Self-Regulation of Cognition in CHARGE Syndrome

Benjamin Kennert[1], Timothy S. Hartshorne[2], Andrea Wanka[3], Heather Dix[4], Jude Nicholas[5]

"Today I intend 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". (Tim Hartshorne)

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. Mental processes involved may include attention, memory, learning (as well as using prior learning), reasoning, problem-solving, decision-making, metacognition, and motivation around goal directedness. A lack of cognitive self-regulation can result in the individual being unfocused. Too much cognitive regulation can result in obsession.

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

[1] Doctoral student Central Michigan University (kenne1bs@cmich.edu)
[2] Department of Psychology, Central Michigan University (harts1ts@cmich.edu)
[3] St. Franziskus, Heiligenbronn, Germany (andrea-wanka@dbicharge.org)
[4] Central Michigan University graduate student
[5] Regional Resource Centre for Deafblind/Haukland University Hospital, Bergen, Norway (jude.nicholas@statped.no)

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 & 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. Hartshorne, Nicholas, Grialou & 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).

Possible Interventions

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 among individuals with CHARGE. According to Florez (2011) the most powerful way teachers can help children learn self-regulation is by modeling and scaffolding it during ordinary activities. Scaffolding involves 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.

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.

Diamond & 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 the working memory. Many of these strategies have been derived from the current understanding of those cognitive processes involved in the information-processing system. These strategies or techniques can be used to minimize working memory failures and enhance the learning opportunities in individuals with CHARGE. Some of these are rehearsal or working memory load reducing strategies.

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. 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 & 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).

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
- Problem solve how to best communicate his wants
- Engage in planning around his goal

Interventions can be planned around building on these skills; helping Matthew to make them more sophisticated. In particular 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 though pictures, story books, role play and modeling, and then helping him to problem solve methods for requesting access to these activities.

References

Brown, D. (2005). CHARGE syndrome "behaviors": challenges or adaptations? *American Journal of Medical Genetics,* 133A(3), 268-272.

Diamond, A.& Lee, K. (2011). Interventions shown to aid executive function development in children 4 to 12 years old. *Science, 333*(6045), 959-964. doi: http://dx.doi.org/10.1126/science.1204529

Florez, I.R. (2011, July). Developing young children's self-regulation through everyday experiences. *Young Children*, 46-51.

Ford, R. M., McDougall, S. J. P. & Evans, D. (2009). Parent-delivered compensatory education for children at risk of educational failure: Improving the academic and self-regulatory skills of a sure start preschool sample. *British Journal of Psychology, 100* (4), 773-797. doi: http://dx.doi.org/10.1348/000712609X406762

Gioia, G.A., Isquith, P.K., Guy, S.C. & Kenworthy, L. (2000). *Behavior Rating of Executive Function*. Lutz, FL: Psychological Assessment Resources.

Hartshorne, T. S. (2011). Behavioral phenotype in CHARGE syndrome. In T. S. Hartshorne, M. A. Hefner, S. L. H. Davenport & J. W. Thelin (Eds.). *CHARGE syndrome* (pp. 317-326). San Diego, Plural.

Hartshorne, T.S., Nicholas, J., Grialou, T. L. & Russ, J. M. (2007). Executive function in CHARGE Syndrome. *Child Neuropsychology, 13*, 333-344.

Hartshorne, T.S., Hefner, M.A., Davenport, S.L.H. & Thelin, J.W. (2011). *CHARGE syndrome.* San Diego, CA: Plural Publishing, Inc.

Jahromi, L.B. & Stifter, C.A. (2008). Individual differences in preschoolers' self-regulation and theory of mind. *Merrill-Palmer Quarterly*, *54*(1), 125-150. doi: 10.1353/mpq.2008.0007

Jarvela, S, Jarvenoja, H. & Malmberg, J. (2012). How elementary school students' motivation is connected to self-regulation. *Educational Research and Evaluation, 18*(1), 65-84.

Luna, B., Padmanabhan, A. & O'Hearn, K. (2010). What has fMRI told us about the development of cognitive control through adolescence? *Brain and Cognition, 72* (1), 101-113. doi: http://dx.doi.org/10.1016/j.bandc.2009.08.005

Magar, E. C. E., Phillips, L. H. & Hosie, J. A. (2008). Self-regulation and risk-taking. *Personality and Individual Differences, 45* (2), 153-159. doi: http://dx.doi.org/10.1016/j.paid.2008.03.014

Scholnick, E.K. & Friedman, S.L. (1987). The planning construct in the psychological literature. In S. L.Friedman, E.K. Scholnick & R. R. Cocking (Eds.) *Blueprints for thinking: The role of planning in cognitive development.* (pp. 1-33). Cambridge: University Press.

Turner, J. E. & Husman, J. (2008). Emotional and cognitive self-regulation following academic shame. *Journal of Advanced Academics, 20*(1), 138-173.