



Factsheet 17

Behaviour in CHARGE

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While not every child with CHARGE develops significant behavioural challenges, it is fairly common (Hartshorne and Cypher, 2004). Because unusual behaviour is often associated with genetic syndromes (Harris, 2006), it is not surprising to find this with CHARGE. The problem has been to understand it and develop interventions to help parents, teachers, and caregivers to address the behaviours.

Fundamentally, it is important to consider behaviour as communication (Hartshorne *et al.* 2005). Particularly, in a syndrome such as CHARGE, where communication is often problematic, interpreting behaviour as meaningful and useful for understanding the experience of the child can be enormously important.

Behavioural phenotype

Hartshorne (2011) introduced a behavioural phenotype for CHARGE. A behavioural phenotype describes the behavioural features of a genetic syndrome so that 'the behaviour suggests the diagnosis' (Harris, 2006, p. 190).

Hartshorne (2011) proposed a CHARGE behavioural phenotype:

- low normal cognitive functioning
- very goal directed and persistent
- a sense of humor
- socially interested but immature
- repetitive behaviours which increase under stress
- high degree of sensation seeking
- under conditions of stress and sensory overload, finding it difficult to self-regulate and easily lose behavioural control
- difficulty with shifting attention and transitioning to new activities
- easily lost in own thoughts.



While describing the behaviour is useful, understanding its source is important for intervention. I have come to consider three primary sources: pain, sensory issues, and self-regulation.

Pain

Pain is likely to be ubiquitous in children and adults with developmental disabilities (Oberlander and Symons, 2006). Untreated, pain can significantly impact on behaviour, relationships and emotional attachment, adaptive functioning, educational experiences, and can produce anxiety and depression (Stratton, 2012).

There are many potential sources of pain for children with CHARGE – from regular medical procedures to chronic ear and sinus infections, gastrointestinal problems of many sorts, and the byproducts of accoutrements such as trachs, gastrostomies, and

implants. Pain can influence mood, sleep, attention, and interaction.

Because individuals with CHARGE may respond to pain differently, there is often a presumption of a high pain threshold. This 'pain insensitivity hypothesis' has been challenged by a number of researchers (Bottos and Chambers, 2006). It may be more useful to consider a high pain tolerance as opposed to threshold. Individuals with CHARGE are likely to have to get used to a great deal of pain.

Sensory Issues

Individuals with CHARGE are multi-sensory impaired (Brown, 2005). That is, besides sight and sound, they often experience deficits in sense of smell, have vestibular and proprioceptive deficiencies, and may be tactile defensive.

Interpreting or making sense of experience and other people is challenging when your senses are challenged, and cognitive development, communication, and relationships may be impaired. This can lead to stress and anxiety. Responses to anxiety can include obsessive compulsive behaviours, rigidity, withdrawal, and frequent outbursts (Ranzon, 2001).

David Brown (2005) notes that some of the behaviours observed in CHARGE are creative adaptations to the sensory limitations. For example, imagine that you had no balance sense, and so you experienced the world somewhat off balance. You might prefer lying flat as opposed to being upright, and you might resist and complain against attempts to get you up. Being up is unsteady and tiring. A lack of balance might interfere with the development of body language and motor coordination, making some forms of communication difficult and frustrating (Brown, 2005).

Self-regulation

Self-regulation is related to goal directed self-control, except that regulation may involve both inhibiting and promoting, and both suppressing and encouraging action on the part of the individual (Vohs and Baumeister, 2004).

Jude Nicholas and I have proposed four areas of regulatory activity: cognitive, behavioural, affective, and physiological. These areas are not completely independent, but may point to the source of poor self-regulation.

Self-regulation is motivated by goal directed behaviour. Consider, for example, the desire to learn to walk, often delayed with CHARGE. For success, individuals must direct their attention to the task and

stay motivated (cognitive self-regulation). They must control their behaviours so that they are directed toward the activity instead of distractions (behavioural self-regulation).

It is also important to manage emotions like fear and discouragement in order to stay on task (emotion self-regulation). Finally, they use what vision, hearing, balance and proprioception they have in order to not be distracted (physiological self-regulation). Self-regulation skills begin developing at birth as the infant learns to deal with emotions (crying) and comfort (eating, sleeping). When the infant has sensory deficits, along with health issues and pain, self-regulation development is impeded.

A lack of self-regulation skills can mean poor motivation for tasks, behavioural and emotional meltdowns, and a physiological system that is not in sync.

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