

# Self-Regulation in Individuals with **CHARGE** Syndrome

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## FOREWORD

Self-regulation is our capacity to manage our responses to things that happen in our lives. Self-regulation as a deficit in individuals with CHARGE syndrome has been on our minds for a number of years. But it was in October of 2009 that we sat together in Jude's office in Bergen, Norway, and created the four-dimensional model of self-regulation.

This Monograph is based on a series of articles that appeared in DbI Review from January, 2014 to January 2016. We hope that the publication of this Monograph might lead to a wider audience.

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## 1. Introduction

*Maria Ramirez, Timothy S. Hartshorne, Jude Nicholas*

People are always thinking, behaving, feeling, and sensing. However, to effectively pursue our goals, we need to organize or regulate these systems, so that we can move toward our ends. This process includes gaining control over our own thoughts, behaviors, emotions and physiological states, i.e. our ability to self-regulate. We self-regulate whenever we adapt our thoughts, actions, feelings and physiological states to situational requirements as well as to social standards or norms we have internalized.

*Everyone has goals. As I (Hartshorne) write this it is Friday late morning, and my mind is wandering to how I might spend my time over the weekend. There is some painting that might be done, certainly gardening, a few newspapers I want to read because I got behind on them, and I should probably plan some time for relaxation and my family. Yes, I have goals, but actually moving towards meeting any of my goals depends heavily on how I regulate myself. I need to focus my mind on what I want to achieve, organize my behaviors around tasks, manage my emotions so that I can experience satisfaction as I move toward completion, and maintain my physical equilibrium and arousal levels.*

*To use an academic example, suppose a person had an exam to prepare for. Focusing attention and thinking about the material is a cognitive task that requires regulation. Engaging in study behaviors such as sitting at a desk, keeping eyes focused, and staying engaged is a behavior task that requires regulation. Exams often produce feelings such as anxiety or apathy, and managing these emotions so that they do not interfere with focus is an emotion task also needing regulation. Finally, avoiding fatigue, eating well, sleeping, and focusing one's senses are physiological tasks to be regulated.*

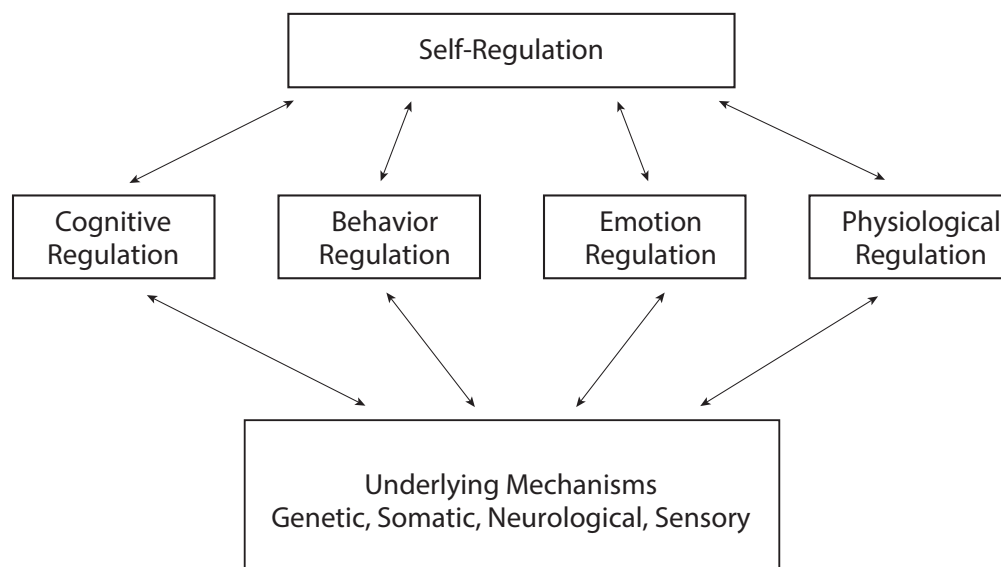
### What is Self-Regulation?

Self-regulation may be defined as the capacity to manage one's own thoughts, actions, feelings and physiological states in adaptive and flexible ways across a range of contexts. Self-regulation involves both the initiation and maintenance of behavioral change in addition to inhibiting undesired behaviors or responding to situational demands.

In order to describe regulation as “self-regulation” two components are necessary: 1) self-awareness of the process and 2) a subsequent goal-directed action. Self-regulation begins with a goal, and that goal describes what you want to have happen and what you must do to make that goal happen. Self-regulation involves gauging internal and external forms of information and responding appropriately under environmental expectations. This feedback-control is important in tracking progress towards a goal (Schmeichel & Baumeister, 2004). Broadly speaking, successful self-regulation entails standards of thought, feeling, or behavior that individuals endorse, mentally represent, and monitor (Hofmann, Schmeichel & Baddeley, 2012).

Self-regulation is composed of multiple components. Nicholas and Hartshorne (2009) have proposed a four-dimensional model of self-regulation: cognition, behavior, emotion, and physiology. This four-dimensional model of self-regulation illustrates that underlying mechanisms such as genetic

predisposition, biology (body & senses), neurobiology (brain networks) and experience interact to determine the individual differences in the ability to self-regulate (see Figure 1).



**Figure 1** The four-dimensional model of self-regulation.

### The Four Dimensions Of Self-Regulation

Self-regulation is the coordination of activity in multiple systems (cognition, behavior, emotion, and physiology) in response to internal as well as external stimulation that can be arrayed along a continuum from effortful to automatic.

*Self-regulation of cognition* is the voluntary regulation of thoughts and mental processes to balance between inhibition and initiation of behavior in order to achieve a goal. The mental processes involved in cognitive self-regulation may include attentional regulation, attention shifting, cognitive flexibility, inhibitory control, updating working memory, metacognition and self-monitoring all of which may affect approaches to learning, problem solving or decision-making.

Attentional regulation is the ability to voluntarily focus attention as needed. Attention shifting is switching a “mental set” while simultaneously ignoring distractions. An example of attention shifting would be counting by different units—tens and ones—and keeping them straight. Cognitive flexibility is similarly involved in avoiding “functional fixedness.” For example, a person looking for a seat may see a wastebasket only as a container that could not also be used as a chair or stool by turning it upside down. People may also show a lack of cognitive flexibility by repeating the same solution strategy even after it has failed. The skill of inhibitory control involves an individual’s ability to override their natural, habitual, or dominant behavioral response to a stimulus in order to implement more adaptive goal-oriented behaviors. Thus, one must focus attention on the task at hand and disregard distracting stimuli. Updating working memory involves maintaining and manipulating relevant information, keeping it in active memory, often while engaging in another cognitively demanding task. Metacognition refers to awareness



of one's own knowledge or thinking processes. Self-monitoring is the ability to both observe and evaluate one's behavior.

*Self-regulation of behavior* is having an awareness of a behavior and choosing those behaviors most adaptive toward achieving a goal. Behavior regulation refers to our ability to use self-control to behave in appropriate ways. Jahromi and Stifter (2008) defined behavior self-regulation as goal directed and purposeful behavioral patterns consisting of one's ability to inhibit activity, regulate pace of movement, and delay gratification. Delayed gratification is the ability to refrain from the temptation for an immediate reward and wait for a later, but larger and more enduring reward (Mischel, 1996). Delay of gratification is important for self-regulation and it is closely related to motivation. For example, Bembenutty and Karabenick (2004) found that students reporting greater delay of gratification were more academically motivated (e.g., had higher self-efficacy and intrinsic motivation).

*Self-regulation of emotion* is the ability to respond to the ongoing demands of experience with the range of emotions in a manner that is socially tolerable and sufficiently flexible to permit spontaneous reactions. When referring to emotional regulation, one is usually referring to the reactivity and regulation of the timing and intensity of emotional responses. Self-regulation of emotion involves the analysis, control, alteration, or prevention of emotional expression and experiences that is adaptive for a situation. This can occur at different times relative to the emotional response. Using emotion regulation, the individual analyzes controls, alters, or prevents behaviors related to the expression of emotions (Lamm & Lewis, 2010). Emotion regulation often is viewed as a social process rather than solely an intraindividual process (Walden & Smith, 1997). For example, the effects of different temperamental characteristics, including dispositional differences in regulation and negative emotionality, on children's socioemotional functioning depend partly on the fit between children's temperament and the social context in which they are embedded (Lerner, 1984).

*Self-regulation of physiology* refers to the ability one has to alter one's own states and responses to meet the needs of the body. This form of regulation is under the control of the somatic, endocrine, and autonomic nervous systems, but is accessible to self-control. An integral aspect of physiological self-regulation is the regulation of sensory systems, including sensory integration, or the ability to process and integrate sensory information (Schaaf & Miller, 2005). According to Kuypers (2011) sensory integration plays a pivotal role in the process and integration of self-regulatory processes. Difficulties in sensory processing may lead to difficulties in social, cognitive, and sensorimotor development (Dunn, 1997). Physiological self-regulation is also involved in the reactivity and regulation of stress response systems (Blair, 2010).

### Relationship Between Self-Regulation And Executive Functions

The four dimensions of self-regulation that we have described may be intricately linked to executive functions. Executive functions refer to the abilities needed to control and regulate organized behavior (Nicholas, 2005; Hartshorne, et al., 2007). They are abilities utilized in monitoring, controlling, and regulating thought and action (Carlson, Davis, & Leach, 2005). This includes goal-directed behavior such as planning, decision-making, self-monitoring (ability to examine one's own behavior), self-perception (using feedback to direct behavior), and ability to make judgments (Nicholas, 2005).

The constructs of self-regulation and executive function exhibit considerable overlap, and both constructs have been characterized as including cognitive processes, emotional responses, and

behavioral impulses (Isquith, Crawford, Espy, & Gioia 2005). If one describes executive functions skills as an umbrella term encompassing the directive roles for purposeful and goal-related behavior, then self-regulation is clearly included. As a result, self-regulation may be described as a key component of executive function skills. For example, Eslinger (1996) defines executive function as including self-regulatory processes such as planning and self-monitoring.

### **How Self-Regulation Develops**

The process of developing self-regulation can be conceptualized as a gradual transition from external control to internal and efficient self-control (Schore, 1994).

Infants begin to regulate arousal and sensorimotor responses even before birth (Florez, 2011). DeGangi (2000) provides a framework for understanding the developmental progression of self-regulation. One of the first steps toward the development of self-regulation is the infant's emerging capacity to attain a level of homeostasis while at the same time exploring his or her environment and regulating arousal and response to stimulation. The development of the mechanisms underlying self-regulation is believed to be the product of the interplay between physiological maturation, caregiver responsiveness, and the child's adaptation to demands posed by the environment (Lyons-Ruth & Zeanah, 1993.; Rothbart & Derryberry, 1981). Other steps that comprise the development of self-regulation include the child's ability 1) to modulate and process sensory experiences, 2) to coordinate simple motor actions, intentionality, reciprocal interactions, and organized effects, and 3) to display self-control, create mental images, pretend play, and use language functionally.

According to Florez (2011) toddlers start inhibiting responses and complying with caregivers at an early age. By age four, children begin to exhibit more complex forms of self-regulation, such as anticipating and modifying appropriate responses. Self-regulation skills develop gradually as children learn strategies to manage incoming information, choose appropriate responses, and maintain levels of arousal that will allow them to actively participate in learning.

To develop self-regulation skills, children need many opportunities to experience and practice with adults and peers (Florez, 2011). Dysregulation is the inability to properly exhibit, inhibit, and alter thoughts, emotions, physiology and behavior (Degangi, 2000). This inability often results in problems with sleep, feeding, stress and sensory processing. These problems may later escalate to challenging behavior, emotional disturbances and deficits in attention and inhibition.

## 2. Self-regulation issues in CHARGE Syndrome

Maria Ramirez, Timothy S. Hartshorne, Jude Nicholas

Self-regulation seems to be compromised in CHARGE. However, different dimensions and several underlying mechanisms come into play in the self-regulation deficits observed in individuals with CHARGE syndrome.

*A few days before a scheduled orthodontic surgery, an 11 year old with CHARGE syndrome was reported to have ‘behavioral issues’ at school. His teachers reported to the parent that he was refusing to do work, was kicking objects, was rude to other children, and was frequently screaming “I am so mad.” All behavior is communication, and so it is worthwhile considering what he might be communicating. In this case it seems likely that he was communicating that he was at least very concerned about the surgery. While it would be tempting to describe this boy as failing to self-regulate, it can be seen that his emotions, behavior, thoughts, and physiological state were all nicely constructed to communicate his attitude toward the upcoming surgery (his goal). First, consider the child’s emotion regulation. Given the upcoming surgery, it is likely that he was scared, anxious, stressed, and possibly confused. He was able to self-identify his emotion as “mad.” The child’s behaviors were refusing to do work, kicking, screaming and not being nice to his classmates. Although not socially appropriate, they nicely served the goal of communicating his concerns. In addition, he may have been having difficulty regulating his cognition (problem-solving other ways of expressing his anger, or reduced ability to monitor the problem-solving process due to poor working memory) and physiologically controlling his internal arousal levels. While we have described these as separate self-regulatory systems, in actuality they impact each other. He may be channeling his anxiety, fear, and anger by engaging in these behaviors, which may be his learned way of regulating emotions such as confusion, fear, anxiety and anger. By increasing his anger, he increases his arousal levels. Once aroused it may be difficult to focus his thinking and so he reacts rather than contemplates. It is important to recognize that this is self-regulation, although deficient, and not wanton misbehavior. But his attempts at self-regulation are not sophisticated and need to develop so that he can more effectively communicate and achieve his goals.*

Hartshorne (2011) proposed a behavioral phenotype for CHARGE which includes: “under conditions of stress and sensory overload, find it difficult to self-regulate and easily lose behavioral control” (p. 323). As DeGangi (2000) states, early deficiencies in self-regulation may lead to challenging behavior, and deficits in attention and inhibition. One source of self-regulatory difficulty is likely the presence of multi-sensory impairments, limiting both the child’s exposure to environmental stimuli and exposure to models interacting and reacting to the environment. This limited exposure may delay or inhibit self-regulatory strategies.

A second source of the challenges with self-regulation is executive dysfunction. Certain problems observed in children with CHARGE syndrome considered related to executive function may result from dysfunction in the regulatory regions of the brain (Lasserre, Vaivre-Douret & Abadie 2013). Hartshorne, Nicholas, Grialou, and Russ (2007) found that children with CHARGE frequently have deficiencies in executive functions, particularly with shifting from task to task, self-monitoring one’s actions, and inhibiting unwanted thoughts, behaviors, or emotions. Thus, individuals with CHARGE syndrome may

have difficulties with using their executive functions for purposes of self-regulation and attaining their goals.

Third, there is emerging literature on self-regulatory strength and depletion (Bauer & Baumeister, 2011). Research indicates that effort expended in maintaining self-control and self-regulation uses up the strength to keep on self-regulating. This suggests that when a child with CHARGE manages to maintain self-control in a stressful situation, they may likely melt down if there is another stressful situation soon after. The physical complications of CHARGE syndrome alone may reduce stamina. Maintaining self-control through self-regulation may prove to be extremely challenging for individuals with CHARGE.

Individuals with CHARGE syndrome experience a constellation of challenges that may be contributing factors to the presence of difficulties across the four dimensions of self-regulation. In understanding their “dysregulation,” it is important to remember that those behaviors we may perceive as difficult or challenging may in fact be a manifestation of their sensory difficulties, executive dysfunction, and fatigue.

Although some behaviors seem “challenging” to us, they may also be an adaptive way for individuals to interact with their environment. In other words, challenging behavior may be self-regulatory behavior, even if the behaviors may not be perceived as socially acceptable.

In the next chapters we will look more closely at the four dimensions of self-regulation. For the sake of simplicity of presentation, the four dimensions are reviewed here separately, although self-regulation in the ongoing daily life behavior of individuals with CHARGE surely involves them all within complex interactive combinations.

### 3. Self-Regulation of Cognition in CHARGE Syndrome

*Benjamin Kennert, Timothy S. Hartshorne, Andrea Wanka, Heather Dix, Jude Nicholas*

The self-regulation of cognition is the voluntary regulation of thoughts and mental processes to balance between inhibition and initiation of behavior in order to achieve a goal. A lack of cognitive self-regulation can result in the individual being unfocused. Too much cognitive regulation can result in obsession.

*Today I (Hartshorne) mean to polish this article. But so far it has been hard to keep my mind focused. When a new email pops up on my computer I am distracted, and if I answer it, I may even forget what I had been doing. I think about an appointment I have later in the day and my eyes leave the manuscript. I read a passage, and then wonder if that is really what we meant to say, and then my mind is no longer on the writing. To counter these forays away from the task, I re-focus my attention, re-energize my motivation, and keep telling myself to stay working and avoid the distractions.*

Cognitive self-regulation allows one to compare alternative choices, stay motivated when thinking about a problem, focus on precision and accuracy, and adapt prior learning to the current problem. It involves planning, modifying, and monitoring thoughts as they occur. In order to plan one must create a mental set of steps and remember the list while performing each step (Luna, Padmanabhan, & O'Hearn, 2010). Planning requires that a goal has been set. Planning can be described as: "The process of formulating an abstract sequence of operations intended for achieving some goal" (Scholnick & Friedmann, 1987). The representation of this sequence is called a plan. A plan can both have an external and an internal representation.

Modifying thoughts is important in order to devote one's attention to the task at hand (Magar, Philips, & Hosie, 2008; Turner & Hussman, 2008; Luna, et al., 2010). A person cannot effectively regulate cognitively without appropriate attention and focus on the goal. Monitoring is also important. It requires feedback, and involves thinking about thinking in order to stay on track.

Another important process involved in cognitive self-regulation is working memory. Working memory is responsible for the allocation of attentional resources during problem-solving monitoring. It is also responsible for cognitive processing that is involved in a range of regulatory functions including the retrieval of information from long-term memory. Working memory keeps us updated on what's happening, and keeps us focused on what matters. Reduced ability to monitor the problem-solving process may result in procedural errors and incorrect associations being formed in long-term memory.

Cognitive self-regulation is difficult to study, as it can only be measured indirectly, but it is important to study. Without cognitive regulation it would be difficult to accomplish even simple tasks such as brushing one's teeth, which involves setting a goal, planning, and focusing attention. Cognitive self-regulation helps children to persist in challenging activities, which increases their opportunities to practice skills required for an activity (Florez, 2011).

Each of the three other domains of self-regulation (physiological, emotional, and behavioral) is related to cognitive regulation. Without the regulation of thoughts and mental processes people would make nonsensical behavioral choices and would have difficulty understanding and controlling emotions.

Jahromi and Stifter (2008) found that a child's competence in each domain of self-regulation was related to performance in other domains. Children with greater cognitive regulation had greater behavioral control and fewer negative and aggressive behaviors related to emotional regulation (Jahromi & Stifter, 2008). Such findings show promise that, by improving cognitive self-regulation, other domains of self-regulation may also improve.

### **Self-Regulation Of Cognition In CHARGE**

Children with CHARGE Syndrome seem to know what they want and persist in their intentions (Hartshorne, 2011). This persistence creates difficulties for parents, for when these children have an idea of what they want, they can have a lot of difficulty letting go of that idea (Hartshorne, 2011). Another challenge is that children with CHARGE often need extended time to process information (Brown, 2005). The impairments associated with CHARGE also may limit a child's awareness of internal and external stimuli, thus limiting regulatory feedback.

Ford, McDougall, and Evans (2009) suggest that an absence of incidental memory may lead to an absence of cognitive self-regulatory skills. The mind works in a systematic way to organize experiences by relating the outcomes of experiences and situations that have previously occurred. If there are problems with executive functioning of the brain, these organizational processes may not occur.

The dimensions of self-regulation are intricately linked to executive functions. Hartshorne, Nicholas, Grialou, and Russ (2007), using a sample of 98 children, investigated executive dysfunction among individuals with CHARGE Syndrome using the Behavior Rating Inventory of Executive Function (Gioia, Isquith, Guy, & Kenworthy, 2000). The study confirmed the presence of executive dysfunction in over half of children with CHARGE. These children displayed difficulty with items measuring shifting from one activity or focus to another, tracking their own behavior and its effect on others, and controlling their impulses and terminating behaviors as required (Hartshorne et al., 2007). Children with CHARGE have difficulty organizing thoughts. This limits the ability to use the knowledge they should have obtained from past experiences. Children with CHARGE may overlook the potential negative consequences of their actions, and may have difficulty weighing the costs and benefits of actions (Magar, et al., 2008). If children cannot organize thoughts in a meaningful way, they may also struggle to remember strategies that have previously helped under a similar circumstance (Jahromi & Stifter, 2008).

### **Interventions To Help With Self-Regulation Of Cognition**

Research is needed to determine successful interventions for improving cognitive self-regulation among individuals with CHARGE syndrome. The following portion of this article will describe ideas for intervention in this area.

Using scaffolding procedures to teach a new task or to achieve a goal may be useful for individuals with CHARGE. Scaffolding describes the process of simplifying a task so that a child can perform it with support. Scaffolding helps the child to be able to identify the problem, learn a complex task, and control their frustrations when they do not complete the task the first time (Florez, 2011). Scaffolding involves providing enough support so that the child is able to achieve a goal or complete a task. The support is gradually reduced as the child becomes more independent (Stone, 1998). Scaffolding helps individuals use what they already know to complete tasks and learn things that they do not yet know. Scaffolding



techniques include: thinking out loud, breaking the task into smaller parts, cooperative learning to promote teamwork, questioning, concrete prompts, and coaching. It is important when using scaffolding with an individual with CHARGE to monitor their stress and frustration levels so outbursts do not occur.

According to Florez (2011) the most powerful way teachers can help children learn self-regulation is by modeling and scaffolding it during ordinary activities or tasks. This cognitive scaffolding is a dynamic process, with the individual with CHARGE interacting with the teacher, who attempts to understand from the individual's responses what cognitive strategies are needed, and accordingly provides guided support to foster cognitive self-regulation. In this way, the individual with CHARGE obtains new strategies or raises new understandings by building on their prior knowledge through scaffolding delivered by the teacher.

For example, cognitive scaffolding may involve helping the child to break down larger goals, tasks, or problems into shorter, more discrete tasks, and then teaching and modeling the task step-by-step. Starting with small tasks and concentrating on one thing allows the child to experience success, which may increase motivation. Having a system that lets the child know what they have done, and what the next step would be, and then getting the child to recognize when they are ready for the next step can be motivating and less demanding for the child. It would be important to recognize when a child is struggling to remain well-regulated and needs a more familiar, less demanding task to maintain control.

It is also important to scaffold metacognitive strategies to support the planning, monitoring and evaluating skills of individuals with CHARGE. Fogarty (1994) suggests that metacognition is a process that spans three distinct phases, and that, to be successful thinkers, students must do the following: (1) Develop a plan before approaching a learning task (2) Monitor their understanding; use "fix-up" strategies when meaning breaks down (3) Evaluate their thinking after completing the task.

Diamond and Lee (2011) suggest several interventions that may help develop executive functions, which could improve cognitive self-regulation. Some of these interventions include using a computer-based program to improve attention and working memory, aerobic exercises or mindfulness activities such as Tai Chi, Tae Kwon Do or yoga, and teaching different thinking strategies.

There are also several specific interventions or strategies for overcoming the limitations of working memory. Many of these working memory strategies have been derived from the current understanding of those cognitive processes involved in the information-processing system. These strategies can be used to minimize working memory failures and enhance instant retrieval of stored information in long-term memory, such as strategies like rehearsal strategies or cognitive load reducing strategies.

For example, rehearsal strategy, as the term implies, consists of using a cognitive strategy that keeps or maintains information in working memory. Rehearsal involves continuously repeating the to-be-remembered material that typically includes rote repetition, either out loud or covertly. For instance, the repetition of information in the same order in which it was presented to keep it available for later recall (e.g., repeating a list of words, such as "dog, tree, fork; dog, tree, fork" over and over; repeating a telephone number over and over again until it is dialed). On the other hand, a cognitive load reducing strategy consists of providing cues to the learner about how to select and organize the relevant information during complex or demanding learning situations, such as an "errorless learning" strategy. "Errorless learning" strategy refers to a learning strategy that decreases or eliminates the opportunity for incorrect choice selection, therefore maximizing the possibility of a correct response. This means

that learning under conditions where errors are prevented (“errorless learning”) compared to those conditions where the participant learns by trial and error (“errorful learning”), may subsequently reduce working memory overload.

Brown (2005) suggests that trying to reduce stress levels, and trying to give the children acceptable strategies for doing so themselves, is one of the most precious gifts we can offer them. Anything that makes learning easier and reduces stress may be helpful. An example may be balancing between new and familiar activities, used flexibly. New activities may be more demanding and increase stress levels, while familiar activities may do the opposite. Modifying distracting surroundings such as noise, light, and people may also be useful for cognitive self-regulation. Providing breaks, letting the individual know that others need breaks, and making use of concrete aids are also examples of possible interventions to reduce stress.

Creating situations in which the child is able to concentrate on the cognitive task, without using their energy for other, different tasks, could improve cognitive self-regulation. Self-regulation, like many other cognitive faculties, is subject to fatigue. It could be useful to prepare the child for what is going to come by having them think about the task ahead and the goal. Allowing movement before, during, and after concentration phases may lower stress and increase motivation. Jarvela, Jarvenoja, and Malmberg (2012) found that motivation is linked closely with active self-regulation among elementary school students. If possible, creating a motivating situation for the child could help greatly. A lack of motivation means that there is no emotional connection with success and then no drive for it (Turner & Hussman, 2008).

### **Self-Regulation Of Cognition In CHARGE: Case Example**

Matthew, almost six, who has CHARGE syndrome, is regarded as highly intellectually impaired, and is thought to be unable to refer to past experiences or separate himself from concrete objects or activities to consider something else, especially at school. One day, when lying on the floor with his mother, she asked him to drink from a blue bowl. He refused at first, then gestured to look at the bowl and carefully examined it as though it was somehow familiar and interesting. Suddenly, Matthew seemed to have a goal in mind. He asked for help standing up, and once erect looked carefully at the bowl. He was now motivated about something, and motivation is a key component of cognitive self-regulation because it involves thinking about what one wants to do and how one is going to do it. As he stood up, it was clear that Matthew needed confirmation of something with the bowl. He touched the water, and after putting it in his mouth began to make a clicking sound with his tongue to help him experience the water in his mouth. He seemed to be recalling a past event and what the experience felt and looked like.

Matthew then looked in a certain direction and pointed to something, knowing his mother would follow his look. He tried to show his mother the direction to go in, which turned out to be toward a large blue, plastic pail. His experience with the small bowl of water seems to have helped him to formulate a thought of wanting to go to the pail. Several days earlier he and his brother had splashed together in the pail. Now Matthew makes a sitting motion, showing that he wants to be in the pail again.

From the situation described, it is clear that Matthew has to work hard with all aspects of self-regulation. However, it is also clear that he has a strong ability to set a goal, sustain his attention on that goal, and show others how to help him achieve that goal. It is also clear that he is able to use his working memory to mentally represent his surroundings. Matthew uses his experience, vision, and touch to help him plan and to help his mother understand what he is thinking. Seeing the small bowl with water reminded him



of the large pail and he became highly motivated toward a goal. Now he needed to regulate his memory to keep focused, and maintain his attention on the goal. Because this has not been viewed as something he is skilled at or does very often, it undoubtedly took a great deal of effort and focus on his part. From this case, we can see the importance of cognitive self-regulation, as it is necessary for problem solving in each individual's own unique way. Matthew was able to

- Use his working memory to maintain a focus on his goal
- Initiate action based on his goal
- Inhibit distracting thoughts or stimulation to get to the goal
- Engage in planning around his goal
- Keep in mind what has occurred in past and integrate information in order to achieve a cohesive understanding of the present
- Problem solve how to best communicate his wants

Interventions can be planned around building on these skills; helping Matthew to make them more sophisticated. In particular cognitive scaffolding procedures might be adopted by choosing simple goals related to Matthew's interests and what he enjoys, and helping him to think about these activities, perhaps through pictures, story books, role play and modeling, and then helping him to problem solve methods for requesting access to these activities.



## 4. Self-Regulation of Behavior in CHARGE Syndrome

*Sarah D. Haney, Timothy S. Hartshorne, and Jude Nicholas*

Self-regulation enables us to set goals, choose from alternatives, control impulses, and regulate behavior. Self-regulation of behavior is our ability to use self-control to behave in appropriate ways.

*Flossing teeth is not an easy habit to acquire. At least it was not for me (Hartshorne). After having to endure a bit of dental surgery due to the state of my gums, I had a new goal in mind: never repeat that. At first I hated flossing. I had to force myself every evening before bed. But my goal was strong and so I stuck with it. Gradually it became something I did pretty much automatically. Even on those late evenings when I just wanted to fall into bed, I found I could not succumb until those teeth were flossed. This is now a very well-regulated behavior. My dentist is very pleased with me.*

Self-regulation of behavior is critical to development as it supports individual success socially, academically, and in everyday life. Self-regulation of behavior means having an awareness of a behavior and choosing those behaviors most adaptive toward achieving a goal. However, what the child views as adaptive and what others in their environment would identify might be quite different. For example, a tantrum might be a great way to get what you want. The tantruming might begin out of frustration, but as goals are achieved, it can become automatic. The main goal of behavior self-regulation is to practice intentional control over behavior and eventually move to automatic regulation (Florez, 2011), as in the case of flossing teeth. In typically developing children, tantruming fades away as their goals become more complex. It is now not simply to get what you want, but to also do so in a way that does not get you in trouble too. Ideally, as the child develops, self-regulation of behavior comes to involve inhibiting useless behaviors, regulating the pace of behavior, and delaying gratification when necessary (Jahromi & Stifter, 2008).

### Self-Regulation Of Behavior In CHARGE Syndrome

Individuals with CHARGE syndrome sometimes have an extremely hard time with self-control, seeming to lack the ability to appropriately regulate their behavior. For example, they may become fixated on an object or thought and not be able to shift away from it, or they may not be able to inhibit an action such as throwing a toy or hitting a friend. Behavioral outbursts get in the way of an individual with CHARGE being able to perform successfully at school, form relationships, express communication, and function independently in everyday living (Lauger, Cornelius, & Keedy, 2005; Smith, et al., 2005). Treatment with psychotropic medications has become widespread, despite concerns regarding effectiveness and the lack of treatment protocols for children and for CHARGE (Wachtel, Hartshorne, & Dailor, 2007).

Hartshorne (2001) has proposed the existence of a behavioral threshold in individuals with CHARGE. There can be a very rapid change from passive to uncontrolled behavior. Hartshorne suggests levels of sensory stimulation, anxiety/stress, or pain may be responsible. However, the behavior itself can be usefully viewed as the child's attempt to self-regulate their behavior through the pain, sensory issues, and/or anxiety. For example, Hartshorne once observed a high school girl with CHARGE on an exercise machine in the gym at her school. She seemed to be doing well, but then suddenly a threshold was crossed and she had a melt down and acted out aggressively. Her behavior abruptly shifted from passive

and cooperative to uncontrolled. How is this self-regulation? The environment in the gym was very busy. There was loud music playing on central speakers, other students were talking loudly and traffic in front of the girl was heavy with students walking in, out, and around the gym. Her goal was to continue working out, but it was harder and harder to maintain her behavioral control. Her sensory system went into overload. Consequently, her attempts to regulate her behavior began to break down. The melt down and outburst was the last option for her, and it led to her rapid removal from that environment.

Children with CHARGE may sometimes feel hopeless and at the mercy of a chaotic environment (Janssen, Risken-Walraven, & Van Dijk, 2003). Sensory impairments and medical conditions can make it frustratingly hard to manage many aspects of their lives, and this can lead to challenging behaviors that are difficult to regulate. According to Nicholas (2005), the inability to self-regulate behaviors in individuals with CHARGE can be attributed to lack of communication skills, sensory/multi-sensory impairment or impairments in executive functioning.

Executive function deficits have been found to be associated with behavior problems (Clark, Prior, & Kinsella, 2002). Hartshorne, Nicholas, Grialou, and Russ (2007) found that executive function can be impaired in CHARGE. Common behavioral problems that arise from impaired executive functioning include: lack of impulse and inhibitory control, difficulty with mental and behavioral shifts, impaired judgment, and decreased self-perception (Nicholas, 2005), and also repetitive behaviors (Jansen, Risken-Walraven, & Van Dijk, 2003). Hence, problems with waiting one's turn to talk, difficulties in anticipation, and the impulsivity of individuals with CHARGE may suggest impaired executive functioning.

Sensory issues may result in many individuals with CHARGE lacking the ability to manage their threshold of arousal. They often do not know how to react to certain stimuli in their environment which consequently causes them to act out. They can often become stressed without the resources to know how to control their emotions and behaviors and calm themselves. Additionally, sensory impairment has been found to limit children's receptive and expressive communication (Lewis & Lowther, 2001), and can be linked to aggressive, self-destructive actions, and other behavior problems (Van Dijk & de Kort, 2005). Common behaviors exhibited from sensory impairments include: hitting, punching, screaming, kicking, and repetitive behaviors such as checking, counting, ordering, pacing, and hand flapping (Brown, 2005). According to Smith, Press, Koenig, and Kinnealey (2005), these types of behaviors "interfere with an individual's ability to communicate, learn and interact adaptively with the environment and are incompatible with the establishment of new skills" (p. 419). The challenging behaviors such as tantrums, outbursts, and self-harming actions are often used because the individual does not know a different way to behave in response to a stressful situation, or is unable to communicate their needs (Van Dijk & de Kort, 2005; Smith, Smith, & Blake, 2010).

Thus challenging behaviors may be adaptive responses to the situation and help the individual respond and function as effectively as they can (Brown, 2005; Hartshorne, Hefner, & Davenport, 2005). Some of these behaviors may serve as self-stimulation for the individual and can be calming (Smith, et al., 2005). Frequently the repetitive behaviors are exhibited as a tool for the individual with CHARGE to calm down and process what is happening around them (Lauger, Cornelius, & Keedy, 2005). They can help the individual to cope with their surroundings and deal with changes in their environment.

## Interventions To Help With Self-Regulation Of Behavior

We self-regulate ourselves in order to achieve a goal. If I want to make a friend, I have to use good social behavior skills. If I want to eat a meal, I have to engage in cooking behaviors. If I want to write an article, I have to actually write. All day long we have goals, some of which we achieve, and others we do not. Children with CHARGE have just as many goals. A goal may be to relax, or to attend to something, or to get something they want, or to get someone to leave them alone. Not all goals are within the child's awareness, but the behavior is still related to the goal. With the goal comes the behavior to get there. Challenging behaviors should be viewed in this context because recognizing the purpose of the behavior is the key to intervention.

A behavior management plan should be based on an understanding of the goals or purpose of the behavior. Care must be taken to evaluate and interpret the child's behavior to have a better understanding of why and when the child engages in these behaviors (Van Dijk & de Kort, 2005; Smith, et al., 2010). The context of the situation is very important. Does the child engage in certain behaviors only in particular social situations or environments? Are there anxiety-provoking stimuli that influence the child to act out in certain ways? Does the behavior seem intentional or out of the child's control? Does the behavior reflect the child's internal emotional state? Is the behavior an attempt to communicate wants or needs? It is also important to note what the child gets from performing the behavior or what the child is attempting to avoid by acting out certain behaviors (Bernstein & Denno, 2005). This can sometimes be identified by looking at what happens after the behavior. Creating an ABC data chart (Antecedence, Behavior, Consequence) is one way to map out behaviors in order to evaluate them. For each behavior, what happened before the behavior (the context) and what occurred right after, are recorded. Such a chart when reviewed often reveals the purpose of the behavior and can be very useful when developing an intervention.

Simply having a goal does not by itself cause people to self-regulate their behavior (think New Year's resolutions). They must really value the attainment of the goal. This has two important implications. The first is that in order to teach a child to self-regulate their behavior we have to convince them about the importance of the goal. Study hard so you can pass the test might be a more meaningful goal for the parent than for the child. This is why parents often have to offer incentives to make engaging in the self-regulatory behavior necessary to do well on the test more desirable (I will give you a dollar if you get an A; you will be grounded for a week if you do not pass). The point here is that if you want to teach a child to self-regulate their behavior, the child has to value the outcome or goal.

A second implication of the role of goals is that the challenging behavior children engage in is generally their attempt to self-regulate around a goal they have. For example, a child engages in tantruming behavior because they do not have access to a toy that is out of reach. A second example would be a child who is experiencing gas pains frantically walks around their house, room to room, throwing objects that they encounter. They may engage in this behavior because it is only thing they have learned that helps them cope with the pain. These two examples of challenging behavior are both forms of self-regulation that allow the child to work towards their goal of either access to the toy or distraction from the pain. Of course there are better choices for behaviors that might lead to the same result, but we cannot teach these if we do not recognize that the challenging behavior has a purpose.

Thus, we want to teach the child to self-regulate their behavior in order to achieve the parent's (or teacher's) goal and we want to teach the child different self-regulation behaviors for achieving their own goals. The former requires making the goal important to the child, and the latter requires that we recognize the nature of the goal. In both cases, we have to specifically teach the child to self-regulate their behaviors in an appropriate and successful manner. It is generally more difficult to convince a child to change their goal than it is to teach them new behaviors to achieve it. However, many "inappropriate" goals may be subsets of more appropriate goals. For example, disruptive behavior to get attention might be a subset of wanting to feel important and valued. If this is the case, the individual can be taught appropriate social skills so that they become more valued in the social setting.

One way to teach new behaviors is through scaffolding. Teaching what kinds of behaviors are appropriate in a variety of settings is very important. Techniques such as modeling, rehearsing, and redirecting negative behaviors to positive ones have been shown to be useful and successful (Florez, 2011; Bernstein & Denno, 2005; Smith et al. 2010). Modeling is demonstrating for the individual how to regulate specific behaviors so they can later do it independently across situations (Florez, 2011). Rehearsing involves discussing appropriate behaviors in situations beforehand. Before going to the store, discuss the behaviors that are acceptable, as well as those that are not. Focusing on positive behaviors is most effective in helping children stop their destructive behaviors (Bernstein & Denno, 2005; Smith et al., 2010). Instead of saying "no", direct the child to alternative behaviors and habits so the person with CHARGE can exhibit them when feeling overwhelmed, frustrated, and over-stimulated. For example, teach the individual to squeeze a soft ball instead of hitting themselves or others when they are feeling upset and stressed.

Teaching choice-making skills and providing opportunities to use choice-making skills could also help reduce negative behaviors. Due to limited communication skills, many individuals with CHARGE may lack opportunities to express preferences and make choices. It is important to recognize that choices are available and encourage individuals with CHARGE to express their preferences related to the available choices.

Parents, caregivers, and educators need to try as best they can to understand the behavioral cues the individual with CHARGE is expressing (Janssen, Risken-Walraven, & Van Dijk, 2003; Lewis & Lowther, 2001). It is important to let the child know that you understand what they are communicating. Some of these behaviors may be the only way the child knows how to convey their wants, needs, and goals (Brown, 2005). Individuals with CHARGE can become upset by change and unfamiliar situations. They may feel overwhelmed, over-stimulated, and at the mercy of their environment (Janssen, Risken-Walraven, & Van Dijk, 2003). Therefore, it is essential to create everyday routines to make the child's life as predictable as possible. Having consistent routines reduces anxiety, as well as the intensity and frequency of challenging behaviors (Smith, et al., 2010). Preparing and planning has been found to be a vital part of self-regulation (Florez, 2011). Use of a calendar system, picture board, hand gestures, and lots of verbal and visual reminders, including social stories to continually prepare the individual for a particular trip or event is extremely helpful (Van Dijk & de Kort, 2005). Scheduling sensory breaks can reduce the negative consequences from over-stimulation, under-stimulation, and stress from anxiety-provoking stimuli in the everyday environment (Smith et al., 2005).

It is important to avoid becoming discouraged when intervening with behavior challenges. Creating a personalized intervention plan and remaining consistent is important, as is giving it time to work. Of course, the best way to reduce challenging behaviors is to reduce the stress levels of those with CHARGE.

Since it can be very difficult to regulate their behavior independently, they need patient, supportive caregivers to aid them in understanding their behavior so they can learn to cope with the anxiety-provoking stimuli in their everyday lives.

### **Self-Regulation Of Behavior In CHARGE Syndrome: Case Example**

James' parents could no longer take him to the store due to his destructive behavior and meltdowns. If his mother turned her back, James might be pulling objects off of shelves and throwing them. When they passed the balloon counter, James would stand there or tantrum until his mother purchased all of the balloons. It was clear that James knew what he was doing. His behavior served to get him what he wanted, particularly balloons. He had a goal, and had chosen behaviors that were generally effective in reaching it.

Trying to change the goal of a young man with CHARGE is no easy task. A different approach is to change the behaviors that led to the goal. James was provided a list of three things that needed to be purchased at the store. To begin, they were items that James liked, such as yogurt. He was told that once these were in the cart he could have a balloon. His parents had never before seen James move through the store so quickly. A similar strategy was adopted at school where James liked to be on the computer and did not like to complete his work. His work was placed into folders that required no more than 10 minutes to complete. Once completed, he was given a token. Once he had acquired three tokens he could be on the computer. James concentrated very hard on his work.

To maintain or strengthen these behaviors, you would gradually increase the challenge. His list of items to get at the store and the number of tokens needed to be on the computer could be gradually increased. But what is important is that James learned there was a way to achieve his goal through a different set of behaviors.





## 5. Self-Regulation of Emotion in CHARGE Syndrome

*Benjamin Kennert, Maria Ramirez, Timothy S. Hartshorne, Gail Deuce, Jude Nicholas*

The self-regulation of emotion is the ability to respond to the ongoing demands of experience with the range of emotions in a manner that is socially tolerable and sufficiently flexible to permit spontaneous reactions. It also addresses the identification and expression of feelings.

*I (Hartshorne) am starting to be stressed by this article. I wanted it completed some time ago. As my emotions get on edge, I become a bit grumpy, and find that I am less pleasant with the people around me, particularly my co-authors. As I notice my feelings become more aroused, I remind myself that this is a group process, we are all busy, we are making progress, and it will be good. Telling myself these things helps me to relax a bit and calm myself down. I have lowered my level of emotional arousal and am better able to focus on the task at hand. I have self-regulated my emotions.*

The self-regulation of emotions is a process that involves the analysis, control, alteration, or prevention of emotional expression and experiences that are adaptive for a situation. Emotion self-regulation may occur at different times relative to the emotional response. Emotions may be regulated either by manipulating antecedents to emotional response tendencies or by manipulating responses to those tendencies (Gross, 1998).

When focusing on manipulating the antecedents to emotional response, self-regulatory strategies may include situation selection, situation modification, attention deployment, or cognitive change (Gross & Thompson, 2007). Situation selection refers to approaching or avoiding certain people, situations, or environments on the basis of their likely emotional impact. This strategy requires an understanding of emotional responses that can be expected from interaction or lack of interaction with these people, situations, and environments. For example, talking with a certain person always leads to heightened, negative emotion, and so you avoid that person. During situation modification, an individual modifies the environment to alter its emotional impact. If you cannot avoid talking to that person, you might bring a friend along with you to help you stay calm. Attention deployment is turning one's attention away from something in order to influence emotions. You are in a situation where there is this person you do not want to talk with, and so you make sure that you are constantly engaged in talk with others. Cognitive change refers to the way with which we mentally appraise a situation to alter its emotional significance. This may be done by changing how we think about the situation or about our capacity to manage the demand it poses. Cognitive change requires strong cognitive self-regulation skills. For example, one can mentally prepare for having a conversation with someone and tell oneself that it will be "no big deal."

When focusing on the emotional response itself, self-regulation strategies include those that intensify, diminish, prolong, or curtail ongoing emotional experience, expression, or physiological responding (Gross, 1998). For example, after a challenging conversation with a person, saying to oneself "That is an idiotic position to take, or thing to say, but I do not have to be bothered by it," could reduce emotional arousal.

It is important to note that emotion regulation is used not only to reduce a negative level of arousal, but also to intensify or prolong. In order to increase one's motivation for doing well on an exam, a person might actively seek to increase emotions of anxiety.

When discussing the self-regulation of emotions, it is important to provide an understanding of what we mean by the term “emotion”. Emotions occur when an individual evaluates internal or external emotional cues, and this evaluation triggers a coordinated set of behavioral, experiential, and physiological emotional response tendencies (Gross, 1998). These tendencies may be modulated, and this modulation gives final shape to outward emotional responses. During emotion self-regulation, a person may increase, decrease, or maintain positive and negative emotions (Koole, 2009).

An interesting study conducted by Barrett, Gross, Christensen, and Benvenuto (2001) showed that individuals with more highly differentiated and more intense negative emotional experience reported greater emotion regulation, while positive emotional differentiation and intensity were unrelated to emotion regulation. Sometimes our emotions can be kind of a mess and difficult to sort out, but for those individuals who are able to be clear about what they are feeling, particularly in the case of negative feelings, self-regulation appears to be easier. This suggests that the regulation of emotions might be considered separately for positive and negative emotions. This may be because negative emotions tend to have more immediate consequences if they are not dealt with.

Developmental studies have shown self-regulation to play a crucial role in children's social competence (Cicchetti, 1994; Eisenberg et al., 2000); in other words, impairments in emotion self-regulation affects children's capacity to regulate their emotions, and emotion dysregulation in turn leads to social difficulties. Difficulties with emotion regulation may result in psychosocial problems, such as high levels of negative affect and escalation of anger, aggressive-disruptive behaviors, antisocial behaviors, addictions, suicidal ideations, and mood disorders such as depression (Wyman, et al., 2010). Emotion self-regulation recruits less cortical activation in the ventral-prefrontal cortex with age, suggesting that individuals are better able to regulate emotions with age and development (Lamm & Lewis, 2010). Activation of the ventral medial prefrontal cortex is associated with successful suppression of emotional responses to a negative emotional signal (Hänsel & Känel, 2008).

Emotion self-regulation skills closely relate to the other dimensions of self-regulation: physiological, behavioral, and cognitive. According to Saarikallio (2010), the regulation of emotion is accompanied by the regulation of physiological and behavioral processes related to the specific emotion. Thinking about the situation one is in and what one wants out of the situation, influences the emotional arousal. Thus, improved cognitive self-regulation will result in a greater ability to assess emotional situations, monitor emotional situations, and respond using cognitive or meta-cognitive strategies. In return, a stronger ability to self-regulate emotions will result in a stronger ability to mentally assess a situation, and respond with appropriate behavior.

### **Self-Regulation Of Emotion In CHARGE Syndrome**

It is often difficult for children with severe disabilities, including CHARGE, to develop self-regulation skills, and challenging emotional outbursts are common. Conditions that may contribute to difficulty with emotion self-regulating include multiple sensory impairments, difficulty and delay in language development, executive dysfunction, communication difficulties, and poor health and pain. Communication and sensory information are important for learning how to regulate through

experiences and feedback, and it is likely that impairments in these areas contribute to poor self-regulation among individuals with CHARGE. Hearing impairment may cause difficulty processing new information, answering questions, and following directions, while vision impairment may cause difficulty in processing facial expressions, imitating socially acceptable behavior, and focusing on other visual stimuli. Learning how to interpret and express emotions is highly dependent on how the experience is shaped through modeling, which is reduced by communication and sensory problems.

A study by Hartshorne, Nicholas, Grialou, and Russ (2007) explored executive dysfunction among children with CHARGE Syndrome using the Behavior Rating Inventory of Executive Function (Gioia, Isquith, Guy, & Kenworthy, 2000). The authors report that about one third of individuals had difficulty on the emotional control scale of the instrument, and half had clinical scores on the behavioral regulation index. Thus it appears that individuals with CHARGE may have some difficulty with self-regulating their emotions. Due to the many challenges faced by these individuals, it may be difficult for someone with CHARGE to understand when they are feeling an emotion, what it is that they are feeling, and how to regulate or control it. As DeGangi (2000) points out, early deficiencies in self-regulation may lead to challenging behavior, and deficits in attention and inhibition.

### **Interventions To Help With Self-Regulation Of Emotion**

An important step in teaching children to self-regulate their emotions is first teaching those children what it means when they are feeling an emotion. Teaching a feeling vocabulary to a child may be useful here. This could be done using scaffolding techniques, or breaking the process of recognizing and responding to emotions down into smaller, discrete tasks. Modeling of emotions and how to respond in specific situations, as well as using role-play activities, is a useful method when teaching feeling words and how to recognize each feeling. When modeling emotions, mirroring feelings using exaggerated facial and body movements may help children understand how to recognize emotions in other people. The use of differential reinforcement can be very effective in teaching appropriate emotional responses by rewarding positive, appropriate emotions in a situation and reducing negative, inappropriate emotions. It is important to take advantage of opportunities to teach a child feelings when they are noticeably feeling an emotion. When you know that a child is feeling happy, angry, or frustrated, for example, this could be a good time to help them understand that emotion by modeling, and by showing them appropriate responses to that emotion. Concrete aids may help children understand or express emotions as well. Examples of concrete aids may be using a color or face chart to describe feelings, or using a “traffic light” to describe the strength of the feeling. For children with more significant difficulties, repetition may be important when teaching about feelings.

While teaching the child a feeling vocabulary is useful, strategies for reducing the strong, negative emotions are necessary. When self-regulating emotions, an individual may either alter or avoid triggers that produce an emotional feeling, or alter the emotional feeling after it occurs. By helping children to recognize situations in which they are likely to have a strong feeling, you may begin to teach them how to engage or avoid those situations, or limit their emotional effect. When dealing with a feeling after it occurs, practicing how to respond to feelings with the child or walking them through how to deal with the feeling will likely be helpful. It is also important to teach the child calming techniques when they are feeling a negative emotion. Calming techniques may include breathing techniques, exercise or mindfulness techniques such as meditation, Tai Chi, or yoga, or having attachment or stress reduction objects available. Having a “safe place” available for the child to go to during an emotional response

may help that child calm down. For children with significant difficulties, maintaining a consistent environment and routine may also be important to reducing inappropriate emotional responses. However, it is also important to teach the child how to respond in different situations and with different people, and to help the child form secure stable relationships with as many people as possible. This will help the child's self-regulation skills to generalize to new environments, situations, and people.

### **Self-Regulation Of Emotion In CHARGE Syndrome: Case Example**

Before starting at a new residential school, Sarah, age 14 and diagnosed with CHARGE syndrome, had been excluded from school for two years as a consequence of her difficult behavior.

Some of Sarah's issues:

- Regular emotional outbursts
- Unable to tolerate being with her peers and very easily annoyed or upset
- Inappropriate attempts to initiate interactions
- Saying hurtful and inappropriate things to others
- Being very angry with herself, with a negative self-image and self-esteem
- Lack of awareness of her own emotions - Sarah had an analytical understanding but was not able to link this information to what she herself was feeling
- Unable to negotiate or tolerate when things did not go her way
- Difficulty coping with the unexpected
- Difficulty controlling her own behavior and impulsivity (saying "I just want someone to make me stop.")

A full team of practitioners were involved in developing a program to support Sarah, which was regularly monitored and strategies developed or adapted in response. Some strategies developed to support Sarah in this area were:

- Providing Sarah with her own space, with a gradual reintroduction to being in the room with her peers
- Ensuring a consistent routine, with any changes kept to a minimum and every effort undertaken to prepare Sarah for changes
- A highly individualized curriculum, using Sarah's strengths and interests to build her confidence and self-esteem
- Modeling and discussing how Sarah might respond in different social situations. For example, in preparation for attending a local youth club, staff discussed with Sarah what was likely to happen; how to respond if someone said hello; how to initiate a conversation; how to move away if the situation became too much. A picture-board sequence was used to provide a concrete visual cue
- Sarah struggled during role play, and so plastic characters were used. Social scenes were enacted, exploring different ways a person might respond in each situation. This was also used to re-enact situations that Sarah had found difficult, exploring what might have been a more positive way of managing the situation
- Weekly yoga was introduced to help Sarah develop and regulate her physiological state more effectively. "Deep belly breathing" became a useful strategy for Sarah to use independently to help her calm

- Pet therapy provided Sarah with the opportunity to enjoy caring for and nurturing Darcy the dog.
- Discussing Sarah's own emotional state. Initially she found this extremely difficult, and staff who knew her well labeled her emotional states for her (e.g. 'I think you are feeling a bit frustrated,' etc.). A break though came when Sarah was in conversation about a forthcoming trip and suddenly said: "I'm feeling something". Unable to label what she felt, the supporting adult explained she was probably feeling a bit excited and also anxious.

Now 18, it has taken time but Sarah has made huge progress. She is a much happier young lady who is fully included in her class and making real friendships. She is much more socially aware and able to socially engage, sharing a joke and coping with gentle teasing. Sarah is more in touch with her own emotional state and is better able to regulate her emotions and behavior. Very importantly, Sarah is now able to talk about how she is feeling which has proved to be vital as she has undergone a period of ill-health requiring hospitalization and surgery. Overall she has grown in confidence and is looking forward to moving on to college in the near future.



## 6. Physiological Self-Regulation in CHARGE Syndrome

*Andrea E. Larsen, Timothy S. Hartshorne, David Brown, and Jude Nicholas*

The self-regulation of physiology refers to the ability one has to alter one's own states and responses to meet the needs of the body. It also includes identifying warning signs and learning how to calm the body down.

*Once in graduate school when I (Hartshorne) was studying at my favorite study carrel in the library, someone sitting right behind me started tapping the desk with his pencil. It slowly drove me crazy. The noise completely distracted me, I began to squirm in my chair, I started to sweat, and my stress levels increased dramatically. I lost the ability to keep myself in a nice, calm state. Several interventions posed themselves, one was rather violent; but, the most reasonable was to move to a different carrel.*

Consider the experience of walking into your living room at home. Is your attention drawn to the paintings on the wall? Or to the peculiar way the furniture is arranged? Most likely you do not even notice. These perceptions are habitual for you. They have faded into the background. But imagine instead walking into the room to show a friend who has never seen it before. Now you notice the discoloration on the wall, and the messy way papers and magazines are piled on the coffee table. Your perceptions have become much more sensitive to the environment.

Humans generally learn how to habituate to their experience, both internal and external. Habituation is the process by which an individual decreases or ceases to respond to an initially novel stimulus after repeated presentations. In other words, we stop noticing that crack in the ceiling because it has become so familiar. Thus, habituation is characterized as the filtering out of sensory impressions which are no longer relevant. If we could not do that, we would end up being highly over-reactive, as with the tapping of the pencil. This is the opposite process to habituation and is called sensitization, i.e. an increase in the elicited response from repeated presentation of a stimulus. However, sometimes it is beneficial to be highly sensitive to what is going on around us so that we do not miss noticing things that could be very important.

Physiological self-regulation refers to the ability to alter internal states and functions to maintain or regain a state of homeostasis in response to demands on the individual (Sedges, 2007). It is important to be able to regulate homeostasis, like a thermostat. Self-regulation involves the ability to efficiently adapt to and alter one's response to over and under stimulation. Physiological regulation describes the way the individual reacts, or their reactivity, to stimulation, which is under the control of the somatic, endocrine and autonomic nervous systems. However, it is also capable of conscious and deliberate management. Self-regulation attempts to manage this reactivity in a manner that allows the individual to still pursue goals. If you need to study, and your arousal level is too high (over reacting) where you are sitting, you might try moving to a less distracting spot.

Dunn (1997) describes how self-regulation can modify the degree of arousal (or reactivity) that we experience. In this model, we may be in a state where we are easily able to ignore much of the external stimulation in our environment, and maintain a low arousal level. We have habituated to the distracting stimulation in the environment (you tune out the pencil tapping). Or we may be in a state of high



alert and notice everything going on around us. We are in a state of sensitization (the tapping seems to become more pronounced). Self-regulation can support habituation, or under arousal, by simply not reacting to or tuning out any stimulation (simply tune out the pencil tapping), or it can support sensitization, or over arousal, by deliberately noticing and reacting to stimulation (react strongly to the tapping). On the other hand self-regulation can actively work against the level of arousal by focusing full attention on all aspects of the situation when under aroused (sensation seeking), or by withdrawal of attention away from stimulation when over aroused (sensation avoiding). The pencil tapping situation involved a highly sensitized state, and the initial response was reacting to the stimulus. Moving to a different, quieter, carrel, would have been the more active self-regulatory strategy of sensation avoiding.

An individual's capability to efficiently self-regulate rests upon several factors related to the maturation of psychological and physiological functioning (Marcovitch et al., 2010). Although limited, infants display regulatory processes from birth, and these become more sophisticated as they develop. Most of the stimulation that infants must first learn to react to is through sensory input. They see, hear, smell, taste, and touch, and they learn to balance (vestibular sense) and they figure out where their body parts are and how they are attached (proprioceptive sense). Using each of these senses they gradually learn to habituate or react as needed for the task in front of them; for example, they can tune a noise in or out depending on their goals at the moment. The responses produced as they self-regulate are generally found to be fairly stable amongst infants, but the sensory threshold level required to generate a response may vary (Calkins & Fox, 2002). Physiological self-regulation requires the active management of these thresholds of habituation and reactivity to allow optimal performance in the pursuit of a goal. Capabilities to self-regulate physiological states become increasingly more controlled as one develops. In children, gains in self-regulation have been found to correlate with increased ability to attend and motor control (Calkins & Fox, 2002).

The proper functioning of many internal and external responses is dependent on the maintenance of a physiological balance referred to as homeostasis. Numerous physical and chemical variables, such as one's body temperature, are constantly maintained within a set range. Maintaining a homeostatic balance is crucial for the effective functioning and survival of all cells. Preserving one's internal environment can further influence one's reactivity, and the ability to uphold a desired level of awareness (Bornstein & Suess, 2000.)

There are several physiological measures of how well the individual self-regulates. Resting heartbeat (Groome et al, 1999), brain electrical activity (Compton, Hofheimer, & Kazinka, 2013), and levels of the hormone cortisol (Calkins & Fox, 2002) have all been used to better understand self-regulatory strength. Researchers have also been interested in how self-regulating can wear a person out, making it harder to self-regulate the next time. If you use your self-regulatory strength to tune out pencil tapping, you may have a harder time later that same day when you have to regulate another event. It appears blood glucose levels are affected. Reductions in self-regulatory strength due to declining glucose levels can be generally reversed through glucose consumption (Galliot et al., 2007).

### **Physiological Self-Regulation in CHARGE**

Individuals with CHARGE syndrome are truly multi-sensory impaired (Davenport & Hefner, 2011.). The initial development of self-regulation in infants is largely dependent upon sensory stimuli. Therefore, a child born with CHARGE is immediately placed at a disadvantage for the development of self-regulatory mechanisms. Problems with being able to completely experience the sensory world can impact the



understanding of the environment, reduce the benefit of modeling, and increase the level of confusion and uncertainty. Children with CHARGE will still make self-regulatory adaptations, but these can take a long time to create. For example, the boy pictured on the cover of this monograph is watching television. This is normal viewing posture when you have no vestibular sense, upper visual field loss, poor tactile and proprioceptive perception, and low muscle tone. This boy has figured out a way to regulate his attention by stabilizing and thus regulating himself. Hanging upside down by children with CHARGE is frequently reported by parents. It can be a way to regulate the physiological state of the individual so that goals may be achieved.

Difficulty with balance is common in CHARGE. Brown (2005) identifies challenges such as memory deficits, managing sensory perception, and utilizing body language. Also impacted are postural control, equilibrium, muscle tone, and motor coordination. Low muscle tone has been related to diminished levels of sensory input and perceptual awareness. Balance difficulties pose delays in the maintenance of a stable visual field and may contribute to attentional and motor development deficits. The possible impact on self-regulation is not known, although research has found that increased attentional capacities and motor control signify advanced self-regulation in infants (Calkins & Fox, 2002). Vestibular problems would most likely make certain kinds of physiological self-regulation more challenging because the person must deal with and attend to their unsteadiness, in addition to whatever else might be confronting them.

Children with CHARGE typically display a variety of anomalies that interfere with proper neurodevelopment (Gilles, 2011.) Multiple cranial nerve anomalies appear to be common. These nerve dysfunctions may be related to absent or reduced sense of smell, breathing difficulties, and problematic issues pertaining to swallowing. Dysfunction of the nerves within the face may result in facial palsy and a significantly reduced ability to taste. The impact of neurological differences has yet to be studied; however, they would likely impact the child's ability to self-regulate.

Executive functions, presumed to emanate from the prefrontal cortex, are those capacities used to organize oneself in the environment so that the individual can sustain attention to tasks, shift attention, inhibit behaviors, and initiate actions. There is evidence linking executive functions with the physiological response and regulation of stress (Williams, Suchy, & Rau, 2009). For instance, cardiac activity (i.e. heart rate), a measure of stress reactivity, is controlled, in part, by the pre-frontal cortex, the same brain region housing many of the cognitive processes encompassed by executive functions (Lin, Heffner, Mapstone, Chen, & Porsteisson, 2014). Thus impaired executive functions are likely to have a direct effect on the regulation of stress response systems. Hartshorne, Nicholas, Grialou, and Russ (2007) used the Behavior Rating Inventory of Executive Function (BRIEF) to study executive functioning in children with CHARGE. High scores were found for shifting attention, the ability to self-monitor one's actions, and inhibiting behavior. The BRIEF has a "behavioral regulation index" on which over half the children received clinical scores.

Self-regulation can also be impeded by pain (Sauer, Burris & Carlson, 2010) and stress (Blair and Diamond, 2008). Children with CHARGE syndrome are at risk for experiencing much higher levels of physical pain and psychological stress throughout their lives (Nicholas, 2011; Stratton & Hartshorne, 2011). Both internal and external stressors such as the presence of emotional stress and physical pain initiate the body's physiological stress response in the attempt to restore homeostasis. Individuals with CHARGE appear likely to experience many threats to their homeostatic balance, and the resulting stress can impede the maintenance of physical, cognitive, and emotional difficulties, and hinder self-regulatory capacities (Sauer et al., 2010).

## **Interventions To Help With Self-Regulation Of Physiology**

### *Sensory Stimulation*

The first means of developing self-regulatory skills during infancy is typically dependent upon one's senses. Promoting sensory stimulation may be beneficial for the self-regulatory capacity in children with CHARGE, due to extensive multi-sensory impairments. However, each child diagnosed with CHARGE possesses their own unique sensory needs. Occupational therapists can create "sensory diets" specifically tailored towards each individual. A "sensory diet" refers to a list of recommended activities and modifications based on the child's daily sensory needs, such as the use of a trampoline, a sandbox, simple exercises, finger-fidgeting, and applying a weighted vest or blanket (Nackley, 2001). Before a suitable "sensory diet" can be generated, the child should be assessed for deficits in motor control and proprioceptive, vestibular, and tactile information processing.

### *Mindfulness exercise*

Interest in mindfulness interventions has been growing for a number of years (Brown, Ryan, & Creswell (2007). The key to self-regulation in this context is the directed attention of the person to their subjective internal, emotional and physical experience (Brown, et al, 2007). Yoga is one mindfulness exercise that has been used with children who are deafblind (Karnad, 2002). Barrey-Grassick (2011) adapted Tai Chi for children who are deafblind and those with CHARGE. These practices have the potential to develop self-regulation skills.

### *Wilbarger Protocol*

A unique treatment plan known as the Wilbarger protocol, also referred to as brushing and joint compression, was specifically designed to meet the needs of children who experience deficits in sensory reception and integration. It is intended to help children achieve and sustain optimal levels of arousal (Kimball, Lynch, Stewart, Williams, Thomas, & Atwood, 2007.) Application of the protocol not only entails sensory stimulation, but promotes relaxation which can further aid in the reduction of stress (Kimball, Lynch, Stewart, Williams, Thomas, & Atwood, 2007.)

Employing the technique involves a specialized non-scratch brush which is used to apply deep pressure. Following the pressure, each of the major joints must then be compressed. This procedure must be used only after proper training, and followed on a routine basis for optimal benefit (Kimball, et al 2007).

### *Diet*

The ability to utilize self-regulation is affected by blood glucose levels, and so a balanced diet and adequate fluid intake is important. It is beneficial to eat several smaller meals throughout the day and focus on including a sufficient amount of carbohydrates into the diet. Foods lower on the glycemic index are preferred because they prolong the release of glucose, which aids in reducing fatigue (Hagger, Wood, Stiff & Chatzisarantis, 2010).

## **Case Example**

The student was an eight year old boy, profoundly deaf, blind in one eye with reduced visual field in the other eye, late independent walking (aged 4), and with continuing feeding issues. He attended a deaf program. The boy had impressive receptive and expressive language in American Sign Language, but showed characteristic difficulties with initiating utterances. Problem behaviors were distractibility, impulsivity, and physical aggression against others. During two days of observation it was apparent

that episodes of high arousal leading to violent outbursts were related primarily to periods of enforced sitting for up to 25 minutes at a time. Recommendations included providing a high desk so that the boy could stand to do his schoolwork – he enjoyed this and showed greatly improved attention span during lessons with fewer aggressive outbursts, swaying his body from side to side and stepping from one foot to the other as he worked. Significantly improved attention was also observed after the weekly 30-minute adaptive physical exercise session, so as a result these sessions were increased to three times a week, and physical activities providing more and stronger proprioceptive and vestibular input were added – mainly swinging by the arms from an overhead beam, trampolining, wheelbarrow walking (student walks on their hands and arms with their legs held up by an adult), and pushing the heavy equipment cart a long distance to and from the exercise room. The student had received an occupational therapy brushing and joint compression program as an infant, and the recommendation that this kind of program should be offered again was taken up by the school, who reported that the student enjoyed the session twice a day and appeared to benefit from it in terms of improved self-regulation with better attention span. A more general recommendation was for classroom staff to wait for the student's visual attention before communicating to him, to sign a little more slowly, and to generally reduce the pace at which activities were carried out. All these suggestions were facilitated by the fact that the student already had an allocated one-on-one classroom aide – originally purely to impose physical control on the student, but increasingly fulfilling the functions of a deaf-blind intervenor as the benefits of these recommended changes became apparent.



## 7. The Importance Of Self-Regulation

*Jude Nicholas, Timothy S. Hartshorne, and Megan Schmitt*

Self-regulation comes in different dimensions. The four dimensions of self-regulation (cognition, behavior, emotion, and physiology) come into play within complex interactive combinations in the ongoing daily life behavior of individuals with CHARGE. Furthermore, the four dimensions of self-regulation are influenced by several underlying mechanisms such as genetic predisposition, biology (body & senses), neurobiology (brain networks) and experience.

Self-regulation is an important skill for all children to develop. Self-regulation of cognition helps children set goals, follow rules, and plan out the appropriate response. Self-regulation of behavior helps children demonstrate control over their actions. Self-regulation of emotion helps children manage how they express and experience emotions. Self-regulation of physiology helps children to alter their own states and responses to meet the needs of the body.

Self-regulation may be particularly difficult for children with CHARGE to develop. Although individuals with CHARGE syndrome may develop the ability to self-regulate, failures are common, and they may lose control of their behavior in a wide variety of circumstances. However, similar to the large variability across individuals with CHARGE syndrome, impairments of self-regulation in CHARGE syndrome also vary widely. Therefore, some individuals with CHARGE may have more difficulty self-regulating than others. Additionally, there is variability in the dimensions with which individuals with CHARGE have difficulty regulating. For example, one individual may have more difficulty self-regulating his or her behavior; whereas, another may have more difficulty self-regulating his or her emotions. When individuals with CHARGE have trouble with regulating their thoughts, actions, feelings and physiological states to situational requirements their behavior may be perceived as difficult or challenging.

Self-regulation is an essential function in each of our daily lives. We all have to regulate ourselves physically, behaviorally, cognitively, and emotionally to achieve our day-to-day activities. Although we may not realize it, each small step in our lives is a tiny goal that requires us to regulate ourselves to achieve that goal. We engage in a variety of behaviors to self-regulate so that we might achieve our goals and live functionally in the world. Children with CHARGE syndrome must also regulate themselves to achieve small goals throughout their daily lives. However, the way a child with CHARGE syndrome experiences the world is generally different than how most of us experience the world. While we all have a different view of the world, a child with CHARGE syndrome experiences things uniquely given their various sensory impairments. Therefore, the behaviors that children with CHARGE syndrome engage in to self-regulate to live functionally within the world will likely be different than the behaviors others use to live in the world. Often the behaviors children with CHARGE engage in may seem inappropriate, extreme, or weird, but we must remember that these behaviors are generally adaptive and creative ways children with CHARGE syndrome have learned to self-regulate to achieve their daily goals and function successfully in the world, because their self-regulation skills are not well developed.

Reflecting back on the different case examples, some of the behaviors upon first consideration seem strange or inappropriate. Think back to six-year-old Matthew (page 21). He engaged in several 'odd' behaviors—examining a bowl from different angles and making clicking sounds with his tongue while

water was in his mouth. Quite often, these harmless behaviors are viewed as inappropriate and needing to be stopped. However, each of these seemingly strange behaviors served a function for Matthew to help him experience his environment so that he might remember a previous experience, which ultimately supported his ability to regulate his cognition in order to ask his mother for a preferred activity. Consider the outcome if Matthew's mother had stopped him from touching the water or from clicking his tongue while the water was in his mouth. Matthew's opportunity to explore his environment and to gain the sensory experience that helped him regulate his cognition to remember a previous event and request a preferred activity may have been limited, and he might have become upset. Therefore, whenever an individual with CHARGE engaging in an odd or inappropriate behavior we need to determine what function that behavior is serving, how that behavior may be helping the child to self-regulate, and what goal the child may be trying to achieve. By doing this we gain a better understanding of the child and the child's wants and needs.

Self-regulation is a powerful skill that has an important impact on individuals with CHARGE throughout their lives. Developing self-regulation takes time as well as the support of parents and other adults. Our roles as professionals and parents should be that of a bi-directional mediator or a bridge, which facilitates the child's interaction with their environment and provides them with the tools to "see" and "be" themselves within their environment. We can start early and plant the seeds of self-regulation so the children with CHARGE can go on to refine and build on them over time.

Researchers have found that self-regulation skills in children can be developed with practice and they can also be influenced by experience. A number of different interventions have been offered in this monograph. . With more research and experience, we can hope to refine our understanding of the development of self-regulation in CHARGE, and improve our approach to this critical process.

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Gail Deuce is a specialist teacher for children and young people who are deaf blind/multi-sensory impaired. She has approximately 30 years' experience in the field of special education in the UK. Gail currently works part-time for a school in a London borough as a specialist outreach teacher for children who are multi-sensory impaired and also supporting inclusion of children with severe learning difficulties. In addition, she works as a freelance consultant and is a tutor for the University of Birmingham MSI distance learning course. Gail explored the education of learners with CHARGE while completing her PhD.

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Tim Hartshorne, Ph.D., is a professor of psychology, specialized in school psychology, at Central Michigan University. He is the grant holder for DeafBlind Central: Michigan's Training and Resource Project, which provides support to children who are deafblind in Michigan. Much of his work is influenced and motivated by his son Jacob, who was born in 1989 with CHARGE syndrome. Tim's particular interests include understanding the challenging behavior exhibited by many individuals with deafblindness, CHARGE, and related syndromes, and also how severe disability impacts the family. In 2005 he was awarded the Star in CHARGE by the CHARGE Syndrome Foundation.

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Andrea Larsen received her bachelor's degree from Central Michigan University in 2012, with a focus in psychology and the general sciences. Her desire to actively engage in the medical profession led her to join the CHARGE Lab while attending CMU. After graduation she became injured while gaining clinical

hours as a certified nurse assistant and has been largely consumed by her rehabilitation and new role as a mother. Her firsthand experience with her injury and the diagnoses of another rare syndrome, referred to as Thoracic Outlet Syndrome, is beginning to drive her passion for the medical field even more so than before.

Jude Nicholas (Psy.D.) is a licensed clinical neuropsychologist, who is attached at Haukeland University Hospital and at Statped Vest center for special education in Bergen, Norway. He has some 20 years of clinical experience working with children and adolescence with sensory impairment and developmental disabilities. He has a longstanding interest in syndromes with sensory impairments and cognitive functions. He is author of several articles and book chapters on these topics, especially on Congenital Rubella Syndrome and CHARGE syndrome. His current research investigates the neuropsychological functions of tactile cognitions, particularly in persons with dual sensory impairment/deafblindness.

Maria A Ramirez is a member of the CHARGE syndrome lab and doctoral candidate from Central Michigan University. She completed her internship at the Children's Hospital of Philadelphia and currently resides in Chicago. She has worked on articles related to self-regulation and emotion regulation in children with CHARGE. Presently she is collecting data for a study which is examining the effects of an adapted form of Tai Chi (fun chi) on sleep and emotion regulation.

Megan Schmittel is a doctoral candidate in the school psychology program at Central Michigan University. She has been a member of the CHARGE Syndrome Research Lab at Central Michigan University since 2013 and is interested in social play in children with CHARGE syndrome. After completing her degree, she plans to continue research in CHARGE syndrome and work in a hospital or clinic providing services to children with low-incidence disabilities.

Andrea Wanka, Ph.D., is commissioner for Deafblindness at the stiftung st. franziuskus heiligenbronn in Germany. She works primarily with congenital deafblindness, but has also specific knowledge and experience on acquired deafblindness. She has a focus on CHARGE syndrome in her work. In her doctoral program she analyzed early parent-child-interactions with children with CHARGE. She received the Young Professional Leadership Award from Deafblind International, and has published five books on CHARGE Syndrome in German.

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