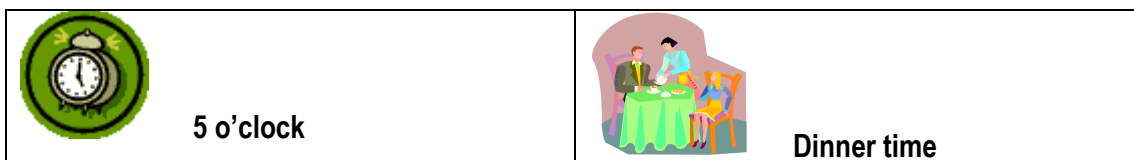
It was of interest to me to find the places where I recognised PWS characteristics also in the autism spectrum. Looking again at the three headings above of socialisation, verbal and non-verbal communication, and repetitive behaviours, one might surmise:

Socialisation

- People with autism are kind, sensitive people, but they may not read signals or know how to respond. *This is also the same as people with PWS*
- Often a person with autism becomes anxious and depressed, has low self-esteem and becomes angry because of this. *Often associated with PWS*
- People with autism are sensitive to sensory things like sounds, smells, touch. Quite often they cannot stand to wear certain clothing against their skin. *Often noted in some people with PWS*
- People with autism can not tolerate loud noises. They actually *hurt* the person. High pitched continuous noises such as hair dryers, or small motors are annoying. Fire alarms, sirens, loud shrill voices, there are many noises that can not be tolerated. *Could this be a reason for sleeplessness at night? Could there be a noise-intolerance level that we are not picking up in PWS?*
- Because of their sensitivity to touch, people with autism do not like to be cuddled, petted, tickled. However, many of them do like to be held closely (not necessarily by another person). Dr Temple Grandin (one of the world's most celebrated people with autism) recognised the sudden docility of cattle when held in a crush. As a child, she wanted the same feeling, so built her own "squeeze machine" which held her tight, safe, and secure. She could control the amount of pressure and release when necessary. This is now recognised as a form of treatment in autism – "sensory

integration therapy” – the deep pressure (rather than a cuddle or pat) allows comforting relaxation. Could swaddling a baby when going to sleep be part of the same feeling of relaxed safety and comfort? Could it help our children with PW whose sleeping patterns are disrupted?

- Sensitivity to taste and texture of food – many people with autism do not like their foods muddled on the plate, can not bear to have more than one taste in their mouths at a time, find the mixture of flavours so obnoxious that they even vomit. *I know that this isn't a common characteristic in PWS, but I have witnessed a person carefully take off all the parsley flakes from a portion of rice on her plate. And I know some with PWS who have an absolute objection to the taste of some foods.*
- Sensitivity to smell will often cause a person with autism to go right off another person because they may have, for example, bad breath, or be wearing a perfume which is just not tolerable to the senses of the autistic person. *I know that many people with PWS are sensitive to smells and can sniff out a fish-and-chip shop at 100 paces, or hear the sound of rustling chocolate paper through deep sleep!*
- People with autism do not like change. They need consistency and routine. If you tell them you are going to the library – that is exactly what it means. It does *not* mean stopping off at grandma's on the way there or back. It's not what you said. Do not automatically expect a person with autism, or with PWS to be able to cope with change or surprise. Surprises can lead to panic attacks.
- Using pictures to tell the sequence of an activity is the best way you can help a child with autism. It's also a way that works in PWS.



This is a way of reducing anxiety, introduces a routine that can be relied upon and teaches sequencing

- Children with autism find it difficult to share their toys. By sharing, they lose control. As it is important for the child to retain control, the concept of sharing does not make sense. *A sense of control is important to any child with a disability. Making sense of their own world (control) is the only way they can start to make sense of the real world*
- People with autism can be seen as being self-centered. *This is essentially recognised in PWS as well*

Verbal and non-verbal communication

- Behaviour is a form of communication. This includes all behaviours whether we regard them as socially acceptable or not. Body language is an important signal for communication and in autism this may take the shape of “fingering”, or “stimming”

(stimulatory behaviour, for example, spinning a lid, flicking a twig, flicking pages).
Many children with PWS will 'stim', or appear hypnotized by repetitive movement.

- Quite often these sorts of behaviours can mean that the person doing them is anxious about something, or may be anticipating an unwelcome surprise or change in routine, it may be that the person is not coping with the noise or things surrounding him/her and so turns to "stimming" in order to put a form of control back into their surroundings.
- A person with autism interprets language in the form of pictures – almost like a video playing over in their mind. It is difficult to grasp the concept of words (squiggles on paper) but easy to grasp a visual image. *Recognised in PWS as well.*
- Repetitive speech in autism is called "echolalia". However, to a person with autism, this can be a normal communication. However annoying it might be to another person to hear someone repeat the same phrase over and over, the repetition to an autistic child is a reinforcement of the whole concept of the word. For example "Mummy" comes to mean "Mummy" because of the whole of the experience, that is, the smell of Mummy, the house where Mummy lives, the shape of Mummy, the sound of Mummy, etc. Most children begin their language acquisition by using this gestalt form but quickly change over to an analytic form. Children begin to analyze the way language is used in other contexts and come to understand that "Mummy" is a word that can represent other things besides the whole of their experience with their specific "Mummy."⁴
Repetitive questioning as PWS communication is very common. Usually we treat it as a difficulty to comprehend the message. We generally answer the question and tell the child to repeat our answer in the hope this will stop the questioning. More often than not, it doesn't work and the repetitive questioning goes on and on.

Repetitive Behaviour

- Being aware of a repetitive behaviour that might lead into further agitation will give the caregiver a 'heads up' or signal. It might also give the person with autism, *or PWS*, a good warning signal as well.
- People with autism like order, predictability and perspective. Many of them love to watch model trains, and when they're older, become train-spotters. Getting down to ground level to watch a model train go round and round on tracks allows the child to become part of the ordered predictability. They are almost mesmerized by this and seem to be in another world. *Although a person with PWS may not become fixated by this repetitive movement, they do like order, predictability and perspective. Some of them are completely happy in an environment of rules and regularity. Some are very tidy – and some aren't!*
- At the 2007 International PWS Conference a paper was given on behaviour where it was suggested that a child with PW might become so fully absorbed in the task at hand (in this case it was a jig-saw puzzle) that they appear to be "in the flow" or "at one" with the object. This was to be encouraged as a time of well-being and wholeness, and could be used as a reward. *Being "in the flow" might also account for the serenity of the autistic child when stimming (self-stimulating) or absorbed in their*

⁴ <http://www.autism-help.org/communication-echolalia-autism.htm>

'special subject'. A person with PWS can also become absorbed in their 'special subject', appear more serene and 'in the flow'.

- Taking away the stimming object (which may be seen as inappropriate) will always be replaced by something else of their choice. Be careful in taking away something considered inappropriate as it might be replaced by something totally inappropriate.
- When a person becomes terrified of a world full of scary sensory experiences, they become almost shell-shocked and need a coping or escape mechanism. They can become mesmerized by a repetitive action or sensation and will do this repetitive action over and over in order to allay their fear and anxiety around what will be their next bad experience. Often the person will spin, or flap hands. This is a clear signal for sensory overload. *This is similar to the experience of a person with PWS whose world becomes out of kilter, a sense of loss of control may erupt with challenging physical behaviours.*
- People with autism like neatness and order. Some people (not all) with PWS show similar need for order and can tell immediately if something has been disturbed, replaced, or is no longer there. For a person with autism this can mean more than just "something's gone", it can mean if a detail is changed, the *whole picture* becomes different. It is *no longer the same*. For a 'normal' person this can be difficult to understand, and words of platitude will not help. *I have witnessed this many times in PWS. If a stitch is out of place, the garment is ruined, useless; if a toy has a blemish, it is wasted, imperfect,*
- Obsessive compulsive disorder: *this seems to be the main area where people with PWS gain the label of having autistic-like behaviours. What does it mean?* In autism, obsessive compulsive behaviours are a means of reducing anxiety; the behaviours themselves become almost irresistible in their repetitiveness and therefore an obsession. Often these obsessive repetitive behaviours are done to reduce a negative experience, to allay anxiety or what could go wrong (eg handwashing), or a fear of contamination. In their extreme, behaviours take over from language and the ability to speak reduces.

Having identified behaviours where PWS and autism cross over, the next question is how to manage the behaviour in a way that is win-win. Some of the strategies offered at the autism seminar were:

Strategies for coping

- Strategies for reducing stress levels: identify the stress, teach deep breathing and relaxation. Anxieties are often at their most profound around puberty.
- Strategies to reduce repetitive questioning or echolalia: don't ask too many questions. Make statements, comments, and praise. Questions confuse a person and raise more questions from them. Repetitive questioning can also give a person a sense of satisfaction or reassurance that you have not changed your mind.

- Disappearing into one's own realm by rocking, spinning, or becoming absorbed in their "special interest" helps to give the person comfort. It alters their state of consciousness (in other words, shuts out all the unwanted things). It is possible to confine some behaviours to a time or place that is more acceptable.
- Self-injury is also common in autism *as it can be in PWS*. However, in autism this can also lead to epilepsy and with severe autism this is as common as 1 in 3 people especially during puberty. Self-injury can be caused from depression ("what's wrong with me/I hate myself") or extreme rage when the intensity of emotion is overpowering. *Skin-picking, pulling toenails or fingernails, even head-shaving and piercing with sharp object can be seen as self-injury. (See more on self-injury)*
- Children and adults who are unable to meet a steady gaze because they feel threatened by it, are often attracted to animals who do not hold a gaze either, and are non-judgmental. Sitting alongside a person means they do not have to meet your gaze and they feel more comfortable.
- Coping with the "special interest" or repetitive behaviour:
 - you can offer "controlled access" by limiting the time with a timer, by offering something else to do (something you know they enjoy), by easing anxiety and explaining when they will have a chance to do their 'special interest' again'; offer the special interest as a reward, "just as soon as you've finished your homework/made your bed/picked up your things, you can continue with your jig-saw"
- Music therapy is proving very successful in children with autism especially when learning to speak. Singing, rather than saying, is a much easier way to learn to vocalize. It's a good way to 'pattern' behaviour and to learn how to sequence. For example, you can have your very own "getting dressed" song which teaches the child the correct sequencing. Songs for other special activities; just starting to sing a line of the familiar song will capture the child's attention. This is also a technique successfully used by teachers.
 - Choice of music for personal down-time. The choice here is interesting – many choose heavy rock or heavy metal music as a release for anger.
- Anger: *the similarities with PWS here are obvious:*
 - Two-thirds of adolescents with Asperger's syndrome have a secondary mood disorder, anxiety, depression, anger.
 - Many suffer bullying and teasing.
 - Many can not understand the emotions.
 - Many show social anxiety about making mistakes and often become controlling as a defense mechanism.
 - Many get exhausted at school from socializing as well as learning and once at home, a different personality arises.
 - Anger is often a 'quick fix' to release energy. Often expelled by breaking something. *I have heard of a young man with PWS who will break glass and once into breaking glass, the repetitive action for him is a release for his anger.*
 - Often the anger is due to a feeling of not being valued (although this in itself is not often able to be vocalized or explained).
 - Anger can be symptomatic of depression.

- Lunar cycle: the full moon can be a time for problems – *I have heard this mentioned often in PWS*. Keeping a diary over the months will indicate whether the lunar cycle is indicative of behavioural disturbances
- Keeping a visual thermometer of anger and sitting alongside the person, you can ask the child whereabouts on the thermometer they see themselves. They can then start to identify emotions of anger from low-level to high.
- Family history of mood disorders. *This is interesting and something that is being explored by researchers in the PWS field*. As far as the autistic spectrum goes, there is a propensity for a child to inherit strong emotions from his/her parents, but is unable to conventionally express these.
 - A survey of 238 children and adolescents showed 64% had more problems with anger; management than typical children, 58% had rapid mood changes more than typical children.
- .Using the web to find resources⁵

In conclusion:

We need to look further at the behaviours in PWS that we might associate with autistic-like behaviours and understand the differences. This may help with better management and understanding of the ways in which people with PWS see their world, see the real world, and try to understand it for themselves.

Understanding behaviour as a form of communication means we have to better understand the behaviours and what they mean.

⁵ www.cat-kit.com/en special education and communication skill tools resource
<http://www.jkp.com> resource for publications on autism, special education, child psychology
www.transporters.tv resource for younger children, teaching emotional interaction and recognition