Evaluating Children with Disruptive Disorders and Intellectual Disability

David Dossetor

David Dossetor is Head of the Department of Psychological Medicine, senior staff specialist and clinical senior lecturer. He trained in the UK and has a special interest in the psychiatry of intellectual disability. He has also presented and published in a number of other areas of interest including psychopharmacology and health related quality of life. He has been involved in the CAPTOS project since its inception. David provides telepsychiatry services and regular visits to Macquarie Area Health Service which includes Dubbo, Mudgee and Wellington.

Disruptive Behaviour Disorders in Those with Intellectual Disability. The Influence of Behavioural Phenotypes on our Understanding of "Challenging Behaviour".

Ronald in a happy mood (Photograph by kind permission of his parents)

Ronald is 8 years old and has had irremediable chronic disrupted and disrupting sleep for many years despite the best endeavours of his parents, a Sleep Investigative Unit and behavioural programmers. On average he sleeps 2 hours a night. He has gross and fine motor coordination problems, can't dress himself, do zips or buttons but is expert at stripping off his clothes. He has been clean and dry for 2 years, but only by day. He needs help wiping his bottom. At night he soils and then smears his faeces round the room. He can express and understand simple single sentences. Yet he has a capacity to read aloud the daily newspaper (without comprehending what he is saying) but he can't recall what his mother asked him to get from the next room. He is constantly on the go. He has no pretend play or social interaction with other young people whom he, at best, ignores. He hits out at his sister, and, if not individually supervised at school, other children. He will do this with minimal

provocation such as when children look at him or smile, something about which he has an obsession. His interests are all repetitive and stereotypic. He loves tipping things over. He will recite or re-enact scenes from a favourite video. He will fill a computer screen with a single digit for amusement. He requires constant individual adult attention. If his constant need for guidance is not heeded, he starts to build himself up into a state of intense agitation during which he repeatedly talks to himself, with increasing tension which often goes on to self-injury, biting himself, or scratching himself until he bleeds. He can also self injure if someone accidentally touches him, scratching that point till bleeding. He may also push or hit adults. He intrudes into people's personal and social space, which makes them vulnerable to unexpected attack.

He has recently had his diagnosis of Smith Magenis syndrome confirmed by genetic probe, which demonstrated a deletion of chromosome 17. At the autistic school his diagnosis was made by his teachers 5 years earlier because he had the same flat face with prominent forehead and small nose, with stumpy hands and feet, as a girl with the same diagnosis and the same terrifying behaviour. Their developmental paediatrician has prescribed Dexamphetamine for 3 years, which helped his language progress, but not his behaviour. A trial of Sertraline had no effect, and recent treatment with Risperidone moderated his sleep problem and gave him significant weight gain but didn't affect his behaviour.

His mother and stepfather are both good-natured people with unusually close family support. They have both had to change jobs because the strain and sleeplessness of caring for Ronald made them unpredictably unavailable for work and at times unsafe in their judgement because of chronic exhaustion. They both had features of worry, anxiety and burnout. They have a strong relationship together, but are concerned for the physical and emotional effects on their 5-year-old daughter. Yet they are among the best-informed people on the details of Smith Magenis Syndrome in Australia. Mother has presented papers at international conferences. Research as well as clinical experience confirms that Ronald's behaviour is completely characteristic.

This is what a behavioural phenotype is - a predictable pattern of behaviour associated with a known genetic problem.

This child illustrates the issues involved in the assessment and treatment of disruptive disorders in those with intellectual disability.

1. Families don't notice how they teach aggression:

The dominant model used to understand disruptive behaviour disorder is the social learning model, that is the inadvertent tendency of parents to reinforce anger and violence, through: -

1. **Coercive cycles of negative interaction:** the child increases the emotional tempo until the parent gives in- as parents we experience these as angry, frustrating battles to get the child to do what we want them to do. Children, especially intellectually impaired children, experience these as overwhelming, negative, critical and sometimes rejecting experiences. They do not increase the likelihood of the child complying. Observers are likely to observe us as parents in a power battle in which anger is displayed to get obedience but authority is not used in a consistent way.
2. **Inadequate supervision and rewards for bad times** - here we may be weary from the demands of our children and fail to give them anything until "something goes wrong". We fail to structure positive activities, we only relate with negative events and so we unthinkingly reward negative events.
3. **Lack of warmth, encouragement and rewarding interactions** - the difficulty of a child who is different, demanding and not doing what we hoped may make it hard to like our children sometimes even though we love them. We may become less loving than our child needs even though it is the best we can do at the time.

All of these messages of the social learning model imply that it is the social environment of

family, school and community that cause problem behaviour. However, sometimes in those with intellectual disability the situation is more complicated. Parents find themselves struggling with the best will in the world to do the right thing by their child. The children do not usually intend to cause harm. Other underlying medical factors may be in play. Anger explodes as an unrefined display of negative feeling that swamps everyone.

'All of these messages of the social learning model imply that it is the social environment of family, school and community that cause problem behaviour. However, sometimes in those with intellectual disability the situation is more complicated.'

2. Being difficult is a positive message:
The implication of "challenging behaviour" as the term for such behaviour may be difficult to accept. That is that the behaviour has a communicative intent, a message from the child, which needs understanding and responding to even if at times it is hard to read. This message can be decoded by a sort of scientific experiment in which the parent joins the scientific team. Everyone has to throw away their previous ideas as to what is causing the behaviour and observe carefully what is actually happening. The technical name for this is functional analysis. Parents and clinician analyse the behaviours and what things function to trigger, what things come out of the behaviour and ways to get the same positive results less painfully The most frequent functional needs are the demand for care, attention, stimulation or other needs such as food and fun. Sometimes the avoidance of stressful demands (such as homework) or social contact (for those who find this stressful) may figure in the analysis.

'The most frequent functional needs are the demand for care, attention, stimulation or other needs such as food and fun.'

3. Behaviour patterns are more related to developmental rather than chronological age:
Behaviour patterns have to be considered in relation to children's developmental profiles. The behaviour described at the beginning of this article behaviour is from an eight year old, but is better understood as from a young person whose self help and communication skills are functionally like that of a 2.5-3 year old. His social development is even more impaired than his communication skills and is like that of a 1-1.5 year old. Any message from the child in abnormal behaviour (communicative intent) has to be considered in this context, rather than like that of an eight-year-old's desire for independence of parental supervision. My own research in a sample of 92 adolescents with intellectual disability, showed that the level of disturbance was developmental age, not chronological age related. Behaviour was worse at a developmental age of between 18 months and 3 years, in keeping with the greater demandingness of average toddlers, before they develop an imaginative internal world and capacity for empathy and social reciprocity.

'the level of disturbance was developmental age, not chronological age related.'

4. Understanding developmental syndromes can help:
Diagnostic developmental and behavioural syndromes may help understand the risk of the disruptive behaviour. Ronald, described earlier, fits the description of Hyperkinetic Disorder or Attention Deficit Disorder (ADHD) and also Autism. Associated with ADHD are the drive of restlessness, the inattention to external signals, the low threshold to disturbance and the high amplitude of emotionality, negativity and distressed feelings. The central feature of autism highlighted by research and reliable assessment is the lack of give and take (reciprocity) in communication, social interaction and the way stereotypic activities or interests are pursued. An alternative description is a lack of flexibility and adaptability to the (social) environment. This explains why the environment needs to adapt to the person with autism, rather than the other way round. Recent research has shown that the strongest predictor of self-injury in an intellectually disabled young person is autistic features. (Oliver, C., personal communication). Further long-term behavioural interventions have been disappointing, in larger studies. Current attempts to reduce self-injury in young

people with intellectual disability have moved to prevention initiatives in high-risk groups.

'Recent research has shown that the strongest predictor of self-injury in an intellectually disabled young person is autistic features'

5. Problems with mainstream psychiatric diagnoses:
Although some of the diagnostic labels mentioned above appear to have predictive validity in those with intellectual disability, this is not clear for widely accepted labels such as depression, generalised anxiety disorder, oppositional defiant disorder or post traumatic stress disorder. What people feel and think inside cannot be reliably elicited in adults with an IQ under 50 (or under a developmental age of 4.5 years), and so diagnoses that depend on the expression of subjective experience cannot be made reliably. Accordingly, our academic understanding based on average populations cannot always help us when we are looking at someone from a different population or individuals with intellectual disability. There have been some attempts to establish different diagnostic categories based on descriptions and on statistical analysis of populations of disabled people. This has been an interesting approach but not widely accepted, as they seem to bear little resemblance to the categories in adult populations. Many psychiatric diagnoses are difficult to elicit reliably in those with an IQ <50 or a developmental age under 4.5 years.

6. Abnormalities of brain biology influence mind and behaviour:
It is known that of over 90% of those with an IQ less than 50 have abnormalities of brain structure.

Until recently, with a few exceptions we could not define any connection between specific anatomical abnormalities and behaviour. The new developments in medical sciences, such as genetics, molecular biology including neurotransmitter studies, functional neuroimaging, and neuropsychology give us new insights into the biological underpinnings of the mind and its problems. There are recognised genes that increase our risk for ADHD. Abnormalities of frontal lobe function have predictable behavioural deficits. Occasionally, neurotransmitter levels in the CSF can inform logical prescribing. However, all too often, we have no such investigations to help us and any brain abnormality can only be considered to be a non-specific risk factor for challenging behaviour.

'It is known that of over 90% of those with an IQ less than 50 have abnormalities of brain structure.'

7. Behavioural phenotypes are a window on the influence of the brain on behaviour.
Behavioural phenotypes, where a particular genetic problem or disorder is associated with a specific behavioural pattern, is a rich source of investigation to establish different underlying causes for outward behaviour patterns. New theories for disruptive behaviour can be explored. Frequently, some of the steps to understanding the links are missing between genetic abnormality and the different behaviour problems. However each condition enables us to look again at the way the brain develops, the connections between different parts of the brain, the chemistry of the brain and the way the working brain is linked to thinking, feeling and acting.

The greater acceptance of the physical and biological fabric underlying behaviour has led to a broader clinical approach to assessments and treatments than traditionally seen by child mental health professionals. We have to take on board the fact that these children and young people with intellectual disability have greater dependency needs and their delays in the developmental sequences of growth give rise to both psychological and brain problems (neuropsychiatric abnormality).

Conclusion
As our understanding of disruptive behaviour grows, the pendulum swings between nature and nurture, between seeing the problem as arising from the way children are made physically on the one hand, or on the other hand, seeing the problem as arising from the way children are raised by their parents. But in those children and young people with intellectual disability, and more particularly

those who show a behavioural phenotype, the evidence suggests that nature needs to be taken more seriously when trying to make a difference in behaviour problems. The behaviours are often more extreme (and sometimes dangerous), pervasive and unaltered by environmental changes. This has also lead to the importance of examining the role of psychotropic medications. Each administration of a drug can be seen as a test of the physical fabric that may contribute to disturbances in the child's life. If the practice of medicine is like sailing the oceans with medical texts as maps, then the psychological medicine of those with intellectual disability is sailing with more maps, even if the accuracy of each map is not known. The greater dependence on carers and families make developmentally sensitive, emotionally encouraging individual and environmental communication important. However even more than usual, each young person with intellectual disability should be considered individually and from a greater number of perspectives to ensure a greater number of helpful options. When we do this we acknowledge greater uncertainty in our assessments and treatments. In practice this means that treatment trials are often helpful contributions to assessment. The more traditional idea that we begin by gathering all that we must know is replaced by the more realistic approach of acknowledging that assessment and treatment go hand in hand.

Treatment in the case example involved:-

- *advocating for funding and additional substitute care*
- *treating the parents for their anxiety and burnout*
- *ensuring the home could provide safe containment*
- *re-examining the quality of communication and behavioural management*
- *recommending melatonin for improving sleep regulation*
- *reviewing his intellectual assessment with a view to suggesting what was going wrong in his brain function*
- *seeking further neuroimaging to understand this better and considering neurotransmitter studies*
- *trialing mood stabilisers (carbamazepine and lithium carbonate) for aggression*

While some of these studies are not available to the coal face clinician most of the things that made a difference are. The extra studies just remind us in case we had forgotten that every one of has to obey the laws of gravity and chemistry as well as the family and social rules that govern behaviour.

REFERENCES:

Harris, James, C. 1995. *Developmental Neuropsychiatry Vols. I and II*; Oxford University Press

O'Brien G, Yule W. 1995. *Behavioural phenotypes.* San Diego: MacKeith Press.

Smith A, Dykens E, Greenberg F. 1998. Sleep disturbance in Smith-Magenis syndrome (del 17 p11.2). *American Journal of Medical Genetics* (Neuropsychiatric Genetics) 81:186-191.

www.omim

Smith Magenis Syndrome: first described in 1986. Following features found in 75-100%:

Genetics: interstitial deletion of chromosome 17p11.2 (diagnosed by fluorescence in situ hybridisation, FISH), may include up to 100 genes deleted (contiguous gene syndrome); sporadic occurrence, incidence estimated at 1/25,000 population.

Appearance: Short (growth retardation), flat midface, and head; abnormalities of nose (broad bridge), ears, palate and eyes (eg. strabismus), short fingers and toes, hoarse voice.

Neurology: poor muscle tone, peripheral neuropathy, decreased deep tendon reflexes, decreased pain sensitivity, hearing loss.

Sleep disturbance: naps during day, difficulty getting to sleep, waking in night and early; absence of REM sleep, inverted melatonin circadian cycle.

Development: Intellectual disability (IQ 20-78), structural brain abnormalities, delay of motor and speech development, expressive language worse than receptive.

Behaviour: Often described as a "good baby". Characteristic self-hugging. Destructive and self injurious behaviour, head banging, wrist biting, pulling out finger and toe nails, putting objects in orifices eg in ear, hyperactivity, impulsiveness, hostility, aggression.

Other less common problems: heart and kidney problems, low active thyroid gland, scoliosis, high cholesterol levels.