

Behavior in CHARGE syndrome

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Unusual behavior is often associated with genetic syndromes, and may constitute a behavioral phenotype. In contrast to providing a psychiatric diagnosis, a behavioral phenotype describes what is unique to the behavior associated with different syndromes. While behaviors in CHARGE are as complex and variable as other aspects of the syndrome, there are some commonalities that raise the question of common sources for these behaviors. This article addresses how pain, sensory issues, and anxiety may impact the behavior of individuals with CHARGE syndrome, and how the development of self-regulation skills might help to mitigate some of the behaviors.

KEYWORDS

anxiety, behavior, CHARGE syndrome, pain, self-regulation, sensory impairment

1 | INTRODUCTION

In 1996, there was a panel on behavior in CHARGE at the Australasian CHARGE conference. The next year at the International CHARGE Conference in the United States, Denno and Bernstein (1997) presented on behavioral characteristics in CHARGE. Concern about and interest in the behavioral aspects of CHARGE have increased since. In 2007, Wachtel, Hartshorne, and Dailor (2007) reported that 37 (43%) of individuals with CHARGE from their sample were on at least one psychotropic medication: 15 were on one, 13 were on two, 8 were on three, and one person was on five psychotropic medications. We suspect the number overall has increased. Behavioral issues are common topics on the various CHARGE syndrome social media pages

and behavioral presentations continue to draw large crowds at international conferences.

Problem behaviors in CHARGE are as complex and variable as other aspects of the syndrome. Physical behaviors may include scratching, hair pulling, biting, pinching, kicking, shoving, throwing objects, smearing feces, undressing, self-injury, and resistance. Verbal behaviors may include repetitive statements or questions, yelling, and complaining. Non-verbal behaviors may include agitation, pacing, invading personal space, and withdrawal. Psychiatric diagnoses are often assigned to individuals with CHARGE, the most common being autism, obsessive-compulsive disorder, and attention-deficit/hyperactivity disorder. But we have also seen individuals diagnosed with Tourette disorder, bipolar disorder, schizophrenia, borderline personality disorder, oppositional defiant disorder, anxiety disorder, and major depressive disorder.

Hartshorne and Cypher (2004) argued that “it may not make sense to add diagnoses on to that of CHARGE, such as CHARGE and autistic disorder, or CHARGE and ADHD, but rather to recognize that a diagnosis of CHARGE may indicate the potential development of behaviors that are like those in other disorders” (p. 50–51). As Waite et al. (2014) point out, “High rates of ASD have been reported in syndromes such as Cornelia de Lange and fragileX. However, there is debate about whether the ASD profile of behaviors that triggers a diagnosis in these syndromes is the same as in individuals with idiopathic ASD” (p. 470). Unusual behavior is often associated with

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genetic syndromes, and may constitute a behavioral phenotype. A true behavioral phenotype would potentially allow for the diagnosis of a syndrome on the basis of behavior that is unique to the syndrome (Harris, 2006). Hartshorne (2011) has described a behavioral phenotype for CHARGE syndrome (Table 1). While these seven characteristics appear to be descriptive of behavior in CHARGE, additional research is needed to compare behavior in CHARGE with behavior in other syndromes (Waite et al., 2014). Nevertheless, they provide an alternative to a psychiatric diagnosis and a foundation for future research.

Challenging behavior in genetic syndromes may be seen as deriving from four sources (Einfeld, 2004). These are vulnerabilities stemming from intellectual disability when present, from the child's life experience, from factors in the immediate environment, and from the specific genetic cause. While we cannot change the genetics, another approach to behavior is to try and understand why it might develop, assuming it has an experiential/environmental etiology. Through our work with individuals with CHARGE and their families we have come to see three sources of behavioral issues in CHARGE beyond what might be genetic. These are the experience of pain, sensory issues, and anxiety. Figure 1 shows these in the form of a triangle. In the middle is self-regulation, as we believe learning to self-regulate pain, sensory systems, and anxiety can help to mitigate the behavioral issues.

This article addresses how pain, sensory issues, and anxiety impact the behavior of individuals with CHARGE syndrome, and also briefly addresses the development of self-regulation.

2 | PAIN

Historically, individuals with developmental disabilities who present with limited communication have been portrayed as having a higher threshold for pain than the general population and some have gone as far as to say these individuals do not feel pain (Sobsey, 2006). Similar statements, without any evidenced based research, have also been proposed for individuals with CHARGE, such as suggesting a high pain threshold. More recent research, however, suggests that individuals with developmental disabilities, including those with CHARGE, experience more frequent and intense pain experiences (Breau, Camfield, McGrath, & Finley, 2003; Stratton & Hartshorne,

TABLE 1 CHARGE syndrome behavioral phenotype

Low normal cognitive functioning
Very goal directed and persistent with sense of humor
Socially interested but immature
Repetitive behaviors, increase under stress
High degree of sensation seeking
Under conditions of stress and sensory overload find it difficult to self-regulate and easily lose behavioral control
Difficulty with shifting attention and moving on to new things; easily lost in own thoughts

From ref. Hartshorne (2011). Used by permission.

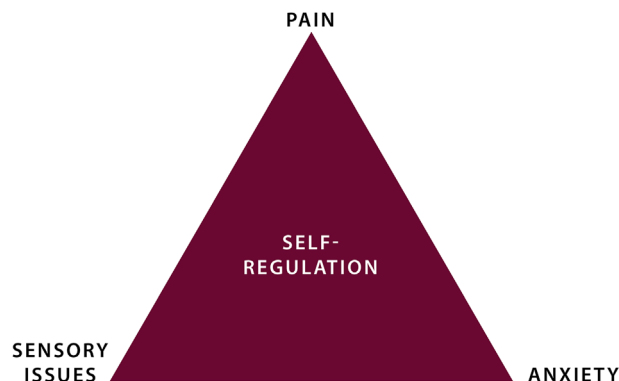


FIGURE 1 Behavior Triangle: Major sources of problem behavior in CHARGE syndrome

2010; Stallard, Williams, Lenton, & Velleman, 2001) and that they may communicate pain in a different way due to communication and sensory deficits (Oberlander & Symons, 2006; Stratton & Hartshorne, 2010).

Individuals with CHARGE endure a significant number of painful experiences including multiple, intensive medical procedures, recovery, acute complications, and various therapies (e.g., physical therapy) to name a few. Stratton and Hartshorne (2010) found that individuals with CHARGE experience considerable amounts of reported intense pain, largely from complications related to CHARGE including migraines, constipation, surgery-related pain, chronic otitis media, sinusitis, gastroesophageal reflux, dental issues, and breathing-related pain. It has also been hypothesized that all 12 cranial nerves can be impacted in CHARGE and the extent of these anomalies' relationship to pain is unknown (Blake, Hartshorne, Laward, Dailor, & Thelin, 2008). Migraine headaches, however, have been implicated in cranial nerve V functioning (Hargreaves, 2007). Blake et al. (2008) report cranial nerve V anomalies in over half of children with CHARGE. Migraines were reported by parents to be the most intense painful experience for children with CHARGE in the Stratton and Hartshorne (2010) investigation.

In addition to substantial and intense acute pain experiences, individuals with CHARGE have chronic pain (Stratton & Hartshorne, 2010). Parents were asked to rate their child's frequency of pain for a variety of identified painful experiences. Of those experiences, gastroesophageal reflux, difficulty swallowing, breathing difficulty, hip/back pain, abdominal migraines, and muscle pain were reported to occur 95 or more days a year, often in combination with one or more other painful experiences. In addition to prolonged conditions eliciting chronic pain, surgery pain is frequent. The reported average number of surgeries was 12 (range of 1–47) between the ages of 1 and 18.

Due to its chronic nature, individuals with CHARGE may develop a high degree of pain tolerance, and so underreport it. Those with communication challenges may not have the tools to indicate the experience of pain. When not reported clearly, underreported, or when communication attempts are missed by others, treatment to reduce pain is not sufficient or even missing. As the gold standard for

treatment of pain remains patient self-report, individuals with CHARGE are placed at greater risk for experiencing long-term, unnecessary pain that can impact their quality of life (Oberlander & Symons, 2006), overall adaptive skills (Breau et al., 2007), mental health (Kassam-Adams, 2006), and behavior.

With pain, problem behaviors increase. For those individuals with CHARGE who have a combination of limited communication and heightened pain experiences, it is expected that problem behaviors will be elevated and at times will be intense. Even with more formal and functional communication in place, pain can impact adaptive functioning including understanding and using language, and socializing (Breau et al., 2007), thereby reducing the self-report of pain. In one investigation, individuals with an intellectual disability were found to have between 21% and 29% reduction in functioning when in pain (Breau et al., 2007). It is unknown to what degree reductions in adaptive behavior and communication occur in individuals with CHARGE when their pain experience intensifies or occurs for a prolonged period. However, it is clear, based on our experience, that these are replaced by problem behavior, an alternative form of communicating one's needs.

As noted, many children with CHARGE develop extremely challenging behavior (Hartshorne, Hefner, & Davenport, 2005; Lauger, Cornelius, & Keedy, 2005; van Dijk & de Kort, 2005). It has been shown that challenging behaviors increase with pain, including aggression and self-injury, and may serve as indicators of pain or alternative ways to communicate pain to others (Cook, Niven, & Downs, 1999; Symons & Danov, 2005). Further, longer periods of time associated with elevated pain have also been found to be associated with elevated ratings of self-injurious behaviors (Symons & Danov, 2005; Symons, Harper, McGarth, Breau, & Bodfish, 2009). Given this, it is imperative for parents, medical professionals, educators and the like to first rule-out pain when problem behaviors have presented, spiked, and/or appear unexplainable. All behavior is communication and challenging behaviors like aggression, property destruction, tantrums, and self-injury, are strongly communicating that there is a problem, and it is often due to pain.

The CHARGE Non-Vocal Pain Assessment (CNVPA) (Stratton & Hartshorne, 2012) is a non-vocal pain measure that can be used to screen for pain based on an individual's presenting behavior. This 30-item measure assesses five subscales including vocal, social, facial, activity/challenging behaviors, and body limbs/physiological and uses a Likert rating scale. The CNVPA is reported to have strong psychometric properties, good social validity, and has been found to discriminate pain well from non-pain behavior for individuals with CHARGE.

In summary, problem behaviors serve a purpose and often in CHARGE are communicating discomfort or pain. Due to multisensory impairments, many individuals with CHARGE present with limited communication that can be further limited by a painful experience. Rather than using the historical model of "They don't feel pain or they need to experience more pain to feel it," more recent research suggests individuals with CHARGE tolerate a significant amount of pain and likely communicate this through problem behavior. Non-vocal pain

assessments can be useful to identify pain early when problem behaviors begin/increase and can lead to earlier treatment of pain/discomfort and a reduction, or elimination in problem behavior. Teaching strategies to indicate and communicate pain at the child's developmental level is strongly suggested, particularly when pain is present. As more formal communication increases, problem behaviors are less likely to develop.

3 | SENSORY ISSUES

As they develop, children learn how to create responses to their sensory experiences (Dunn, 1997). According to Dunn, sensory processing difficulties can affect social, cognitive, and sensorimotor development. They can also impact the child's reactivity to their environment, and this can lead to behavioral issues. Fox, Snow, & Holland (2014) found sensory processing difficulties in well over half of 38 five- to nine-year-old children at risk for conduct disorders.

Sensory impairments and differences in response to sensory stimulation are frequently associated with genetic syndromes, and understanding these and how they influence behavior is important (Waite et al., 2014). Many of the puzzling behaviors demonstrated by children with CHARGE syndrome originate from the complex multisensory impairments that are prevalent in the condition (Davenport & Hefner, 2011). The behaviors begin because they serve important functions for the children, and it is necessary to understand the functions of these behaviors before attempting to modify them (Murdoch, 1997). The brain is only connected to the body and to the outside world through the sensory systems (Coren, Porac, & Ward, 1984) and in CHARGE syndrome all of the sensory systems may be malfunctioning for various reasons. There may be specific problems with the sensory receptors (the eyes, the ears, the vestibular apparatus, etc.), there may be problems with the nerve pathways that connect the sensory receptors to the brain, specifically, frequent impaired function of the cranial nerves (Blake et al., 2008), and there may be malformations of the brain itself (Feng et al., 2017), and all of these will have a significant negative impact on overall sensory functioning. It is not unusual for a child to have all three kinds of these problems, (with sensory receptors, with nerve pathways, and with the brain), which makes functioning (for them), and understanding and interpretation (for us) very difficult.

We rely on intact sensory systems to gather information about our environment. It is important to remember that damage to any one sensory system can present functional challenges to integration with other, apparently unrelated, sensory systems, because information is missing. In this context even minor issues with a sensory system, things which should be easily managed and compensated for by most people, may have a magnified impact on overall sensory functioning and behavior for a child with CHARGE syndrome. For example, vestibular problems may contribute to functional vision deficiencies (Möller, 2011). Clinical assessment might suggest that a child's vision or hearing or balance are fine, particularly if examination of eyes and ears

suggests that everything is anatomically intact. However, close observation of the child is likely to reveal significant problems with function, not least because of the interdependence of the senses (Smith, Smith, & Blake, 2010). As a result it is most helpful to consider children with CHARGE syndrome as multi-sensory impaired, and to utilize well-established educational practices from the broad field of deafblindness.

The sensory systems of children with CHARGE syndrome are unusually vulnerable to deterioration or further damage. A sensory problem may be temporary because of an infection (e.g., of the eye or the ear, joint pain, or a skin infection). But it may also be the case that the sensory system itself has sustained permanent damage as a result of such things as persistent and severe infections over time, unfortunate side effects of surgical intervention, falls resulting in impact to the head, or the child's own self-stimulation behaviors which can develop in intensity and frequency until they become self-abusive (e.g., head banging, teeth grinding, eye poking, skin picking). Any worrying change in behavior (e.g., not looking at things, not responding to sounds, moving less, resisting being touched, or lifted and carried, sleeping too much or not enough, self-stimulating with more intensity or more frequency) should result in an investigation of sensory status.

Significant variability of sensory functioning may also result from internal factors (such as fatigue, hunger, chronic constipation, pain, poor body temperature control, anxiety) and external factors (such as light levels, noise levels, sensory distractions, postural insecurity). Sometimes aggressive behavior towards self or others may be reduced, or eliminated altogether, by measures taken to change these internal and external factors. Pain control, deep pressure massage, removal of excessive environmental stimuli, regular movement, and better physical support in sitting, can all help a child to become less distressed, and more available for social interaction and learning.

An additional consideration is that many of the children have sensory systems that are not integrated together effectively so that they do not respond to stimulation in a well-modulated way (Brown, 2005). Such sensory processing issues are considered to underlie behavioral and functional problems in Autism Spectrum Disorder (Case-Smith, Weaver, & Fristad, 2015). This is likely the same in CHARGE given the challenges of multi-sensory impairment. As a result, apparently minor incidental sensory inputs (air movement or quiet noise from a heater or air conditioner, shadows created by people walking between the child and a window, a wet or sticky patch on the tabletop where the child places their hands) may overload the child and create strong aversive responses, high levels of over arousal, or a significant increase in possibly damaging self-stimulation behavior. Good deafblind educational practice is founded upon a multi-sensory view of the child and their environment, so that all of these internal and external factors should be automatically considered, particularly when a child's behavior is causing concern.

Just like all infants, young children with CHARGE syndrome self-stimulate in order to learn about their bodies, to extend and practice skills, to self-regulate, to amuse themselves, and to make themselves feel more confident and comfortable (Murdoch, 2000). With

multi-sensory impairment the need to self-stimulate becomes more intense and more persistent because the brain is not in good contact with the body. Characteristic self-stimulation behaviors in early childhood include rocking side to side (just the head or the whole body), kicking the legs, sucking the fingers, grinding the teeth, rubbing or scratching the body with the hands, staring at light sources (often while waving a hand in front of the eyes to create shadows and visible movement), and adopting postures which create strong proprioceptive stimulation (crossing the fingers and the legs, arching the back, curling into a fetal position, pressing the head, squeezing into tight spaces). These behaviors can serve many important functions (Moss, 1993), including confirming that the body has postural security, reducing the impact of joint pain, helping to maintain an open airway, reducing discomfort from constipation, stabilizing the visual field, calming down, and gaining and maintaining alertness. In the early days all these behaviors give the child's brain a better idea of where the body is, what it is doing, and what it is capable of doing. As a result these kinds of spontaneous behaviors are inherently self-regulating, and they offer us important insights into the child's challenges and needs, and valuable pointers for our interventions (Haney, Hartshorne, & Nicholas, 2015). But if we do not notice and value self-stimulation behaviors, or even worse, if we consider them nonfunctional or negative and try to eliminate them as our only goal, then this is likely to have a negative impact on the child's self-confidence and sociability, and in turn their behavior. If we believe that children with CHARGE engage in unusual behavior because it is in some way functional for their self-regulation (Ramirez, Hartshorne, & Nicholas, 2014), then attempts to block these behaviors may frustrate and challenge the child to engage in even more serious behaviors. Our goal must be to understand why the child might have chosen these behaviors, and then help them to find alternatives that still provide, among other things, the necessary sensory stimulation.

4 | ANXIETY

Anxiety, anxious behavior, and diagnosed anxiety disorders are common concerns among individuals with CHARGE and their caregivers. A survey of 87 families found that anxiety disorders were the most common mental health diagnoses reported in individuals with CHARGE (Wachtel, Hartshorne, & Dailor, 2007). Blake, Salem-Hartshorne, Daoud, and Gradstein (2005) surveyed caregivers of individuals with CHARGE and 43% indicated that their child was diagnosed with obsessive-compulsive disorder while 37% percent indicated an anxiety disorder diagnosis. Similarly, Hartshorne et al. (2016) found that about 50% of their participants with CHARGE indicated difficulties with anxiety and/or obsessive-compulsive behaviors. Overall, it is clear that anxiety is a prevalent concern in this population.

Anxiety has many different causes. Individuals with CHARGE syndrome are exposed to circumstances that contribute to a higher level of anxiety (Wachtel, 2011) including concern over explained or

unexplained pain, sensory overload or underload, and lack of predictability in the environment. A meta-analysis of children with chronic illnesses found a small but meaningful difference in anxiety compared to levels of anxiety in the general population (Pinquart & Shen, 2011). Specifically, they found that there were higher levels of anxiety in individuals with sensory impairments. The authors noted that lack of control over an illness can lead to elevated levels of anxiety. Anticipation of pain can also lead to higher levels of anxiety in individuals with CHARGE and these higher levels of anxiety can lead to deficits in tolerance of chronic pain (Nicholas, 2011).

Sensory impairment and difficulties gathering and processing information can lead to a lack of understanding of one's environment and the progression of events. This lack of predictability can result in higher levels of anxiety and increases in behavior aimed at communicating distress or obtaining security. Use of interventions that help mark changes in routine and anticipated events can make the environment more predictable (Blaha, 2001), which may help to reduce anxiety.

Anxiety can have a direct positive or negative impact on behavior; moderate amounts can prove useful while too much can be debilitating and too little can be dangerous. Children with CHARGE syndrome exhibit behaviors that are common with anxiety: negative thinking patterns, anger, aggression, tantrums, crying, physical complaints, avoidance behaviors, sleeping difficulties, eating disturbances, and withdrawal from activities or family interactions (Eugster, 2007). Research indicates that while anxiety is a concern for individuals with varying genetic syndromes, there are differences in how that anxiety is exhibited (Crawford, Waite, & Oliver, 2017; Leyfer, Woodruff-Borden, & Mervis, 2009). CHARGE syndrome has a behavioral phenotype (Table 1) (Hartshorne, 2011) that may represent a unique profile of anxious behaviors.

Individuals with CHARGE syndrome often exhibit obsessive-compulsive behaviors and many are diagnosed with Obsessive Compulsive Disorder (OCD) (Blake, Salem-Hartshorne, Daoud, & Gradstein, 2005; Hartshorne, 2011; Hartshorne et al., 2016). These behaviors include repetitive behaviors, sorting objects, linking up objects, checking, and an all-consuming focus on one idea, activity, or item (Wachtel, 2011). Hartshorne and Cypher (2004) asked one hundred parents of children with CHARGE about the behaviors that their child exhibits and about a quarter to a third of participants indicated that their child engaged in behaviors linked to a typical OCD diagnosis, including the need for items to be arranged in a certain order or symmetrically, seeking reassurance on doing or not having done an activity, the need to touch certain parts of the body or to blink the eye, and doing the same things over and over again. There can be an adaptive function to such behaviors. For example, ritual organizing and ordering of items may reduce the anxiety over where things are for someone who is multi-sensory impaired. Repetitive behaviors can provide a sense of regularity to the environment and the individual's experience.

Individuals with CHARGE syndrome sometimes exhibit over-responsivity to sensory input (Hartshorne, 2011). Sensory over-responsivity (SOR) is defined by behavioral responses to sensory stimuli that are atypical or exaggerated. While the cause of SOR in

individuals with CHARGE and how it might be related to sensory impairment are not known, evidence shows that SOR is associated with anxious behavior, and anxiety diagnoses. In a study of college students, individuals with high levels of sensory responsiveness were more likely to be anxious (Levit-Binnun, Szepeswol, Stern-Ellran, & Engel-Yeger, 2014). Conelea, Carter, and Freeman (2014) found 88 children with a clinical diagnosis of anxiety were likely to exhibit behaviors related to SOR. They also found that SOR was significantly correlated to an OCD diagnoses. Similarly, Lewin, Wu, Murphy, and Storch (2014) found that sensory concerns and SOR were linked to OCD-like behaviors. Therefore the heightened levels of OCD behaviors present in the CHARGE population could partially result from their sensory over-responsivity.

Individuals with CHARGE syndrome exhibit behaviors that are common in children with anxiety. Anxiety may be the result of pain, sensory impairment, and the unpredictability of the environment. Interventions should assist with pain management, sensory issues, and making the environment more predictable.

5 | SELF-REGULATION

Self-regulation is the ability to monitor and change one's responses to achieve a desired goal. This process involves overriding the body's automatic responses to achieve an outcome or state. There are two components to self-regulation: 1) having a desired goal; and 2) self-awareness. Together these two components allow individuals to monitor their progression toward a goal and to alter their cognition, emotions, behavior, or physiology to maintain movement toward the goal (Ramirez et al., 2014).

There are four major systems involved in self-regulation: cognition, behavior, emotion, and physiology. The body must coordinate these systems to achieve a goal after receiving input from internal and external sources. Cognitive self-regulation is the ability to regulate one's thoughts and mental processes through attentional regulation, shifting, inhibition, and working memory to achieve a desired goal. Self-regulation of behavior requires an individual to monitor one's behavior to achieve a goal, which may be done by inhibiting activity, regulating movement, or delaying gratification. Emotional self-regulation is the ability to react to situations with an emotional response that is appropriate in timeliness and intensity. Physical self-regulation involves the body's ability to maintain homeostasis after receiving internal and external stimuli. This includes maintaining equilibrium within the somatic, endocrine, and autonomic nervous systems (Ramirez et al., 2014).

Self-regulation develops through the interplay of physiological development, caregiver responsiveness, and the child's interaction with the environment (Ramirez et al., 2014). As all three may be diminished for children with CHARGE, self-regulation strategies must be deliberately taught. Interventions that teach self-regulation skills may help individuals with CHARGE better regulate pain, sensory systems, and anxiety in order to alleviate negative behaviors that may arise from these issues.



FIGURE 2 Upside down: A child with CHARGE hangs upside down in her chair

When individuals experience pain, they often experience maladaptive thought patterns, negative emotions (i.e., depression, irritability), and compulsive and repetitive behaviors (Nicholas, 2011). Improving self-regulation skills may help individuals with CHARGE to become better aware of negative thoughts and to shift attention away from ruminating on feelings of pain. Self-regulation skills may support an individual's ability to identify and express emotions about pain in ways that do not include behavioral outbursts. For example, self-regulation skills can help an individual identify where the pain is occurring and communicate the intensity of the pain with a preferred mode of communication. Additionally, improved self-regulation skills may allow individuals to inhibit repetitive, or compulsive behaviors that may arise when in pain. Interventions like biofeedback, cognitive behavioral strategies, and relaxation skills can teach self-regulation to address pain (Sauer, Burris, & Carlson, 2010).

Individuals with CHARGE syndrome experience sensory issues including difficulties perceiving balance, experiencing touch, and sensing pressure (Brown, 2005). These can result in poor memory, difficulty coordinating hearing and vision, maladaptive thinking, and stereotypic behaviors. A behavior such as hanging upside down, commonly seen in children with CHARGE (Figure 2), may be a form of self-regulation, as it can support upper visual field loss, vestibular challenges, and provide proprioceptive feedback. Self-regulation can help the child to manage shifts from a calm sensory state to hyperarousal (Dunn, 1997). Acquiring additional self-regulation skills can help an individual to better process stimuli, inhibit triggering sensations, and manage body movements. Interventions like physical therapy and

occupational therapy can help improve self-regulation when experiencing sensory issues (see Brown (2005), for other interventions).

Anxiety can result in maladaptive thought patterns and repetitive and compulsive behaviors (Campbell-Sills & Barlow, 2007). Improved self-regulation skills may help an individual inhibit negative thoughts about past or present anxiety provoking situations, and reframe these into more adaptive thought patterns. Additionally, self-regulation may allow an individual to inhibit compulsive or repetitive behaviors that occur because of anxiety, or help individuals to communicate to others the purpose of their adaptive repetitive or compulsive behaviors. Regulating by developing a predictable routine and being prepared for changes may also lower anxiety. Calendar systems are one example of how assistive technology could be used to make the environment more predictable and encourage social-emotional regulation (Blaha, 2001). Interventions like deep breathing, mindfulness, development and availability of a "safe place" and a predictable environment may help individuals with CHARGE regulate their physiology, cognitions, emotions, and behaviors when experiencing anxiety (Kennert, Ramirez, Hartshorne, Deuce, & Nichols, 2015).

6 | CONCLUSION

Challenging behavior is not inevitable in individuals with CHARGE syndrome, but it is highly predictable given problems with pain, sensory issues, and anxiety. Pain should always be checked first, particularly if there has been a change in behavior. Helping children to compensate for multi-sensory impairment is essential. Anxiety can be reduced by increasing routine and predictability. Children need to be supported in the development of self-regulation skills.

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How to cite this article: Hartshorne TS, Stratton KK, Brown D, Madhavan-Brown S, Schmittel MC. Behavior in CHARGE syndrome. *Am J Med Genet Part C Semin Med Genet*. 2017;175C:431–438. <https://doi.org/10.1002/ajmg.c.31588>