11th International CHARGE Syndrome Conference Information Central Program Handouts

This session is scheduled over Breakout Sessions #3 and 4 and over multiple rooms. You can wander through the posters and Exhibitor Area stopping wherever you like to visit with presenters and exhibitors. Some may have flyers or handouts available at their poster location and some have submitted PDF handouts for download.

CHARGE Syndrome Foundation, Inc.

Information Central Presenters	Poster Title
Angela Arra-Robar	**It's All About Teamwork**
Marguerite Bilms, Martha Majors, Sharon Stelzer	**Preschoolers: Tomorrow's Teenagers- Educational Strategies that Bridge the Gap**
Susan M. Bruce	**Using the Holistic Communication Profile to Capture the Role of Pivotal Milestones in Language Development** **Supporting the Development of Self-Determination in Pre-linguistic Individuals with CHARGE Syndrome**
Mary Zatta	**Online Course for Care Providers**
Danielle Bushey , Mary Poblete	**Physical Therapy for a Child with CHARGE Syndrome**
Ching-Pin Chang	**Epigenetic regulation of neural crest cell development by Brg1 and Chd7**
Catherine Rose	**Lights for Learning** **Piloting Your Journey**
Gail Deuce	**A CHARGE information pack for practitioners**
Gail Deuce, Steve Rose, Simon Howard	**A Study of CHARGE Syndrome in the UK**
Amy Parker, D. Jay Gense, Peggy Malloy	**Intervener Training Modules**
Emily Maxwell, Kirsten Corbett, Leslie Buchanan	**Collaborative Approach to Behavior**
Julia Benstz, Andrea Wanka	**CHARGE in German-Speaking Countries**
Robert Hevner	**Cerebral Cortex**
Lauren Lieberman, Pamela Haibach	**Balance Workshop** ***Note: parents can retrieve campers from camp to participate in the balance workshop***
Tate Jenkins	**Headaches in CHARGE**
Susan Bruce, Sharon Stelzer	**Socialization in Different Age Dyads of Students with CHARGE Syndrome**
Shantell Johnson, Kirsten Hissong	**Fathers in CHARGE: Work and Friendships**
Sarah Haney	**Behavioral Self-Regulation**
Benjamin Kennert	**Cognitive Self-Regulation in CHARGE Syndrome**
Benjamin Kennert, Maria Ramirez	**Self-Regulation of Emotion**
Maria Ramirez, Tim Hartshorne	**Models of Conceptualizing Self-Regulation in CHARGE syndrome**
Maria Ramirez, Seraphim Mork, Tim Hartshorne	**Microenterprises**
Tim Hartshorne, Andrea Larson	**Physiological Self-Regulation in CHARGE**
Lori Swanson, Sheri Stanger	**CHARGE Families in the Know Read and Listen to …**

Gloria Rodriguez-Gil, Myrna Medina	**Educational Services for Children with CHARGE Syndrome (Spanish)**
David Brown	**Why self-stimulation is a good thing, and how and why we should interpret it**
Steve Rose	**Challenges of Mealtimes**
Wendy Bridgeo	**Futures Outcomes**
Holly Cooper	**Technology for Learning, Literacy and Fun**
Gloria Rodriguez-Gil	**iPad & Young Students with CHARGE**
Tim Hartshorne	**Behavior as self-regulatory adaptation, or "I can't believe my child just did that!"**
Maria Ramirez	**Self-regulatory strategies for children with CHARGE syndrome**
James Schwark	**They're not children anymore: Communicating effectively with adult providers**
Meg Hefner and Emily Fassi	**CHARGE Syndrome Clinical Database**

CHARGE Conference July 2013

Preschoolers: Tomorrow's Teenagers-Educational Strategies that Bridge the Gap

Marguerite Bilms, M.Ed.
Martha Majors, M.Ed.
Sharon Stelzer, M.Ed.
Perkins School for the Blind
Deafblind Program

Program Components for Students with CHARGE

- LANGUAGE AND COMMUNICATION
- Use of Total Communication
- Supports to use Speech
- Supports to use Sign language

- CURRICULUM
- Must include **teaching strategies** that are effective for students with CHARGE Syndrome

Accommodations to Support Access to the CURRICULUM that is FLEXIBLE

- Supportive Environments that include
- Communication
- Vision and Fatigue related to use of Vision
- Hearing and access to information
- Cognition and adapted content as needed
- Motor (both fine & gross motor)
- Sensory and Sensory Breaks

What Students with CHARGE Syndrome Should Learn

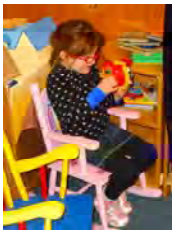
- How to be an effective communicator
- How to be social
- How to be part of a group

Preschool: Frequently Used Teaching Strategies

- Choice Making
- Negotiation
- Clear Expectations
- Clear Beginning-Middle-End
- Behavioral Strategies

Curriculum: Pre-School

Play & Cognition



Curriculum: Pre-School
Language & Communication


Curriculum: Pre-School
Sensory Integration

Curriculum: Pre-School
Social Development

Curriculum: Pre-School
Self-Help Skills


Teaching Strategies

Beginning-Middle-End	Choice Making	Organizational Skills
Partial vs. Full Participation	Task Analysis	Prompt Levels
Motivation	Clear Expectations	Active vs. Passive Learning
Follow Student's Interest	Needs Board	People Preferences
Signals (verbal, auditory, visual)	Pause time for response	Structure and Routine

VI. Teaching Strategies
Pre-School

Beginning-middle-end	Choice Making	Organizational Skills	Partial vs. Full Participation	Structure and Routine
Prompt Levels	Motivation	Clear Expectations	Follow Student's Interest	Pause time For response

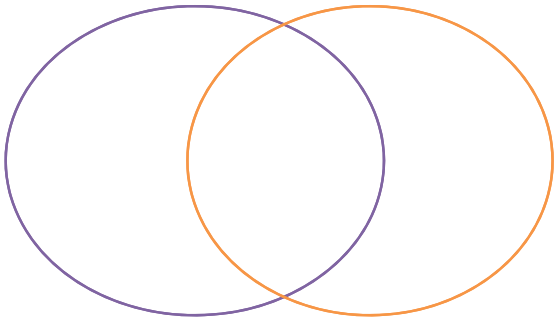
Functional Academics (Teenager): Frequently Used Teaching Strategies

- Negotiation
- Structure & Routine
- Following Student's Lead
- Choice Making
- Clear Beginning-Middle-End
- Teaching in Natural Environments
- Behavioral Strategies (environmental management, etc.)

Similarities in Teaching Strategies

- Negotiation
- Choice Making
- Behavioral Strategies

Today's Pre-Schoolers, Tomorrow's Teenagers



Implications of Similar Teaching Strategies

- Students' needs for strategies are "Life long"
- Starting young can provide time to "practice" skills
- Skills can develop and mature over time
- Provides structure early on that can be a life long tool
- Provides consistency throughout all environments

Questions & Wrap-Up

Contact Information

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- Martha.Majors@perkins.org
- Sharon.Stelzer@perkins.org

Physical Therapy and CHARGE syndrome

Danielle M Bushey, PT, MS, NCS
Mary Poblete, PT, DPT

Physical Therapy

- *Physical therapy is "a dynamic profession with an established theoretical and scientific base and widespread clinical applications and the restoration, maintenance, and promotion of optimal physical function."*

Role of a physical therapist

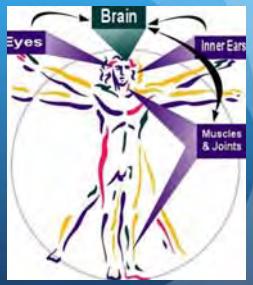
- *Diagnose and manage movement dysfunction and enhance physical and functional abilities*
- *Restore, maintain and promote, not only optimal physical function, but optimal wellness and fitness and optimal quality of life as it relates to movement and health*
- *Prevent the onset, symptoms, and progression of impairments, functional limitations, and disabilities that may result from diseases, disorders, conditions, or injuries*

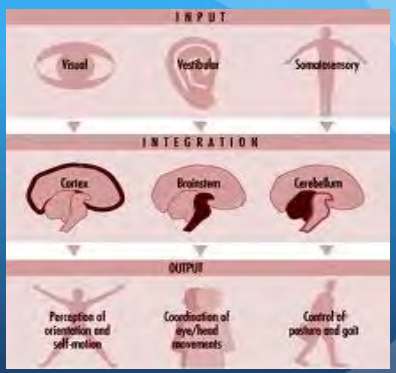
What are the physical challenges of individuals with CHARGE?

- Cardiac
- Respiratory
- Tone
- Range of motion
- Posture
- Balance
- Coordination
- Vision
- Hearing
- Cranial Nerve
- Other comorbidities:
  - Surgical intervention
  - Developmental delay
  - Medication side effects
  - Nutrition

The complex BALANCE system

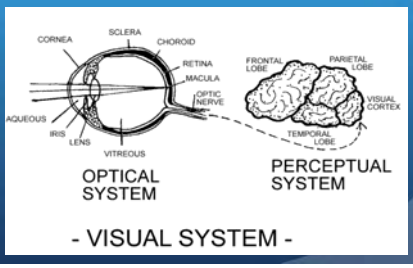
- Balance is maintained through a complex interaction of the Central Nervous System (CNS) and the three primary sensory systems:
- VISION
- VESTIBULAR
- SOMATOSENSORY





Visual System

- A strong inhibitor of the other two systems (vestibular and somatosensory)

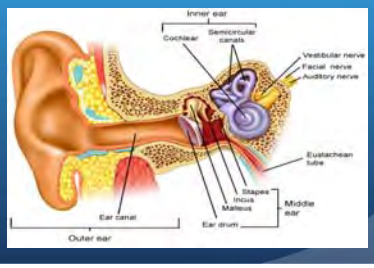


Somatosensory System

- The perception of sensory stimuli from the skin and internal organs of the body
- It is the fastest of the three sensory systems
  - light touch
  - proprioception
  - deep pressure
  - temperature
  - kinesthesia
  - vibration

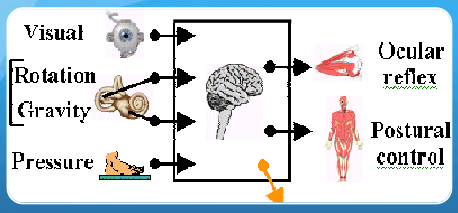
Vestibular System

- The conflict resolver between the other two systems (vision and somatosensory)



Why do individuals with CHARGE have such a difficult time with balance?

- Vision
  - Colobomas
  - Retinal Detachments
- Somatosensory
  - Low tone (joint laxity and decreased proprioception)
  - Sensory processing issues (defensivenes, regulation)
- Vestibular
  - Semi-circular canals
  - Cranial Nerve VIII impairment



If only it were so simple:

Neural connections
System coordination

Motor Control

- Area of study dealing with understanding of the neural, physical, and behavioral aspects of movement.

Motor Control

- Stimulus identification =>
- Response Selection (stimulus processing) =>
- Response Programming (motor output)

Motor Learning

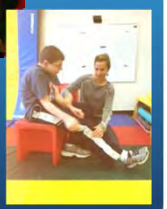
- The improvement of a given functional task resulting from practice or experience. In order for the motor learning to be meaningful, the performance of the new task must be reproducible over time, performed in multiple environments and generalized to similar activities

Principles of Motor Learning

- Amount of practice
- Learner needs to be actively involved
- Task conditions (speed, timing, and environmental conditions)
- Adaptability
- Reinforcement is required
  - Knowledge of results
  - Feedback can be decreased over time

CHARGE and Learning

How do you make a PT session successful with students with CHARGE syndrome?



UNDERSTANDING and KNOWING your student

- Mode of communication
- Behaviors and behavior plan
- Need for structure and routine
- Obsessive Compulsive Disorders
- Limitations

Methods of Developing a Relationship

- Team approach – Communicate with teacher, family members, speech therapist, occupational therapist, Orientation and Mobility specialist, Behavioralist
- Communicate with your student
  - Sign language (visual and tactile)
  - Photos
  - Verbal

Methods of Developing a Relationship

- Avoid power struggles
- Give lots of positive feedback
- Clear expectations
- Sandwiching non preferred activities in between preferred activities
- Choice making
- Mood induction

How should we approach our students?

- Be flexible
- Be patient
- Limit expectations
- Accept slow progress

How to justify physical therapy

- Need to have measurable and functional goals.
  - Need to show progress
- They need to relate to a "disability"
  - Unable to participate in a school sports team
  - Unable to safely play on playground equipment
  - Unable to use stairs for fire safety
  - Unable to walk outside without falling
  - Unable to negotiate curbs or ramps in the community
  - Strength and endurance limit the ability to participate in vocational work

Expanded Core Curriculum

- How does physical therapy relate to access to education?
  - It's not just about being able to walk from one class to another or negotiating stairs!

Expanded Core Curriculum

- The National Advisory Council of the Nation Agenda states that blind and visually impaired students have two sets of essential educational experiences: (1) regular curriculum offered to all students-the core curriculum – and (2) learning experiences required because of vision loss – the expanded core curriculum.

ECC Components

- Compensatory, Functional Skills, Lang/Communication
- Career Education
- Social Interaction/Group
- Independent Living Skills
- Leisure and Recreation/APE
- Orientation and Mobility
- Assistive Technology
- Sensory Efficiency
- Self-Determination

How does the ECC relate to physical therapy?

- Compensatory, Functional Skills, Language/Communication
- Career Education
- Social Interaction/Group
- Orientation and Mobility
- Sensory Efficiency
- Self-Determination

How can physical therapists advocate for their students with CHARGE syndrome

- Understand the importance of functional activities
- Understand the importance of improving balance and coordination for safety in all environments
- Understanding the importance of improving and maintaining flexibility, strength and endurance

Recommendations:

- Exercises: strengthening and stretching
- Follow-up with an orthopedist and/or physiatrist for monitoring of scoliosis, hip integrity, kyphosis, and need for foot bracing.
- Proper seating: to decrease postural fatigue (armrests, feet on floor), prevent postural mal-alignments
- Changes in position for sensory input and to decrease postural fatigue

Balance training in a setting in which the individual can be an active learner. Give the individual with CHARGE syndrome the opportunities to explore, fall, progress and LEARN.

CHARGE Syndrome Clinical Database Project

Principal Investigator: Meg Hefner, MS
Saint Louis University School of Medicine
Department of Pediatrics, Division of Medical Genetics

We have developed a comprehensive database of clinical information on CHARGE syndrome (CS). If you or your child has been diagnosed with CS, you may be eligible to participate.

What is this for? The purpose of this study is to create a comprehensive clinical database and registry of individuals with CS of all ages from all over the world. Information from this database will provide meaningful contributions to CS knowledge and research.

Who can do this? Any adult with CS or parent/guardian of an individual with CS is eligible.

How would I do it? Participation in this project involves entering data (mostly medical information) into a web-based questionnaire. There are opportunities to directly upload photographs and certain medical records in some sections.

How long will it take? Completing the entire questionnaire will take several hours. It can be done in multiple sittings over several weeks. We may ask for yearly updates.

What do I need? You must have email and Internet access to participate in this project. You will need access to your/your child's medical history. The study is in English only.

Will I get paid? No. Your participation is strictly voluntary.

I want more information. What do I do now? If you are interested in learning more about this project, or if you have questions, you can go to the Clinical Database Project link at ***chargeysndrome.org***, or contact Meg Hefner directly at ***hefnerma@slu.edu***. Thank you for your interest in this study.

This project is endorsed by the CHARGE Syndrome Foundation and Saint Louis University.

Cognitive Self-Regulation in CHARGE Syndrome

Benjamin Kennert & Tim Hartshorne, Ph.D. • Psychology Department, Central Michigan University

What is Cognitive Self-Regulation?

- Regulation of thoughts and mental processes to balance between inhibition and initiation of behavior in order to achieve a goal.
- It is voluntary, and requires both an awareness of the process, and a goal-directed action
- 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 requires feedback, and involves thinking about thinking in order to stay on track.

In What Way Might This Be Difficult for Individuals with CHARGE?

- 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 difficulty self-regulating for individuals with CHARGE.
- Hearing impairment may cause difficulty processing new information, answering questions, and following directions.
- Vision impairment may cause difficulty in processing facial expressions, imitating socially acceptable behavior, and focusing on other visual stimuli.
- Children with CHARGE often need extended time to process information (Brown, 2005)
- Executive dysfunction (Hartshorne et al, 2007): Difficulty shifting from one activity or focus to another, tracking own behavior and its effect on others, and controlling impulses and terminating behaviors
- An absence of incidental memory, due to limited environmental feedback, may lead to an absence of cognitive self-regulatory skills (Ford et al, 2009).

Interventions/What Might Help?

- Using scaffolding procedures to learn a new task or to achieve a goal
  - Scaffolding involves helping the child to break down larger goals, tasks, or problems into shorter, more discrete tasks, 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.
- Anything that makes learning easier and reduces stress may be helpful
  - Balance new activities with familiar ones.
  - Modifying distracting surroundings such as noise, light, and people
  - Providing breaks and letting the individual know that others need breaks
  - Use concrete aids
- Relaxation techniques or exercises
  - Breathing
  - Going to a safe place
  - Mindfulness activities such as meditation, Tai Chi, or yoga

- Allowing movement before, during, and after concentration phases of a task may lower stress and increase motivation.
- Create a motivating situation, if possible

Poster design and printing by the Faculty Center for Innovative Teaching at Central Michigna University

References

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

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

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

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CMU
CENTRAL MICHIGAN UNIVERSITY

PHILIPS

Perkins

LightAide

Catherine Rose
July 25, 2013

PHILIPS Perkins

Agenda

- Identify new areas of learning for children with visual impairments
- Identify new ways to assess vision capabilities for children with visual impairments
- Identify new ways to motivate children for therapies

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Inspiration Technology Partnership

Alexis, age 7

Philips Color Kinetics
(architectural lighting)

Perkins

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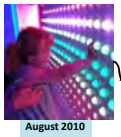
How did we get here?
Engaging Innovation Journey



August 2010
An Idea

April 2011 **3 prototypes**
Product Testing with Children with Vision & Combined Vision/Hearing Impairments

Sept 2012 **5 Alpha Units**
Product Testing with Children with Vision & Combined Vision/Hearing Impairments

May 2013 **20 Beta Units**
More Beta Testing

Customer feedback was incorporated into the ongoing design iterations.

Contact: catherine.rose@philips.com

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What is it?
LightAide Product Overview

Key Benefits
- Helping to teach the children with low vision and multiple disabilities

Key Features
- 224 bright multicolored LED lights
- 4 interactive switch connections to:
  - turn on and off lights
  - move lights
  - learn and engage
- Leader controls via LCD or switch
- Upload new activities via USB
- Portable and Lightweight
- Easy to Operate

Product Launch Fall 2013

Perkins

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LightAide Uses

- **Anticipated:**
  - **Visual Awareness, Cause & Effect**
  - **Eye Tracking, Social Interaction (Turn-Taking)**
  - **Diagnosis (visual field, color awareness)**
  - **Creating Calm Environment**
  - **Matching Lessons (Color, Shapes, Letters)**
  - **Augment current therapies**
- **Unanticipated**
  - **Students used new language**
    - "Light" (verbally)
    - "Buy" (with POD system)
  - **Interactive Play between children**
    - regardless of abilities
  - **Readiness for Switch Use**
    - OT said, "I hadn't planned to introduce switches until next year. She's ready!"
    - TVI said, "a typically distracted student was using the LightAide for 45 minutes using the switch, fascinated!"

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Email me if you have additional questions and comments:
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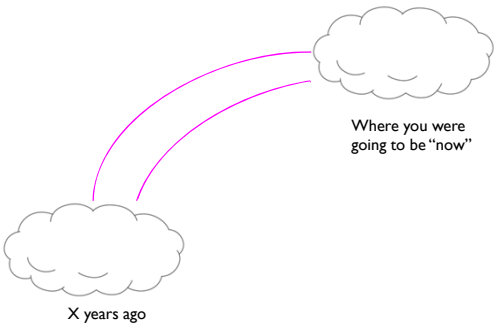
7

Piloting the Journey

Catherine Rose
Catherine.Rose@philips.com

"Inspired Paths
Are Not Linear"
~Anonymous

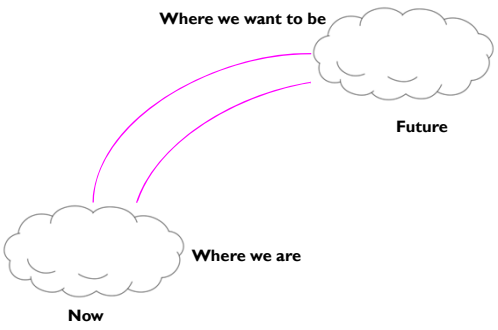
Imagine you can go back in time…



Our Experiences Shape Us

- How we "arrive"
- What baggage we have "in tow"
- Memories
  - 25% great things
  - 75% losses where you learn resilience
- Embrace your story

Where we are Now and Future

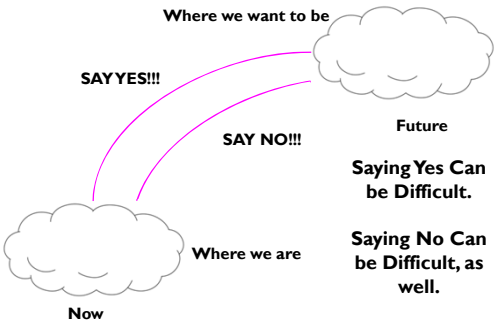


What I really want is….

Step 1:
Spend 5 minutes
Starting all sentences with
"What I really want is"

Step 2:
Re-read and underline &
highlight trends you see.

Where we are Now and Future



Requires Calm & Confident Conversations

- Preparation: Know what you need to say
- Practice: Balance real with rehearsed
- Plexiglass: Strong but unbreakable

Powerful Conversations

- I _____ to _____.
  (Choice language; Verb)
- By that I mean _____.
  (Concise Explanation)
- And as a result _____.
  (Positive Impact to Person)

Choice Language Options			
I demand	I need	I prefer	I would like you to consider
I expect	I want	I would like you	I wonder if you could

Use I - because when you don't you give your power away.

Things May Still Turn Out Unfavorable

- **Three Possible Reactions**
  - **Influence: Make a change**
  - **Acceptance: Accept and move**
  - **Removal: Get out of situation**

Nothing is Worse than being Stuck

Choose to be empowered

Pilot your own Journey.

For More Information

- On this topic:
  - http://caringcalm.blogspot.com
  - Catherine.Rose@philips.com
- On my family:
  - http://schnaderbeck.blogspot.com
- At 2013 CHARGE Conference, also
  - Presenting: **"We Are in CHARGE"**
  - Sharing the **LightAide in "Lights for Learning"**

Self-Regulation of Emotion in CHARGE Syndrome

Benjamin Kennert, Maria Ramirez, & Tim Hartshorne, Ph.D. • Psychology Department, Central Michigan University

What is Emotional Self-Regulation?

- A process that involves the analysis, control, alteration, or prevention of emotional expression and experiences that are adaptive for a situation.
- Emotions may be regulated either by manipulating antecedents to emotional response tendencies or by manipulating responses to those tendencies (Gross, 1998)
- 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)
- It is voluntary, and requires both an awareness of the process and a goal-directed action.
- It is controlled by feedback within ourselves and in the environment (Vohs & Baumeister, 2011)
- When focusing on manipulating the antecedents to emotional response, self-regulatory strategies may include situation selection, situation modification, attention deployment, or cognitive change (Gross, 1998)
- Deficiencies in emotional self-regulation may lead to challenging behavior and difficulty expressing emotions.

In What Way Might This Be Difficult For Individuals With CHARGE?

- 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.
- Hartshorne et al (2007) reported that about one third of individuals had difficulty on the emotional control subtest and half had clinical scores on the behavioral regulation index of the Behavior Rating Inventory of Executive Function (BRIEF)
- 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.

Interventions/What Might Help?

Teach Feeling Vocabulary to Develop an Awareness of Emotions:

- Modeling: Show appropriate responses, mirror feelings
- Scaffolding: Break larger tasks down into smaller ones and gradually move from providing more assistance to less
- Role-playing: Act out situations and how to respond to them
- Use positive reinforcement by rewarding appropriate responses (examples: stickers, fun toys or activities)
- Repeat often when teaching
- Take advantage of opportunities to teach feelings when child is noticeably having an emotional response
- Use tools to assist in teaching feeling vocabulary, such as feelings charts with colors or faces representing feelings, or traffic light to indicate degree of emotion being felt
- Teach emotions in different environments and with different people

To Help Before or During an Emotional Response:

- Recognize triggers that produce emotions and methods for avoiding those situations or limiting their emotional effect
- Try to keep a consistent routine/environment
- Reward positive, appropriate behaviors in new environments
- Work to maintain a secure, stable relationship with the child
- Calming techniques such as breathing techniques, having attachment items available, having a safe place to calm down, and exercise or mindfulness activities such as meditation, Tai Chi, or yoga

References

Gross, J.J. (1998). Antecedent- and response-focused emotion regulation: Divergent consequences for experience, expression, and physiology. *Journal of Personality and Social Psychology, 74*(1), 224-237.

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

Vohs, K.D., & Baumeister, R.F. (2011). *Handbook of self-regulation: research, theory, and applications second edition.* New York, NY: The Guilford Press.

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Fathers in CHARGE Syndrome: *Work and Friendships*

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The Invisible Parent

- Everybody knows a father
- Everybody knows that fathers are different than mothers
- Then why are they often assumed to be synonymous?

How are Fathers Different?

- Gender roles and expectations(Knox & Schacht, 2010)
  - Traditionally deliver discipline and life lessons
  - Spend more time with work than family
  - Often deal with school administrators, medical personnel, other professionals
  - Often do not deliver direct care

Fathers and Work

- May feel the need to prioritize work-related obligations over family obligations (Maume, 2006)
- May not want to jeopardize promotions and raises
- Spend more work-related time and nights away from home than mothers (Wyosocki & Gavin, 2003)

Fathers and Friendship

- Work life may translate into fewer male-male friendships (Vail-Smith, 2007)
- Activity-specific, less permanent friendships (Hartshorne, 1994)
- Side-to-side friendships (Hartshorne, 1994)
  - Less intimate than women's face-to-face friendships

Research Questions

- What are fathers reactions to the diagnosis of their child with CHARGE Syndrome?
- What impact has CHARGE syndrome had on their work life?
- What impact has CHARGE syndrome had on their friendships?

Diagnosis

- Aware of sperm mutation
  - Yes 33% No 67%
- Words to describe reaction to diagnosis
  - Shocked
  - Scared/Fearful
  - Anger
  - Sad
  - Concern
  - Love

Work

1=not at all; 7=very much

Sacrificed leisure time prior	3.82
Sacrifice leisure time now	3.74
Career central before	3.89
Career central now	2.77
Take allotted vacation	Some 51.2%; All 31%
Changed career/job	No 64.3%; Yes 31%
Father friendly employer	5.76
Change in importance	4.01

Friendships

1=not at all; 7=very much

Quality friendships prior	4.99
Quality friendships since	4.13
Quantity friendships prior	4.94
Quantity friendships since	4.02
Change in number	3.30
Importance of time spent	3.32
Desire for more friends	3.87
Miss quality from before	3.07

DADS

- Top six tasks performed
  - Make medical appointments
  - Recognize and respond appropriately to child's symptoms
  - Talk to health professionals about child's symptoms
  - Help others understand your child's medical condition and treatment
  - Exercise, play sports, leisure activities or supervise with your child
  - Attend support group or educational research about medical conditions
- Top five that made a difference
  - Take over other household tasks to give spouse more time
  - Pick up prescriptions
  - Administer medication to child
  - Recognize and respond appropriately to child's symptoms
  - Give up sleep if your child's medical condition requires it

Life Satisfaction and Meaning

- How happy taking all things together
  - Range: 1 = very happy; 4 = not at all happy
  - Average = 1.94
- How satisfied with life as a whole
  - Range: 1 = dissatisfied; 10 = satisfied
  - Average = 6.75
- How often think about meaning and purpose
  - Range: 1 = often; 4 = never
  - Average = 2.71

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National Consortium on Deaf-Blindness 2.0

Family Experiences With Intervener Services

Experiences with intervener services vary considerably from family to family. They encompass whether a child has intervener services, the process of determining a need for those services and obtaining them if appropriate, and perceptions of the usefulness of the services for a particular child. This document summarizes some of the information gathered from NCDB's survey of parents and guardians.

Some Details About the Parent/Guardian Survey

- 119 individuals completed the survey
- 81% have a child between the ages of 6 and 21
- 47% first learned about intervener services from their state deaf-blind project (the next most common source—22%—learned about these services from a parent organization)

The children of approximately half of the survey respondents either currently, or used to, have intervener services.

Child's Current Status Related to Interveners	Percent of Respondents
a. My child currently has an intervener	42%
b. My child used to have an intervener	6%
c. My child does not have an intervener but I would like him or her to have one	25%
d. I have attempted to acquire an intervener for my child, but have not been successful	7%
e. My child does not need an intervener	4%
f. My child has a one-on-one paraprofessional aide, but this person has not had training in deaf-blindness	14%
g. Don't know	3%

Additional Responses for Those Who Reported That Their Child Does Not Have Intervener Services (But They Would Like Him or Her to Have Them)

Thirty-three percent of parents or guardians who chose "c" or "d" above, responded to this follow-up question, "To what extent do you agree or disagree that the following factors are reasons that your child does not have an intervener?" Below is the percentage who "agreed" or "strongly agreed" with each statement.

Possible Factors Influencing Provision of Intervener Services	Percent of Respondents Who Agreed or Strongly Agreed
My child's school says my child does not need an intervener	44%
My child's school says they cannot pay for an intervener	47%
Educators at my child's school do not understand what an intervener does	64%
My child's school would provide an intervener for my child but say they cannot find a qualified person to take the job	12%

Relevant Comments:

- *I don't believe our school knows what our son's needs really are as a deaf-blind student, so they just scrape by day after day.*
- *They offer him a person who knows sign language but he is not with [her] all the time. She does not know anything about his visual impairment and doesn't care to be trained.*
- *We have identified a person who has expressed an interest/commitment to becoming our son's intervener. She does not currently have intervener training, but wants to begin the training as soon as she can. In our state, interveners are practically unheard of, so educating the school district has been the biggest hurdle.*
- *I really need assistance in my attempts to get an intervener in place for my 6-year-old son so that he, too, can reach his fullest potential.*

Results for Respondents Who Reported That Their Child Has (or Previously Had) Intervener Services

The information in this section comes from the parents and guardians who told us that their child currently has an intervener (50 children) or used to have an intervener (7 children). For 82%, provision of an intervener was written into the child's IEP. Although the parents/guardians who took this survey described variation in the level of difficulty they encountered when obtaining intervener services for their child, more than half reported they found the process easy.

Level of Difficulty	Percent of Respondents
Very difficult	15%
Difficult	27%
Easy	40%
Very easy	18%

Relevant Comments:

- *Easy at first, but once the agency realized that the intervener would naturally need to communicate with all members of the team the process became very, very difficult. Around IEP time, I always got sick.*
- *To get an intervener was not as hard as getting a "trained" intervener. Our school district has been wonderful and supportive; it is just that we could not find a trained intervener to fill the position.*

Respondents were asked about a variety of situations that might occur during interactions with school or school district personnel when intervener services are considered. The responses indicate that many parents/guardians who took the survey had positive interactions related to intervener services, but more than half perceived that the educational personnel they encountered had limited knowledge of deaf-blindness and intervener services.

Interactions With Educational Personnel	Percent of Respondents who Agreed With Statement
School or school district personnel said my child did not need an intervener	15%
School or school district personnel said that training in deaf-blindness was not necessary for paraprofessionals who work with children who are deaf-blind	15%
School or school district personnel said the school could not afford an intervener for my child	17%
My child's school had difficulty finding a qualified person to be an intervener for my child	37%
There were educators and/or administrators at my child's school who understood deaf-blindness	42%
There were educators and/or administrators at my child's school who understood what an intervener does	46%
There were educators and/or administrators at my child's school who were willing to learn about interveners	73%
There were educators and/or administrators who were open to trying an intervener for my child	65%
My state deaf-blind project provided ***information*** about interveners to personnel at my child's school or school district	65%
My state deaf-blind project provided ***training*** about interveners to personnel at my child's school or school district	58%

The next two tables show the respondents' impressions of their child's level of improvement in communication, learning, and quality of life after obtaining intervener services. The majority indicated there was a great deal of improvement in most areas.

Improvement in Communication Since Having Intervener Services	not at all	somewhat	a lot	don't know
Overall ability to express his or her wants, needs, and feelings	3.7%	33.3%	61.1%	1.9%
Ability to express himself or herself using a method or methods of communication (e.g., objects, pictures, gestures, signs, speech)	5.6%	37.0%	57.4%	0.0%
Ability to understand others' communication (e.g., objects, pictures, gestures, signs, speech)	1.9%	37.0%	61.1%	0.0%

Improvement in Other Areas	not at all	somewhat	a lot	don't know
Participation in classroom activities	0.0%	24.1%	68.5%	7.4%
Awareness of what is going on around him or her in the classroom	3.7%	24.1%	63.0%	9.3%
Exploration of environment	0.0%	31.5%	63.0%	5.6%
Independent movement	3.7%	35.2%	53.7%	7.4%
Relationships with peers	3.8%	30.2%	62.3%	3.8%
Behavior	3.8%	22.6%	69.8%	3.8%
Happiness at school	1.9%	11.1%	83.3%	3.7%
Participation in social activities	3.7%	20.4%	70.4%	5.6%
Involvement in community (e.g., volunteering, a job, participation in community recreational activities)	26.9%	21.2%	32.7%	19.2%

Relevant Comments:

- *My son has been able to be integrated in a special classroom in his neighborhood school due to an intervener. Otherwise the district would insist on a special school setting due to his level of disabilities. Without his intervener he would not be able to be maintained in his classroom environment.*
- *Keeping an intervener on my child's IEP has been a fight every year. So far, we have prevailed even though they tried to remove it by substituting an interpreter. Intervener is, however, still on the IEP. I believe an intervener is the difference between success and failure for deaf-blind children.*

The next three tables show respondents ratings of a number of miscellaneous issues or needs that are sometimes associated with intervener services. They include experiences with intervener services in schools, supports related to intervener services, and the importance of parent advocacy.

Experiences with Intervener Services	disagree or strongly disagree	neutral	agree or strongly agree	don't know
My child is still isolated because the intervener is the only person he or she interacts with at school.	83.6%	7.3%	3.6%	5.5%
My child's intervener has not received adequate training.	74.1%	11.1%	9.3%	5.6%

Importance of Additional Supports	not at all important	somewhat important	very important
Getting training for myself that is similar to the training interveners receive, so that I can better support my child at home.	1.9%	31.5%	61.1%
Opportunities to communicate directly with my child's intervener.	1.9%	13.0%	81.5%
Having an intervener at home, in addition to at school.	13.0%	33.3%	48.1%

Importance of Parent Advocacy	Percent of Respondents
Not at all important	7%
Somewhat important	30%
Very important	63%

Relevant Comments:

- *I'm speechless, it is overwhelming at how important the parent advocating is!*
- *The school knew what my child needed and provided from the beginning. We didn't know what an intervener did.*
- *I did not even know I needed one (way back in early childhood), so I'm glad it was all done for me.*
- *Children are now getting interveners without parents having to request - IEP teams are making the suggestions more and more. I have seen the evolution of understanding/acceptance in my state over the past 20 years.*
- *At different stages my advocacy has been important in keeping the intervener full time, in educating new team members, and in working with the team to learn to work with an intervener.*
- *D/B children live in a world of snap shots that are all out of order. Nothing makes sense. An intervener arranges those snap shots into an album, in order, and labeled so our children can take their time, process the information and move on to the next page at their own pace. These children will simply not succeed without one. There is not a standard at present to measure the difference an intervener makes, and that may very well be intentional. I have been astonished at the resistance to this service by educators who should know better. We have lost sight of the child in all the bureaucracy.*

For details about the process used to gather this information, go to
http://interveners.nationaldb.org/developmentProcess.php
For details about the Intervener Initiative Surveys, go to
http://www.nationaldb.org/documents/products/survey details.pdf

Balance Strategies for Children who have CHARGE Syndrome

The following activities are some strategies to help improve your child's balance. These strategies are separated by progressively increasing levels of difficulty. Only practice the activities that are appropriate for your child's capabilities. With time, your child may be able to complete the more challenging activities. Stay close to your child during these activities, so that you can assist him or her in case of a loss of balance. Some equipment that is helpful for multisensory balance training is an exercise ball and a compliant (less stable) surface, such as a BAPS board or soft exercise mat. These can be purchased at a sporting goods store or through the internet. You can purchase an Airex balance pad, through Amazon for ~ $60. You can even place several folded towels under your child's feet to make the surface of support less stable.

These exercises are designed to train each of the sensory systems that affect balance (somatosensory system, vestibular, and visual systems) to function more efficiently. The **somatosensory system** provides information on touch, pressure, and body position relative to the ground. The **vestibular system** provides information in regard to head position and movement. The **visual system** provides information in regard to the position of the body relative to the environment. The focus can be placed on the sensory systems by doing the following:

- Compromise or remove vision to improve function of the somatosensory system
- Compromise the somatosensory system (stand on a moving or compliant surface) to improve the function of the visual system
- Compromise both the somatosensory and the visual system to improve the function of the vestibular system by standing on a compliant surface and altering or removing vision.

These exercises are suggestions and can always be modified to fulfill the specific needs and capabilities of your child. Make the exercises fun by doing things that your child enjoys. Spread these exercises throughout the day, by incorporating them into your daily lifestyle, rather than spend a solid 30 minutes on the exercises.

Activities to help your child use their somatosensory system to control balance

Level 1 Seated Balance Activities

Exercise progressions:

a. Instruct your child to sit in a chair with no backrest with the arms in one of four positions (a. holding onto a support surface, resting on the thighs, folded across the chest, or extended out at sides) the feet are hip-width apart and in contact with a firm surface. Have your child maintain balance with vision reduced or absent for 30 seconds (blindfolds). **Instruct your child to focus on the pressure in the buttocks and feet.** Talk to your child during the activity to keep them interested. Sometimes even counting down or singing with your child can make the activity more interesting.

b. Instruct your child to close his/ her eyes, and lean the trunk forward, backward, lateral, and diagonally. To help your child do this you can use a soft stuffed animal and tell your child to lean in the direction of where they feel a tickle from their stuffed animal. Instruct your child to focus on feeling the pressure as its shift under the buttocks and from one region of the foot to another. Have your child progressively increase the angle of trunk lean so that the pressure under the buttocks and feet progresses from light to moderate to heavy.

c. Instruct your child to reach for objects of different sizes, weights, shapes, placed at various distances and heights relative to the body. You can blow bubbles and instruct your child to pop as many bubbles as he/she can while remaining seated. If your child has low vision, you can use a ball with bells. Instruct him/her to focus on using trunk movement. Progress to throwing objects to your child, varying the throwing heights, weights, and sizes of the objects. Start with larger, lighter balls and progress to smaller and heavier balls. Use bright balls or balls with bells if this works better for your child.

Level 2. Standing balance exercises

Exercise Progressions:

a. Complete all of the same tasks standing. If it benefits your child, you can have your child stand near a wall for stability or assist your child by holding his/her arm. As your child improves and becomes more confident he/she can progress to completing the activities without assistance.

b. If your child is doing very well with these activities, he/ she can progress to completing all of the same tasks with one foot. Be sure to spot your child during these activities.

Level 3. Moving exercises

Exercise Progressions

a. Instruct your child to walk across the room on a firm surface while reaching for objects that you are passing to him or her. You can use rings or koosh balls. You can walk alongside of your child and instruct your child to reach to the side. Be sure to practice this with both sides.

b. Instruct your child to walk across the room on a firm surface while throwing and catching an object with a partner. Begin with larger, lighter balls and progress to smaller, heavier balls. Some children may enjoy swinging a ribbon or yarn strips.

c. Instruct your child to walk across the room on a firm surface while wearing a blindfold. If it benefits your child, he/she can walk along a rope or a wall initially, and progress to walking independently. Your child can even

progress to walking over a rope, which can help him/her learn how to jumprope.

Activities to help your child use their visual system to control balance:

Level 1: Seated Exercises

Exercise Progressions:

a. While seated on an exercise ball (Note: the exercise ball should be of appropriate size for your child. Their knee should be at a 90º angle.) and feet on a compliant surface (BAPS Board or exercise mat). Instruct your child to look at something in front of him or her at eye level.
b. Repeat a) with only one foot on the ground
c. Instruct your child to look at the target while raising arms up and down and alternating feet on the ground. You can give your child ribbons to raise up and down. You can instruct your child to march and flap their arms like they are flying. Putting hula hoops on your child's arms or wrists may help your child continue this motion.

Level 2: Standing Exercises

a. Repeat all exercise progressions performed satisfactorily in a seated position while standing on a compliant or moving surface (BAPS Board or exercise mat)
b. Introduce selected weight shifts and transfer activities (leaning from one side to another, and forward and backward) while looking at an eye level target.

Level 3: Moving Exercises

a. Instruct your child to walk across a compliant surface (padded mat) while focusing on a visual target immediately in front of him/her.
b. Instruct your child to walk across a compliant surface (padded mat) with alternating on toes and heels while focusing on a target directly in front of him/ her.
c. Instruct your child to try this again only walking with heels and toes with one foot directly in front of the other (tandem walk). For heel walking, instruct your child to not let his/her toes touch the ground. For toe walking, instruct your child to not let his/her heel touch the ground. It may be easier for your child to understand this task by instructing them to walk along a line.

Activities to help your child use their vestibular system to control balance

Level 1: Seated Exercises

a. Instruct your child to sit on a balance ball with only one foot on the ground and the other foot raised. Place a blindfold on your child and instruct him or her to maintain the position for as long as he/she can. The time has ended if he/she moves the ball to the side or if his/her extended leg moves down. Progress to having your child do this activity with their other foot extended parallel to the other leg. To make this activity more interesting you could sing with your child or count together.
b. Instruct your child to sit on balance ball while wearing a blindfold and move his/her arms around and alternate his/her legs up and down. You could use stretching bands with their arms and instruct your child to stretch it up and

down while marching with his/her feet. Your child could also hold onto a hula hoop and move as if he/she was steering a car.

c. Instruct your child to sit on balance ball and pass a ball from one hand to the other. Tell your child to watch the ball move from side to side. If it benefits your child, you can use a ball on a string or place a ball in a sock to make catching less difficult. If your child can do this successfully, instruct him/her to gradually increase the height of the tosses. Ask your child, how high he/she can toss the ball. Children can also progress to throwing against a wall or tossing a ball back and forth with another person. They can also pass the ball in a figure 8 pattern (catch with left pass to right throw to other person's left, they pass to right, etc).

Level 2: Standing exercises

a. Repeat the same steps as in the seated exercises
b. While standing on a compliant surface and wearing a blindfold, instruct your child to begin marching in place with knees coming up. As they become comfortable with this activity, instruct him/her to lift his/her knees higher.

Level 3: Moving Exercises

a. Instruct your child to walk across the padded mat while wearing a blindfold. Your child can begin by walking while holding onto a rope, wall, or beside you, and progress to walking independently.
b. Instruct your child to walk forward across the mat on their toes while wearing a blindfold. Your child can begin by walking while holding onto a rope, wall, or beside you, and progress to walking independently.

*Remember, **you can modify any of these activities** to fit the specific needs, capabilities, and personality of your child. **Make these activities fun for your child.** For instance, some activities may be more fun with music. You can also turn these activities into games or be completed with friends or siblings. For the walking activities, you can design obstacle courses. If you make these activities fun and enjoyable, your child will be more interested in completing them and will benefit even more from them.*

Share these activities with your child's physical education teacher. Instruct them to integrate some of these activities into games for the participation of all children in the class.

Contact us or your child's physical therapist or physical education teacher if you have further questions.

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Behavioral Self Regulation in CHARGE

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What is Behavioral Self-Regulation?

Behavioral self-regulation involves engaging in intentional behavior and eventually moving to automatic regulation (Florez, 2011). To intentionally control behaviors, the individual must have an awareness of behavior and choose those behaviors most adaptive toward achieving a goal. Self-regulation of behavior is critical to development as it both inhibits and supports behaviors related to an individual's success socially, academically, and in everyday life (Galinsky, 2010).

Why is it hard for individuals with CHARGE to self-regulate their behaviors?

- Lack of executive functioning, communication skills, and sensory impairments (Nicholas, 2005).
- Central nervous system disturbances (Hartshorne et al., 2007; Nicholas 2005).
- Some challenging behaviors may be adaptive responses to over or under stimulation (Brown, 2005; Hartshorne et. al., 2005; Smith et. al., 2005). They may represent the child's "best" option for coping with stimuli in their environment.

Interventions:

- **Redirection of Behavior:** Instead of saying "no" or "stop that" teach the individual another way to act out their frustrations positively. Focus on positive behaviors rather than only pointing out the bad ones. For example: teach them to grab and squeeze a rubber ball instead of kicking and screaming when they are upset. It is also important to praise good behaviors in order to increase their likelihood.
- **Create a Barrier:** When exhibiting challenging behaviors create a physical barrier between the individual and the negatively arousing situation so they have time to calm down. For example, hold them tightly on your lap. Parents and caregivers should teach the child to remove themselves from a stressful situation so they are able to do so appropriately and independently.
- **Teach appropriate behaviors:** Help the individual to understand what behaviors are and are not appropriate and why. Ex. Modeling appropriate behaviors and using visual aids such as picture boards or puppets.
- **Scaffolding**: Start by boosting the individual's confidence by having them perform a task they can easily complete. Then model the correct way to do more complex tasks and use cues and hints to aid the individual in successful completion. Gradually reduce involvement so the individual can complete the task independently without eliciting any negative behaviors. Ex. Break the task into smaller parts, encourage verbal thinking processes, ask questions, coach them through the task.
- **Teach a new form of communication:** Often individuals with CHARGE have difficulty verbally expressing their wants and needs. Teach them a different way of communicating what they want that both of you can understand. Ex. a sign, gesture, picture, behavior.. When a caregiver responds to the individual, they feel understood and are more likely to exhibit positive behaviors.
- **Create Routines:** Create a specific daily routine that the individual can perform and become accustomed to. Make sure the routine is consistent because it can help reduce anxiety, as well as the intensity and frequency of challenging behaviors.
  - It is also important to reinforce flexibility in schedules and offer choices to increase independence. Ex. Offer different food choices for each meal or let them choose a new activity to do a certain day.
- **Prepare and Plan:** use verbal and visual prompts and reminders of events coming up that stray from the normal daily routine. Ex. Use a calendar, social story, picture board or hand gestures to repeatedly prepare the individual for a change in their routine.
- **Transition Training and Situation Sequencing:** Create lists of everything the child will do that day and present time slots for each activity. This can be done with a picture board, white board, hand gestures, or other visual aids. It is important for the individual to understand what they will be doing that day to reduce anxiety and stress. Ex. First Jani will eat lunch, second Jani will play at the park for 2 hours, third Jani will return home, etc.
- **Sensory breaks:** Schedule breaks throughout the day as needed so the individual can escape or avoid a stressful situation and have time alone in a calming environment. This is especially useful for days at school. Ex. Dimly lit environments, fluffy shaggy rugs, calming colors, etc. The environment should appeal to the individual's sensory needs. It is important to teach the child to request sensory breaks when they feel they need one, but be sure the child is not using the break to escape from performing a task and only using it to escape from being overwhelmed.
- **Sensory object:** Make an object readily available that they can access whenever they feel anxious, stressed, or frustrated. Sensory objects help relax and calm the individual and give them time to process. Common sensory objects include: squishy balls, soft blankets, weighted blankets, and brightly colored objects that uniquely appeal to the individual's sensory needs.

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Headaches in CHARGE

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CHARGE and Headaches

- Little is known about the experience of individuals with CHARGE Syndrome and Headaches.
- In a study of 30 participants with CHARGE, 8 reported experiencing Migraine, 26.6% (Blake & Salem-Hartshorne, 2005).
- In her study of the identification of pain in CHARGE Stratton reported that 15 of 61 parents indicated their child had migraines, 24.6% (Stratton, 2011).
- Reported incidence in general population varies but is about 7.7% (Abu-Arafeh, et al., 2010)

Types of Headaches

Migraine

5 attacks lasting 4-72 hours.

- With at least two of the following:
  - Unilateral Location
  - Pulsating Quality
  - Moderate/Severe Pain Intensity
  - Aggravation by Routine Physical Activity
- With at least one of the following:
  - Nausea and/or Vomiting
  - Photophobia and Phonophobia

(International Headache Society 2004)

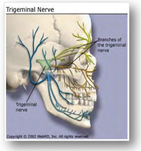
Chronic Tension-Type

Daily episodes lasting minutes to days.

- With at least two of the following:
  - Bilateral location
  - Pressing/tightening quality
  - Mild or moderately intensity
  - Not aggravated by routine physical activity
- Both of the following:
  - No moderate or severe nausea nor vomiting
  - No more than one photophobia or phonophobia

(International Headache Society 2004)

Cluster/Trigeminal Autonomic Cephalalgias:



Severe and unilateral. Lasting 15-180 minutes. One every other day up to 8 per day.

- With at least one of the following:
  - Ipsilateral conjuctival injection and/or lacrimation
  - Ipsilateral nasal congestion and/or rhinorrhoea
  - Ipsilateral eyelid oedema
  - Ipsilateral forehead and facial sweating
  - Ipsilateral miosis and/or ptosis
  - A sense of restlessness or agitation

(International Headache Society 2004)

Abdominal Migraine:

- Typically children ages 5 to 9
- Linked to adult migraines
- Lasts 1 to 72 hours
- Acute stomach pain with
  - Nausea
  - Vomiting
  - Light sensitivity
  - Diarrhea
  - Loss of appetite
- Related to adult migraines

Pain Assessment

- Pain is subjective making it hard to measure.
- Scales such as the Wong-Baker

FACES Pain Rating Scale have been used to measure pain.

- CHARGE Non-Vocal Pain Assessment
  - Based on non-vocal behaviors found to be indicative of pain in CHARGE (Stratton, 2011)

Quality of Life

- Adults who suffer from Chronic Migraines are 2x as likely to have anxiety, chronic pain, and depression (Lipton 2011).
- Women are more likely to experience Migraines or severe headaches (Smitherman et al 2013).
- Children with frequent severe headaches are more likely to have high levels of distress regarding emotional, conduct, inattention-hyperactivity and peer problems (Strine et al 2006)

Research Project

- Does the person with CHARGE have headaches?
- How do you know?
- How often?
- How severe?
- Impact on quality of life, learning, and behavior
- Medications used
- Are the medications helpful?
- What type of headache is it?

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Using the Holistic Communication Profile to Capture the Role of Pivotal Milestones in Language Development

Susan M. Bruce, Ph.D., Boston College

Holistic Communication Profile

The holistic communication profile displays the child's expressive and receptive characteristics, six closely related pivotal milestones, and play (Bruce, 2010a, 2010b).

What is a Pivotal Milestone?

- A milestone is considered "pivotal" if its achievement influences multiple areas of development.
- Some milestones are necessary to the achievement of language, such as object permanence. One cannot converse in speech, sign, or with a speech generating device about events in the past or people in another location without having object permanence.
- Direct instruction can accelerate mastery of pivotal milestones, having peripheral benefits to communication and language development.

Teaching Joint Attention

- Joint attention is shared between people.
- Joint attention includes sharing attention over an object.
- Know how long it takes the child to establish visual or tactual attention.
- Work on increasing joint attention in the context of preferred activities.
- Hand under hand exploration may support sustained joint attention.
- Introduce contact points, point approximations, and distal points as mechanisms for establishing joint attention.

Teaching Imitation

- Imitation is essential to efficient learning
- Deferred imitation means that the child can produce an imitation in a different time and place. This is a necessary condition to symbolic communication.
- Gain child's full attention prior to offering an imitation opportunity.
- Imitation may be grounded in vision, hearing, or touch.

Teaching Object Permanence

- Knowing that an object still exists when it cannot be seen, heard, or touched
- Closely related to person permanence
- The field of visual impairment uses the term "object concept" synonymously with object permanence.
- Visual tracking and visually directed reach & grasp are precursors to visually based object permanence.
- Initially create opportunities for child to observe you (visually or tactually) partially conceal an object.
- Later, fully conceal the object while the child watches.
- Auditory input is poor basis for object permanence. Think tactile.

Teaching Means-End

- Correlated with onset of intentional communication
- Provide opportunities for child to use objects as tools: in play and in daily living skills. For example, demonstrate how one object can be used to move another object.
- Simple switches are cause-effect tasks, but directional switches are means-end tasks.
- Provide problem solving opportunities- including posing physical obstacles-such as placing a preferred object just out of reach
- Provide opportunities for child to observe you problem solve

Teaching Discrimination & Categorization

- Discrimination is essential to differentiating one representation/symbol from another.
- Categorization is important to vocabulary development.
- Provide matching tasks: Give child one object at a time to potentially match to model. Emphasize vocabulary: "same" and "different."
- Later, provide sorting tasks Sorting involves the formation of groups. Provide two or more models. Provide child with objects to place in correct group. Later, provide the child with a pile of objects to sort according to the models.

Teaching Self-Recognition

- Regard self-recognition as part of the larger construct of individuation.
- Provide ample opportunities for child to view self in mirrors, paired with naming child and gesturing on child's body.
- Provide opportunities for child to view self while engaging with objects.

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Micro-enterprises - Building on Strengths and Promoting Self-Sufficiency: *Factors to Consider*

Seraphim Mork, Maria Alejandra Ramirez & Tim Hartshorne, Ph.D. • Psychology Department, Central Michigan University

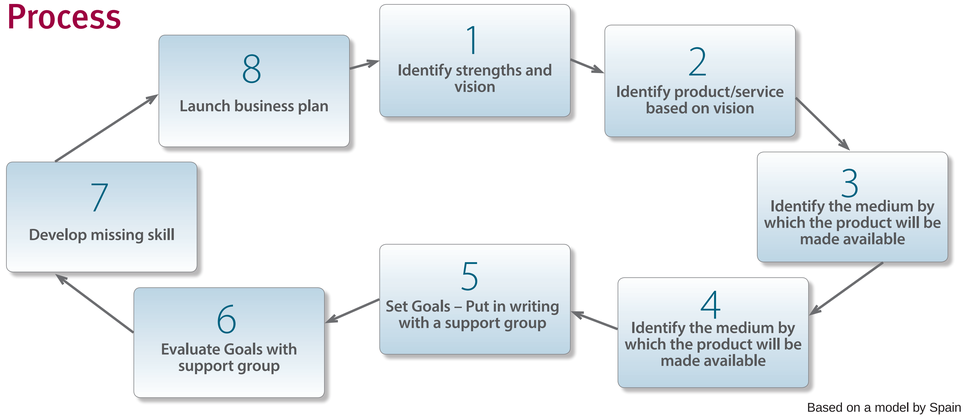
A matter of respect and dignity

- Young adults with disabilities need to develop a life after leaving K-12 school
- Most adults are engaged in some type of work
- Sheltered workshops provide work opportunities, but is the work meaningful, deserving of respect, or dignified?
- Starting one's own business is another option for individuals with disabilities

Micro-enterprises:

- Fewer than 5 employees; typically one owner, non-employer businesses with over $1,000 in gross receipts and subject to taxes according to the Small Business Administration
- Individuals with disabilities like other vulnerable populations can benefit from micro-enterprises.

Process



Based on a model by Spain

Law/Regulations:

Section 504 of Rehabilitation Act:

Any federal funding requires any agency receiving federal financial assistance to provide persons with disabilities an opportunity to be fully integrated into the mainstream

Americans with Disability Act

Public and private educational institutions, and service providers regardless of presence of any federal funding, are required to provide persons with disabilities an opportunity to be fully integrated into the mainstream

Social Security

Individuals with disabilities can get funds to start up business using funds such as Social Security Work Incentives a Plan for Achieving Self Support (PASS) and Supplemental Security Income (SSI)

Benefits

- increased range of choices
- equal or more income
- more time and engagement
- Increased integration
- enhanced self-esteem
- improved quality of life
- less costly than traditional models
- possibility of growth into serious income
- chance of creating passive income
- prospect of becoming an employer

(Conroy, Ferris, & Irvine, 2010).

Examples of products and services provided:

- Scented candles and products
- Shredding paper
- Baking goods
- Gardening
- Jewelry
- Painting

Barriers

- Social stereotypes about people with disabilities
- Limited information about starting up business for individuals with disabilities
- Lack of choices or options
- Low expectations of success

(Conroy, Ferris, & Irvine, 2010).

Resources

Vocational rehabilitation and workforce investment act agencies:

Social security administration

Advocacy center for persons with disabilility:

www.advocacycenter.org

Network180-offers resources for creating microenterprises for persons with disabilities:

http://www.network180.org

Small Business Administration:

http://www.sba.gov

Self-Employment for Individuals with Disabilities -- Online Seminar with Cary Griffin

http://www.ttap.org

Small Business and Self-Employment for People with Disabilities -- Office of Disability

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Socialization in Different Age Dyads of Students with CHARGE Syndrome

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Impact of Deafblindness

- Vision and hearing provide us with information that supports social skills development & interactions (Kekelis, 1992).
- Children who are deafblind will require direct instruction on social skills due to limited opportunities to observe.

Action Research

- Recursive, problem solving form of research usually involving multiple cycles of action & reflection (Bruce & Pine, 2012)
- Types: Classroom, Collaborative, Critical, and Participatory (Hendricks, 2012)

Self-Determination

- Being a causal agent in one's own life to improve quality of life (Wehmeyer, 2005)
- Components: Self-awareness, problem solving, goal setting, decision-making, choice-making, self-regulation, self advocacy, and self-efficacy (Wood, Fowler, Uphold & Test, 2005)

Intervention & Data Sources

Intervention

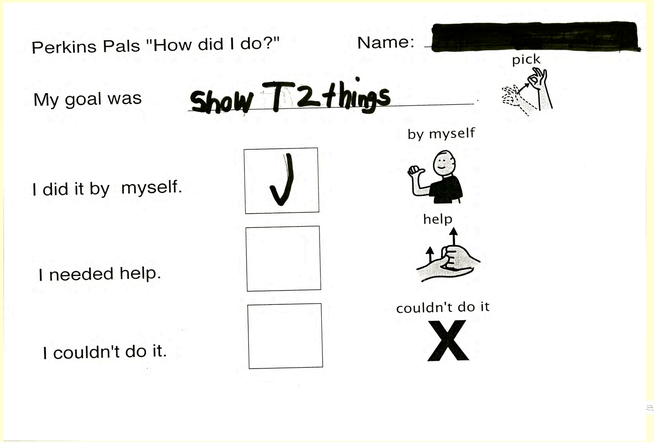
- *Six interaction sessions in four interaction spaces: books, music, pretend play, & cars/trucks*
  - Younger students viewed interactions as play
  - Older students also engaged in self-evaluation and problem solving, which are components of self-determination
- *Six secondary student reflection/evaluation sessions*
  - Secondary students viewed videos of their interactions with younger students
  - Secondary students evaluated their own performance
  - Secondary students set interaction goals for the next interaction session

Data Sources

- Observation videos of interaction sessions (younger and older students)
- Observation videos of secondary feedback sessions (only secondary students)
- Student Documents: Secondary student evaluation & goal form and self-evaluation
- Teacher Document: Teacher evaluation form of secondary students

Evaluation Forms

Student Self-Evaluation



Teacher Evaluation Form

	0	1	2
Greeting	- No greeting	- Staff prompted greeting	- Independent greeting
Conversation	- No conversation	- Staff assisted with conversation	- Independent conversation
Closing	- No closing	- Staff assisted with closing	- Independent closing

Perkins CHARGE Pals Staff-Evaluation Rubric

Findings/Strategies

Joint Attention Strategies

- Gain attention of younger partner before communicating
  - Try tapping shoulder
  - Try saying/signing pals' name
- Wait for pal's visual attention & know how long to wait
- Take turns & defer to wishes of younger pal
- Less movement helps with joint attention

Improved Play Strategies

- Greet pal
- Watch younger pal to determine his preferences
- Select interaction toys the younger pal will like
- Bring preferred toys to younger pal
- Follow the younger pal's lead-build on what he does
- Being seated sometimes helps

Other Findings/Strategies

- Physically arrange room so that pals don't easily gain "rescue" from adults
- While the goal is for the older pals to problem solve, on occasion they need adult support during the interaction sessions
- Secondary students' self-evaluations became more accurate over time

Research Methods

Collaborative Action Research, Collective Case Study Design

Sample Research Questions:

- How will interactions between older students with CHARGE syndrome and younger students with mixed etiologies change, over time, in the context of an engineered interaction space?
- How will younger students change their interactions with objects over time?
- What socialization goals will older students select?
- How will a self-evaluation procedure impact socialization goals established by older students with CHARGE syndrome?
- What strategies will teachers suggest when discussing the dyads during the feedback sessions with the older students?

Participants

Dyad #1: *Secondary Student W*: CHARGE syndrome, profound deafness; 20/40 left eye, 20/150 right eye; linguistic (sign language and pictures)
Elementary Student I: Leber's Congenital Amaurosis & Cystic Fibrosis; moderate hearing loss left ear, moderate-severe in right ear; 20/400 & nystagmus; linguistic (speech)

Dyad #2: *Secondary Student MM*: CHARGE syndrome; 75-85 db loss (unaided): 20/20 corrected; linguistic (speech & Signs)
Elementary Student T: Brain stem & cerebellar dysgenesis; moderate-severe bilateral hearing loss; 20/94; pre-linguistic (few signs, gestures, pictures)

Dyad #3: *Secondary Student MS*: CHARGE syndrome; profound loss in left left ear, 15 db loss in right ear; 20/100; linguistic (speech & some signs)
Elementary Student D: CHARGE syndome; moderate-severe hearing loss; 20/94; pre-linguistic (few signs, pictures, line drawings, body language)

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National Consortium on Deaf-Blindness *

Recommendations for Improving Intervener Services

GOAL 1: RECOGNITION

Increase recognition and appropriate use of intervener services for children and youth who are deaf-blind.

RECOMMENDATION 1

Develop a coordinated and expanded national approach to provide state and local early intervention and education agencies with information and tools needed to understand and use intervener services.

Implementation Strategies

- The National Consortium on Deaf-Blindness (NCDB), state deaf-blind projects, and other stakeholders (e.g., families, early interventionists, teachers, related service providers, early intervention and educational administrators, interveners, and university faculty), will join forces to implement a comprehensive national intervener initiative. The initiative, coordinated by NCDB, will:
  - Develop and disseminate a consistently applied *national definition* of intervener services, including clarification of the occupational role of the intervener.
  - Organize workgroups to implement the recommendations in goals 1 through 3 and identify additional needs and recommendations to improve intervener services. Workgroup topics will include, at a minimum:
    - intervener preparation and training;
    - continuing education needs of interveners;
    - coaching and supervision of interveners;
    - credentialing or certification of interveners;
    - interveners in community and home settings; and,

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        - interveners for infants and toddlers.
    - Create a Web-based platform on which state deaf-blind projects, NCDB, families, and other organizations and individuals can interact and share knowledge. For example, this platform could be used to:
        - communicate ideas and concerns;
        - highlight intervener training and support models; and
        - access a shared video library related to intervener services (e.g., parent and professional insights, examples of interveners working with children).
    - Identify and implement strategies to increase collaborative efforts between agencies and organizations within individual states (e.g., state deaf-blind projects, PTIs, family organizations) to improve intervener services at the state level.
- Develop and make available a core set of publications that increase understanding of intervener services and promote their development and use. These materials should, at a minimum, include:
    - concise fact sheets that a) promote an enhanced understanding of intervener services, and b) explain the occupational role of a well-trained intervener;
    - publications that highlight promising intervener-training and support programs and provide strategies that describe how they can be replicated; and
    - publications that describe effective practices for intervener services.
- Design and launch a national data collection program to collect, compile, and make available data about the use of intervener services, such as:
    - characteristics of interveners (e.g., how many, where, education level), and
    - characteristics of children and youth who receive intervener services.

RECOMMENDATION 2

Coordinate and expand efforts to inform and influence national, state, and local policies and practices so that they reflect and support the provision of intervener services for a child or youth who is deaf-blind when needed.

Implementation Strategies

- Produce and disseminate guidelines that IFSP/IEP teams can use to make informed decisions about the need for initial or continued use of intervener services for an individual child or youth.
- Using the core products described in Recommendation 1:
  - promote best practices for intervener services via information dissemination and technical assistance activities; and,
  - systematically disseminate resources to lawmakers and other policymakers to inform and influence policies related to intervener services.
- Work with OSEP to encourage U.S. Department of Education cross-agency (e.g., OSEP, Rehabilitation Services Administration, National Institute on Disability and Rehabilitation Research) recognition of intervener services.
- Work with state and national special education organizations and centers (e.g., Regional Resource Centers, the National Association of State Directors of Special Education, Parent Training and Information Centers) to design and implement strategies that inform and influence policies and practices related to intervener services.
- Work with state special education advisory councils to raise individual states' awareness of intervener services.
- Contribute to the growth of knowledge related to intervener services in the following ways:
  - develop professional publications including technical reports or peer-reviewed journal articles that summarize available data about interveners and describe the history and current status of intervener services in the U.S.;
  - promote research on intervener services by:

- facilitating discussions among graduate students and researchers within the field of deaf-blindness;
- assisting researchers in identifying children and families who can participate in research studies;
- providing library support (e.g., literature searching) to researchers working in this area; and
- identifying possible funding sources for intervener services research.

GOAL 2: TRAINING & SUPPORT

Establish a strong national foundation for intervener training and workplace supports.

RECOMMENDATION 3

Develop national open-access intervener-training materials that align with the Council for Exceptional Children's *Specialization Knowledge and Skill Set for Paraeducators Who Are Interveners for Individuals with Deaf-blindness*.

Implementation Strategies

- ❖ Establish a workgroup of individuals with expertise in intervener training to collaborate with NCDB on the development of intervener-training materials.
- ❖ Invite professionals from the field of deaf-blindness to submit intervener or general deaf-blind education training materials for review by the workgroup and possible incorporation into the materials.
- ❖ Develop the training using new and existing materials.
- ❖ Create a web-based platform to house and provide free access to the materials.

RECOMMENDATION 4

Develop strategies to ensure that interveners have knowledgeable supervisors and access to experts in deaf-blindness who can provide consultation and coaching.

Implementation Strategies

- Use the intervener-training curriculum described in Recommendation 3 to train teachers and other team members about deaf-blindness and the role of the intervener.
- Identify successful models used by state deaf-blind projects, university programs, and school districts that provide on-the-job support to interveners.
- Replicate these models to support an increasing number of interveners.
- In partnership with a broad group of stakeholders, examine the causes of the shortage of local experts in deaf-blindness, including teachers of the deaf-blind, and identify strategies to alleviate the shortage.
- Design and implement strategies to provide distance consultation, coaching, and mentoring through the use of technology applications.

RECOMMENDATION 5

Expand opportunities for interveners to obtain a state or national certificate or credential.

Implementation Strategies

- Invite input from a broad group of stakeholders regarding:
  - preferred characteristics of credentialing processes and criteria;
  - current and future needs for an intervener credential; and,
  - short- and long-term goals of intervener credentialing.
- In partnership with stakeholders, including the NRCP and the National Intervener Task Force, determine the necessary criteria for an intervener credential.
- Identify credentialing bodies that could offer a national credential that meets those criteria.
- Determine the most feasible credentialing options and move forward with efforts to expand pathways to a national credential that are applicable to interveners with a variety of training backgrounds.

RECOMMENDATION 6

Establish a national intervener jobs clearinghouse to assist in intervener recruitment and job placement.

Implementation Strategies

- Convene a workgroup of interveners and other individuals who have knowledge of intervener hiring practices (e.g., educational administrators, state deaf-blind project personnel) to determine the design elements needed for an online jobs clearinghouse.
- Develop a secure online jobs clearinghouse reflecting those design elements.
- Publicize the availability of the clearinghouse through current intervener training programs, state deaf-blind projects, and other relevant agencies and organizations.
- Maintain the clearinghouse data on an ongoing basis.

RECOMMENDATION 7

Provide resources (e.g., technology applications, technical assistance) that assist interveners to establish organized online and face-to-face communities where they can improve their knowledge and skills by sharing ideas and experiences with each other.

Implementation Strategies

- Convene a workgroup of interveners, state deaf-blind project personnel, and university faculty to determine desired features of an online community of interveners.
- Develop and maintain a Web-based platform providing those features.
- Publicize the availability of the site and train interveners in its use.
- Explore opportunities for interveners to occasionally meet in person (e.g., state meetings, national or regional conferences).

GOAL 3: FAMILIES

Build the capacity of families to participate in decision about intervener services for their children and in efforts to improve these services.

RECOMMENDATION 8

Develop and disseminate information resources and tools to family members that increase their knowledge of intervener services and enhance their ability to communicate effectively with educators, administrators, and others about those services.

Implementation Strategies

- Review existing family-focused resources related to intervener services.
- Use existing resources (if available) or develop new products that families can use to:
  - promote communication about intervener services with early interventionists, educators, and administrators;
  - inform decisions related to intervener services for their child; and
  - inform and influence state and local policies to encourage and promote high-quality intervener services.
- Collaborate with family organizations to distribute information to families who have limited knowledge of interveners. This will include efforts to reach out to groups who are typically underrepresented (e.g., racial and ethnic minorities, families who live in rural areas, and families who are socioeconomically disadvantaged).

RECOMMENDATION 9

Develop and implement strategies that create opportunities for families to share ideas and experiences and work together to impact intervener services at local, state, and national levels.

Implementation Strategies

- Establish accessible Web-based and/or telephone groups where family members of children who are deaf-blind can share ideas and experiences about intervener services.
- Partner with key family organizations (e.g., NFADB, NDBII Parent Group) to implement strategies for recommendations that promote appropriate effective intervener services for children who are deaf-blind.
- Develop a curriculum module about intervener services to supplement current family leadership curricula that family members can use to educate themselves and to mentor others.

GOAL 4: SUSTAINABILITY

Sustain high-quality intervener services across the nation through the inclusion of intervener services in national special education policy.

Recommendation 10

Congress should ensure the long-term sustainability of intervener services for children and youth who are deaf-blind by including "intervener services" as a related service and as an early intervention service in the next reauthorization of the Individuals with Disabilities Education Act (IDEA).

Physiological Self-Regulation in CHARGE

Andrea Larsen and Timothy Hartshorne • Central Michigan University

Physiological Self-Regulation

The ability to self-regulate can be regarded as the strategies one attains to monitor and adjust their behavior in response to the cognitive, emotional and social demands encountered through daily living. Physiological variables previously used in assessing self-regulatory capacities incorporate cardiac functioning; which includes the parasympathetic and sympathetic divisions of the autonomic nervous system and adrenocortical activity; or the responsiveness of the stress hormones. Dysfunction in these areas can inhibit one's ability to obtain optimal levels of arousal and effectively utilize self-regulation.

EX: Children with CHARGE syndrome are placed at a higher risk for experiencing stress. The constant challenges posed by CHARGE can cause an increase in the sympathetic nervous system response and trigger the release of the body's stress hormones. If these physiological responses persist, children with CHARGE may have difficulties employing self-regulation and displaying accurate states of arousal.

Sensory Stimulation

The first means of self-regulatory development is based on sensory input. Each child with CHARGE possesses unique sensory needs. Occupational therapists can help create a "sensory diet" tailored towards each individual's needs. Integrating items off of the prepared "sensory menu" can help children in executing daily chores and tasks.

Sensory Activities

Allow for intermittent daily activities that promote sensory stimulation. Sensory activities can be obtained from a devised "sensory menu."

EX: Playing on a trampoline or in a sandbox, teaching basic yoga poses or exercises, and the application of a weighted vest.

Sensory Environment

Modifications can be made to the child's everyday home and school environments to aid with their sensory needs.

EX: Provide modified chairs, stabilizing items with a dycem, the use of beanbag chairs, and creating areas free of breakable items that allow for rough house play.

Relaxation Therapy

The promotion of relaxation can help diminish stress and reduce sympathetic nervous system activity, also referred to as the body's "flight or fight" response.

EX: Increase blood flow to the extremities by practicing hand warming techniques and helping to reduce tension on muscles by practicing simple stretches and yoga poses.

Wilbarger Protocol

The Wilbarger protocol is an integrative approach that includes both sensory stimulation and relaxation therapy. This program can aid children in reducing stress and help in obtaining optimal levels of arousal.

EX: First apply deep pressure utilizing a specialized non-scratch brush, followed by compression to each of the major joints. Treatments must be followed on a routine basis for benefits to become observable.

Diet

Self-regulatory capacities can be influenced and fluctuate based upon one's blood glucose levels. Maintaining glucose will ensure that the brain has the energy it needs to employ self-control. A balanced diet and adequate fluid intake help to preserve one's blood glucose levels.

EX: Offer several smaller meals throughout the day that incorporate a sufficient amount of carbohydrates. Foods lower on the glycaemic index are most preferred which can include fruits, vegetables and whole grains.

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Addressing Sensory/Oral Placement/Feeding Difficulties Associated with CHARGE Syndrome

TALKTOOLS®

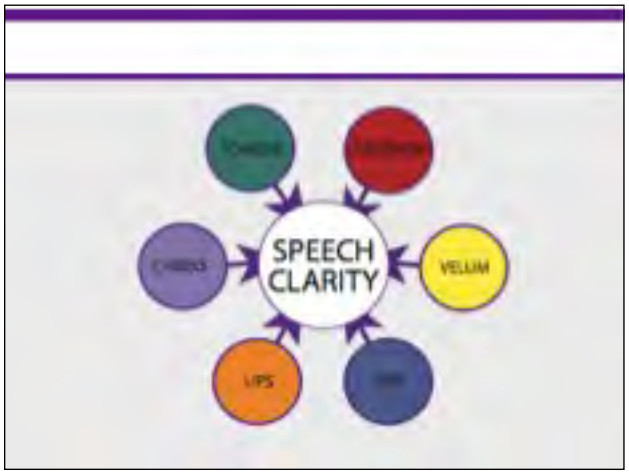
DISCOVER THE FEEL OF FEEDING AND SPEECH

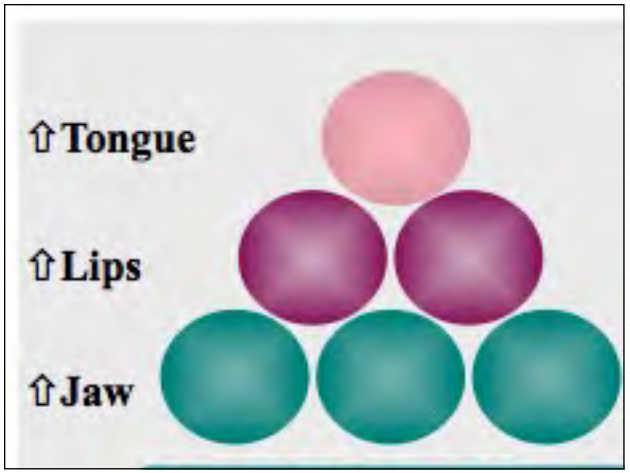
Presented by Whitney B. Rotterdam
MA, CCC-SLP

The Feel of Speech

What is OPT vs Traditional Speech Therapy:

Traditional speech therapy presents information through visual and auditory stimulation. However, many individuals have difficulty learning through their eyes and their ears. OPT adds the "feel" of speech.





Common Deficits In CHARGE Syndrome

Deficits which effect sensory, feeding and speech clarity; each of these deficits may range from non-existent to severe.

1. Hypotonicity
2. Visual Impairment
3. Hearing loss-ear infections
4. Sensory deficits may range from minor to severe: smell, taste, touch
5. Oral tactile sensitivity: hyposensitivity, hypersensitivity, mixed sensitivity fluctuating sensitivity. The "feel" of feeding and the "feel" of speech
6. Tactile defensiveness
7. Cognitive deficits

Common Deficits In CHARGE Syndrome

Deficits which effect sensory, feeding and speech clarity; each of these deficits may range from non-existent to severe.

8. Gap between expressive and receptive language skills
9. Weak jaw musculature: Symmetrical or Asymmetrical
10. Incomplete lip closure, decreased tongue mobility/grading results in limited retraction, lateralization, and tongue-lip pointing
11. Weakness is the muscles of the velum
12. Blocked nasal passages: /m, n, ng/
13. Motor planning deficits
14. Difficulty coordinating oral airflow with vocalizations to initiate speech sounds production

Goals of Oral Placement/Feeding Therapy

- To improve feeding skills and nutritional intake
- To improve speech sound production and improve intelligibility/clarity

Goals of Oral Placement/Feeding Therapy

- To increase awareness of the oral mechanism
- To normalize oral tactile sensitivity
- To teach more normal movement patterns
- To increase differentiation of oral movements

a. Dissociation: The separation of movement, based on stability and strength, in one or more muscle groups.
b. Grading: The controlled segmentation of movement through space based on dissociation.
c. Fixing: An abnormal posture used to compensate for reduced stability which inhibits mobility.

Some Statements to Consider

- "We do not monitor our speech clarity by how it 'sounds' or how it 'looks.' Instead we base our assumption that we are speaking intelligibly on how it 'feels.'"
- "Why does my child bite and put everything in his/her mouth but avoids certain foods?"
- "Why does my child grind his teeth, suck his thumb, etc.? How can we eliminate these behaviors?"

The answer is SENSORY!

So how do we get started??

Muscle-Based Exercises(Oral-Motor)

Begin with a sensory program and a stable posture:

Work from whole body to mouth to achieve acceptance of touch and to develop trust.

Before you Begin to work on feeding or speech:

1. Establish a supported feeding position:

Stability in the body allows for mobility within the oral cavity

Before you Begin to work on feeding or speech:

2. Evaluate the Sensory System:

a. Toothette w/Vibrator
b. Sensory Bean Bags
c. Jigglers

Sensory Diagnosis/Tactile System

Tactile Hyposensitivity:
An under-reaction to tactile input.

Sensory Diagnosis/Tactile System

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Mixed Sensitivity:
Any combination of hyper, hypo or normal sensitivity.

Sensory Diagnosis/Tactile System

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Fluctuating Tactile Sensitivity:
Responses that change over time.

Sensory Diagnosis/Tactile System

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An over-reaction to tactile input

Mixed Sensitivity:
Any combination of hyper, hypo or normal sensitivity

Fluctuating Tactile Sensitivity:
Responses that change over time.

Tactile Defensiveness:
A learned tendency to respond negatively or emotionally to tactile input

Before you begin to work on either feeding or speech:

1. Evaluate the Sensory System and make diagnosis
2. Eliminate tactile defensive behaviors by establishing trust.
3. Treating Sensory Deficits: Sensory Integration Deficits: Begin with a sensory warm-up as prescribed by an S.I. trained Occupational Therapist

Before you begin to work on either feeding or speech

Oral Placement (Muscle-Based) Activities:

1. Speech: Use non-food items to increase strength and stability in the muscles of the abdomen, velum, jaw, lips and tongue as a prerequisite for the emergence of standard speech sound production

2. Feeding: Teach these movements prior to introduction of foods to improve bolus control, confidence in oral feedings and feeding safety.

Before you begin to work on either feeding or speech:

Why is feeding so important:

*Nutritional Concerns

*"The muscles that are used in feeding are the same muscles used in speech"

*How will Oral Placement Therapy help my child to speak more clearly?

Before you begin to work on either feeding or speech:

Exercises for tube-fed children:

1. Associates movements in the mouth with feeding

2. Exercises in the mouth: The muscles that are used in feeding are the same muscles that are used in speech

3. When the child receives medical approval for oral feedings they will be ready to eat both from a sensory position and a strength position. Working on oral-phase feeding exercises improves swallowing proficiency

Before you begin to work on either feeding or speech:

Preventing tactile defensive behavior patterns associated with feeding:
These exercises will also improve jaw strength and teach the motor plan for chewing.

Exercises for infants without teeth:

1. Gloved finger
2. Infadent

Muscle-Based Exercises (Oral-Motor)

2. Jaw Exercises:
   a. Chewing on Back Molars
   b. ARK's Z-Vibe or Probe
   c. Chewy Tubes (Red – Yellow)
   d. ARK's Grabbers (Purple – Green)
   e. TalkTools® Jaw Grading Bite Blocks

Overview of Optimal Feeding Positions and Techniques

1. **Purees**: Spoon Feeding

a) Placement of the spoon: Side, Front or pointed tip at lip midline?

b) Wait for your child to close his/her lips before you remove the spoon

Overview of Optimal Feeding Positions and Techniques

2. Liquids

a) Cup Drinking: Sippy- cups, are they right for my child? What are the alternatives

b) Straw Drinking: Why is the Straw Hierarchy so important?

Straw #1

Overview of Optimal Feeding Positions and Techniques

3. Solids

a) Cube or julienne stick shape?

b) Why is it so important that my child learn to chew on his/her back molars?

The Need for Practice

*OPT activities must be practiced a minimum of 3 times per week

*Once the skill is mastered it must be transitioned into function

*OPT and traditional speech and language work together

*Have fun and enjoy the successes!

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FOSTERING SELF-REGULATORY STRATEGIES IN CHILDREN WITH CHARGE SYNDROME

Maria Alejandra Ramirez
School Psychology Doctoral Student
Central Michigan University

Agenda

1. Overview of 4 areas/windows of Self-Regulation
2. Example of Self-Regulation
3. What we know about Self-Regulation in children with CHARGE syndrome
4. Using the Windows of Self-Regulation to *identify strengths* and *identify difficulties*
5. Using the window of self-regulation to *prioritize areas to target*
6. In Detail: Specific strategies to foster each window of self-regulation

Discussion Goal 1:

Quick Overview Self-Regulation and the 4 Windows of Self-Regulation

What is Self-Regulation?

- Adaptive and flexible management of four domains.
  - Physiological, behavioral, cognitive, and emotion
- In order to describe regulation as "self-regulation" two components are necessary:
  - 1) self-awareness of the process
  - 2) a subsequent goal-directed action.
- Self-regulation must begin 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 stimuli and responding appropriately under environmental expectations.

Self-Regulation:

→ **Goal** →

What are the 4 Windows of Self-Regulation?

Physiological

Behavior

Cognitive

Emotion

What is Physiological Self-Regulation?

Physiological

- Self's ability to react and alter its own states and responses to meet the needs of the body.
- In great part under the control of the somatic, endocrine and autonomic nervous systems.

What is Behavioral Self-Regulation?

Behavior

- Awareness of a behavior
- And choosing those behaviors most adaptive toward achieving a goal.
- Goal directed and purposeful behavioral patterns consisting of:
  - one's ability to inhibit,
  - regulate, pace, and delay gratification
    - Jahromi and Stifter (2008)

What is Cognitive Self-Regulation?

Cognitive

- Voluntarily regulating thoughts and mental processes by balancing inhibition and initiation of behavior in order to achieve a goal
- Involves
  - Attention
  - Memory
  - Learning (as well as using prior learning)
  - Reasoning
  - Problem-solving
  - Decision-making
  - Metacognition

What is Emotion Self-Regulation?

Emotion

- Using emotion regulation, the individual analyzes, controls, alters, or prevents behaviors related to the adaptive expression of emotions
  - (Lam & Lewis, 2010).
- May occur at different points in time during the emotional response

Discussion Goal 2:

Example of Self-Regulation

Example: Dentist Appointment

- **Physiological:**
  - You make sure to brush and floss your teeth really well a couple days before the appointment
- **Behavioral:**
  - You make sure to put the appointment time/day in your planner
  - You get a babysitter
- **Cognitive:**
  - You mentally prepare yourself for the procedure
- **Emotion:**
  - You tell yourself the pain will be over soon
  - You ask for a lot of pain medicine to make sure you feel no pain

Discussion Goal 3:

What we know about Self-Regulation in children with CHARGE syndrome

What do we know about Self-Regulation in CHARGE?

- Multi-sensory difficulties may limit exposure to environmental stimuli, their exposure to interactions and reactions to the environment.
- As DeGangi (2000) states, early deficiencies in self-regulation may lead to challenging behavior, and deficits in attention and inhibition.

What do we know about Self-Regulation in CHARGE?

- **Physiological:**
  - Brown (2005) notes that individuals with CHARGE syndrome are truly multi-sensory impaired, often having challenges with vision, hearing, balance, touch, temperature, pain, pressure, smell, breathing, swallowing, eating, drinking, digestion, and temperature control

What do we know about Self-Regulation in CHARGE?

- **Behavior:**
  - May display behaviors typical of individuals with: Autism Spectrum Disorder, ADHD, OCD, Tourette's syndrome, and Deaf Blindness (Hartshorne & Cypher ,2004).
  - These may include: restricted range of interest, stereotyped movements, fidgeting with objects, preference for certain objects or people, tactile defensiveness, staring at lights, vocal stimulation.

What do we know about Self-Regulation in CHARGE?

- **Cognitive:**
  - Children with CHARGE may present with executive *dys*function.
  - Specifically in the areas of *shifting, monitoring, and inhibiting.*
    - Hartshorne, Nicholas, Grialou, and Russ (2007)

What do we know about Self-Regulation in CHARGE?

- **Emotion:**
  - This area is much less explored than the other areas of self-regulation
  - Given that children with CHARGE have difficulty in the other areas of self-regulation and all the areas are related….it is possible that this may also be an area of difficulty.

Discussion Goal 4:

Using the Windows of Self-Regulation to:

1. *identify strengths*
2. *identify difficulties*

Identifying Regulatory Strengths/Difficulties in YOURSELF

Physiological

Behavior

Cognitive

Emotion

Identifying Regulatory Strengths/Difficulties in YOUR CHILD

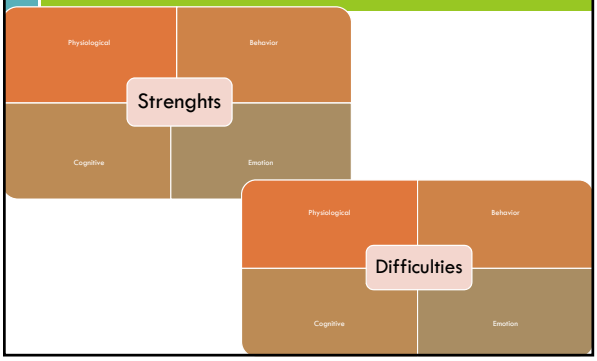
Physiological

Behavior

Cognitive

Emotion

Complete handouts for yourself and child



Discussion Goal 5:

Using the Windows of Self-Regulation to

Prioritize areas to target

Prioritizing Areas to Target

Things to consider:

- What would increase the most function?
- What would increase quality of life?
- What would save time?
- What would save resources?
- Do I have time to target this area?
- Do I have the resources to target this area?
- Are the final outcomes worth it?

Example:

Discussion Goal 6:

Specific Regulatory Strategies to Foster Each Window

Strategies: Behavior

Factors to consider:

- ID problematic Behavior
- What preceded the behavior?
- What follows the behavior?
- Who is present when the behavior occurs?
- Where does the behavior take place?

Strategies: Behavior

- Modeling, Role-playing, and Reinforcement
- Predictable routines
- Visual Schedule
- Increasing choices
- Pre-correction
- Red Dot Timer
- First-Then Statements
- Scaffolding
- Requesting breaks with visuals
- Pressure vests and deep tissue massages

Strategies: Physiological

- Sensory stimulation
- Environmental modifications
- Relaxation Therapies
- Diet
- Feeding
- Toileting
- Sleep issues

Strategies: Cognitive

- Modeling thinking, planning, and inhibitory strategies
- Modification of stressful environments
- Preparation to enter stressful environments
- Use of mind-body and technology practices to foster concentration and inhibition
- Take advantage of the child's favorite activities to imbed teaching of waiting and engaging.

Strategies: Emotion

- To develop awareness of emotion: teaching feeling vocabulary (modeling, scaffolding, and reinforcement)
- Mirroring feeling and modeling appropriate emotional responses
  - "I am happy, this is what happy looks like"
  - "You look upset, like this"
- Repetition and rehearsal of skills
- Use of visuals: colors, faces, traffic lights – to represent feeling or state
- Recognizing triggers that produce emotional response
- Teaching strategies to deal with emotions (deep breathing, location to calm down, attachment objects, etc.)
- Using all daily events to teach feelings, reactions, and modeling appropriate ways of coping

Strategies: Emotion

Remember to:

- Teach in different environments (home, school, grocery store)
- Teach with different people (peers, parents, siblings, etc.)
- Reinforce all appropriate behaviors in new environments

Thank you for your time!!!

If you have questions, please contact me at:

ramir1ma@cmich.edu

They're Not Children Anymore:

communicating effectively with adult providers

James Schwark RN, MSN, MDiv, CRRN
Simulation and Skills Coordinator
Azusa Pacific University
School of Nursing
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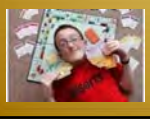
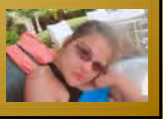




objectives

- Discuss the different perspective adult practitioners possess in contrast to pediatric practitioners.
- Identify the knowledge and expertise contained in families who live with CHARGE.
- Encourage a willingness to ask for clarification and explanation.
- Propose methods of communication which promote hearing.
- Discuss the need for persistence.
- Encourage a willingness to seek providers who are willing to learn.

The Charge of the Light Brigade

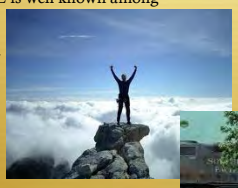
Half a league half a league,
Half a league onward,
All in the valley of Death
Rode the six hundred:
Theirs not to make reply,
Theirs not to reason why,
Theirs but to do & die,
Into the valley of Death
Rode the six hundred.

Cannon to right of them,
Cannon to left of them,
Cannon in front of them
Volley'd & thunder'd;
Storm'd at with shot and shell,
Boldly they rode and well,
Into the jaws of Death,
Into the mouth of Hell
Rode the six hundred.

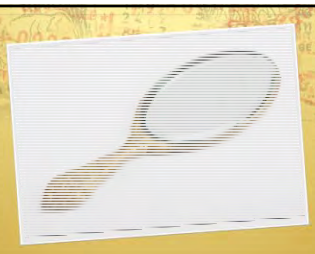
Lord Alfred Tennyson (excerpted)

Great job! (so far)

CHARGE is well known among pediatric providers with their focus . . .



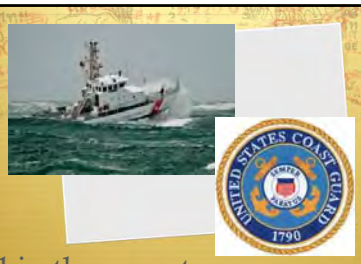
. . . but not among adult providers who have a different focus



Call in the expert

Call in the expert

- You are the experts



Call in the expert
Hit the ground running

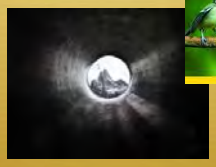
Hit the ground running

- Keep good records
  - comprehensive file
  - focused file
- Know the history
  - diagnoses
  - treatments
  - surgeries
  - -medications

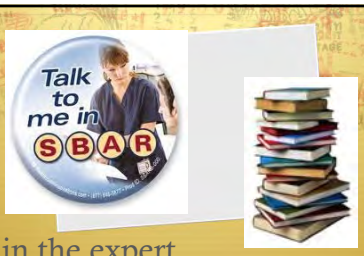
Call in the expert
Hit the ground running
Ask questions

Ask Questions

- Responsibility to educate
- Responsibility to learn



- Potential for tunnel vision



Call in the expert
Hit the ground running
Ask questions
Report & Resources

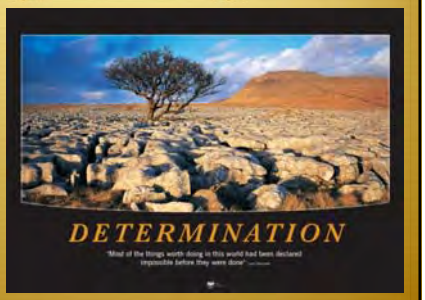
Report & Resources

- SBAR communication
- Suggest websites
- Provide resources
- Identify research
- Participate in studies

Call in the expert
Hit the ground running
Ask questions
Report & Resources
Grind away

Grind Away

- Commit yourself to spending the time and expending the effort to get what you need. Expect that what you are asking for will take more than one request and more than just a little of your time.



Call in the expert
Hit the ground running
Ask questions
Report & Resources
Grind away
Explore

Explore

Your doctor, case worker, therapist . . .

. . . is not the only one in the world.

Call in the expert
Hit the ground running
Ask questions
Report & Resources
Grind away
Explore

Supporting Self-Determination in Prelinguistic Individuals with CHARGE Syndrome

Susan M. Bruce, Ph.D., Boston College

Defining Self-Determination

Self-determination is having control over what happens in your life. (Wehmeyer, 2005).

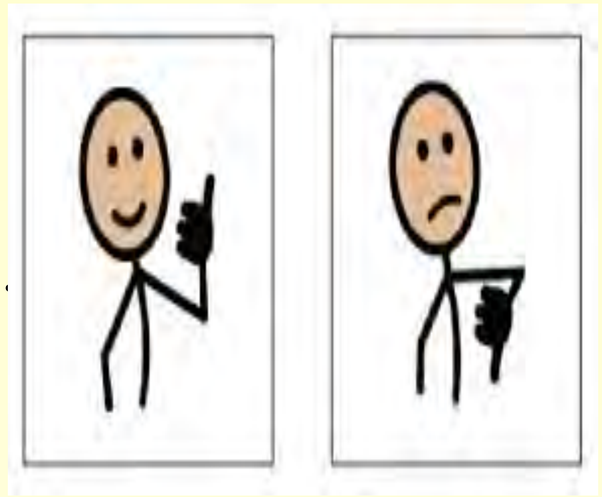
“All people have the right to an education that supports their capacity to take greater control, and deserve the supports that enable them to assume greater control” (Wehmeyer, Bersani, & Gagne, 2000, p. 114).

Components of Self-Determination

- Self awareness
- Self-regulation
- Probleenable them to assume greater control” (Wehmeyerm solving
- Goal setting & attainment
- Choice-making
- Decision-making
- Self-efficacy
- Self-advocacy

(Wood, Fowler, Uphold, & Test, 2005)

Making Decisions

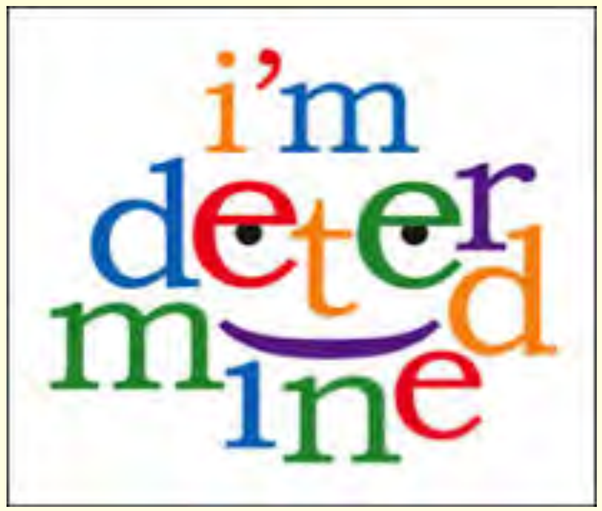


- Decision-making starts in early childhood, with risk increasing over time
- Decisions about what to wear
- Decisions about what to eat
- Decisions about who to work or play with
- Decisions about materials within an activity
- Decisions about when to do something
- Decisions to be “all done”
- Decisions about leisure
- Decisions about vocation
- Decisions about risk

Authentic Choice-Making

- Participation in choice-making routines is different from making AUTHENTIC choices
- Authentic choice-making requires:
  - Knowing your options
  - Knowing what the representations/ symbols for your options mean
  - Having options that are at different levels of preference
  - Having a means to intentionally communicate your selection (e.g. an “indicating response”)

(Shevin & Klein, 2004)



Teach skills for self advocacy including collective and political self-advocacy.

Preference Assessment as Basis of Choice-Making

- Individuals with severe disabilities change their preferences more often.
- Their preferences are more context bound.
- Adults think they know their preferences, but systematic preference assessment indicates otherwise.
- Systematic preference assessment addresses all the senses-vision, hearing, touch, taste. (See Logan & Gast, 2001.)
- Consider the size of the array & means of presentation.

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Models of Conceptualizing Self-Regulation in CHARGE syndrome

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CMU
CENTRAL MICHIGAN UNIVERSITY

Areas of Self-Regulation

Physiological:

the self's ability to alter its own states and responses to meet the needs of the body

Key: sensory integration, which is one's ability to register, orient, interpret, and respond appropriately to a stimulus (Myles et al, 2000).

Behavioral:

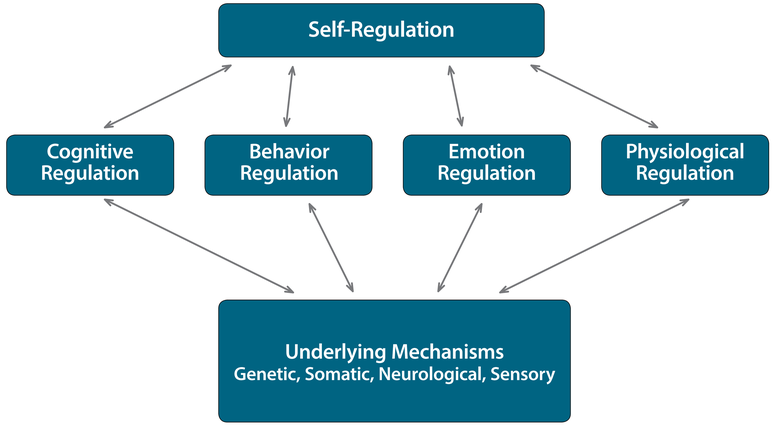
Having an awareness of a behavior and choosing those behaviors most adaptive toward achieving a goal.

Cognitive:

voluntarily regulating thoughts and mental processes by balancing inhibition and initiation of behavior in order to achieve a goal.

Emotion:

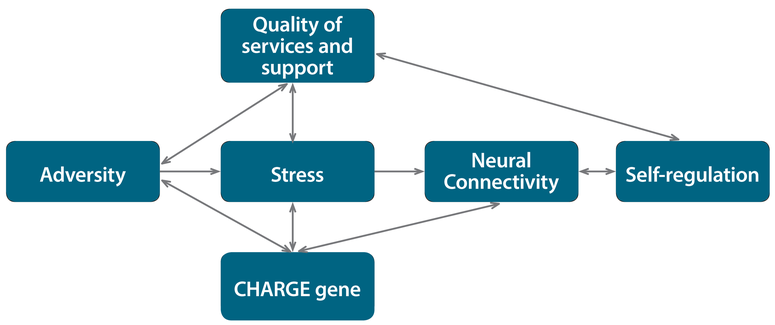
a process involving the analysis, control, alteration, or prevention of emotional expression and experiences that are adaptive for a situation



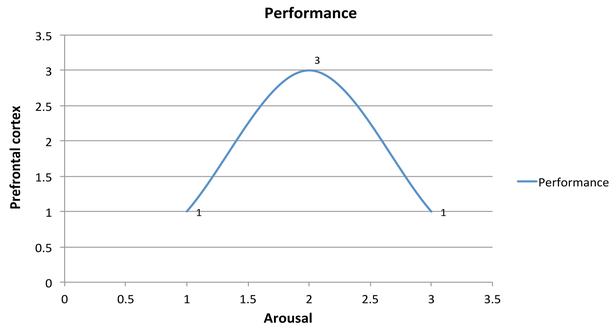
Jude Nicholas & Tim Hartshorne, 2009

Changes in thoughts, behavior, feelings, sensations	PASSIVE Self-regulation Strategies	ACTIVE Self-regulation Strategies
Habituation	Non-reactive Tune it out	Sensation Seeking
Sensitization	Reactive to Stimuli	Sensation Avoiding

After Dunn, 1997



After Blair & Raver, 2012



Self-regulation is used to manage arousal levels

With a regulatory disorder, child is challenged to manage

Cognitive – unfocused vs. obsessive

Behavior – hyperactive vs. hypoactive

Emotion – reactive vs. passive

Physiological – overwhelmed vs. underwhelmed

References

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People with CHARGE Syndrome often have congenital cardiovascular defects. Mutations in *CHD7*, the gene encoding chromodomain helicase DNA binding protein 7, have been identified in CHARGE Syndrome in a majority of cases. In an effort to understand more about this syndrome, scientists have created mice that have only one functioning copy of the *Chd7* gene. Mice with a loss of *Chd7* function display CHARGE-like phenotypes and are an excellent model of human CHARGE Syndrome. Several of the murine models with one functioning *Chd7* gene display congenital cardiac abnormalities. In patients with CHARGE Syndrome, observed congenital heart problems are likely a result of abnormal development of the conotruncal region (outflow tract- i.e. where the pumping chambers of the heart give rise to pulmonary artery and the aorta) and the great vessels (aorta and pulmonary artery).

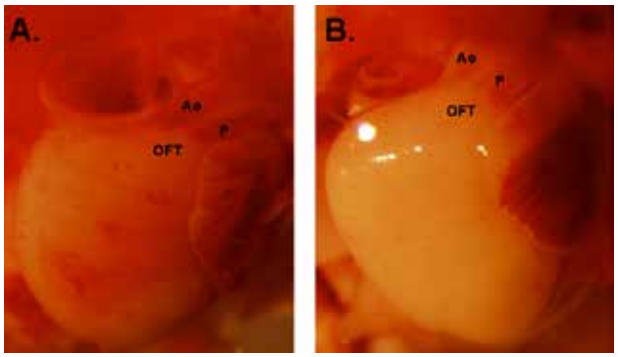

Figure 1. Grossly intact septation and structure of the outflow tract (OFT), aorta (Ao), and pulmonary trunk (P) in mice with *Chd7* still present in NCCs (Panel A) and with *Chd7* deleted in NCCs (Panel B).

During development, the conotruncal region and the great vessels are derived from several different populations of early progenitor cells. Neural crest cells (NCCs) are one developmental cell population that is necessary for the proper development of these cardiac structures. NCCs are an interesting cell type that originates near the neural tube. NCCs migrate to many different parts of the developing embryo where they differentiate into a diverse array of tissues. Thus, it is plausible that many of the clinical features of CHARGE Syndrome may be a result of an inability of NCCs to migrate and differentiate into various tissues. Interestingly, it has been shown that one

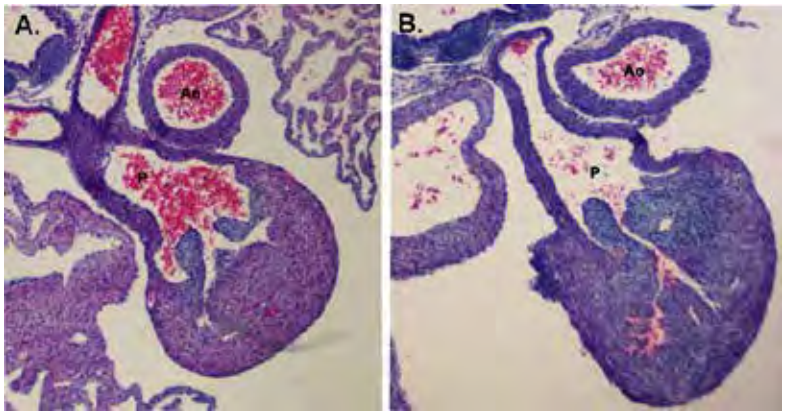

Figure 2. H&E staining of embryonic sections from mice with *Chd7* present in neural crest cells (Panel A) and in mice with *Chd7* absent from NCCs revealed intact septation and overall structure of the pulmonary trunk (P) and the aorta (Ao).

group of NCCs, namely cardiac NCCs, migrate from the neural tube to the heart where they are important for the normal development of the heart and the great vessels. The objective of our study was to determine whether *Chd7* is important for NCCs to migrate to the heart region and successfully participate in the development of the outflow tract and the great vessels.

We created a unique mouse model in which we could breed mice to selectively delete *Chd7* (*Chd7*^{flox}) from NCCs (*Wnt1-Cre*). We found that we were not able to generate viable pups that have *Chd7* deleted from the NCC population (*Wnt1-Cre:Chd7*$^{flox/flox}$). The pups died shortly after birth, and our observations revealed that the pups likely died due to abnormal brain development and oral palate defects that prevented them from feeding properly. In order to determine if mice without *Chd7* in the NCC population have abnormalities in the development of the conotruncal region and the great vessels, we studied at embryonic mice just before birth (e16 and later). As shown in figure 1, at embryonic day 18, we observed normal development of the outflow tract and normal septation of the aorta and pulmonary trunk. In order to further visualize the structure of the outflow tract, pulmonary trunk and aorta of these structures, we fixed and cut tissues from mice with and without *Chd7* in the NCCs. As shown in figure 2, staining revealed that the aorta and the pulmonary trunk were septated.

Thus far, our results suggest that *Chd7* deletion in NCCs using a *Wnt1-Cre* driver is not critical for the development of the outflow tracts and the septation of the pulmonary trunk and the aorta. We are currently looking at earlier time points to see if the development of the pharyngeal arch arteries is impacted by the deletion of Chd7 in NCCs. We are also using other murine models to delete *Chd7* from a variety of early cell populations that participate in the development of the cardiac structures that are often impacted in patients with CHARGE Syndrome.

Behavior as self-regulatory adaptation, or "I can't believe my child just did that!"

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Typical Deafblind Behavior

- Eye pressing
- Finger flicking
- Rocking
- Tapping body/objects
- Self-injurious behavior
- Mouthing objects
- Tactile defensiveness
- Clinging
- Spinning
- Vocal tics
- Feces smearing
- Lining things up
- Extreme preferences
- Darting/running off
- Learned helplessness
- Submissive
- Stare at lights
- Inappropriate vocalization

How to make sense of it

- The kid has a syndrome!
- It's pathological and should be eliminated
- It's due to frustration and pain
- It's communication
- It works for the kid

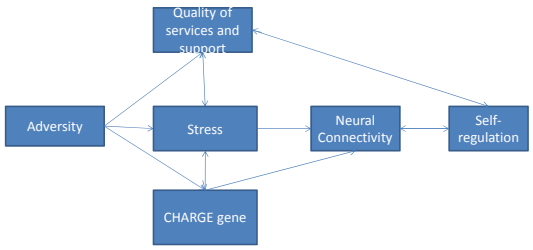
Not because they guarantee success, but because they serve a purpose

Self-regulation problems in CHARGE

- Rapid changes in arousal levels
- Melt downs
- Unfocused behavior
- Diagnoses
  - OCD – a way to reduce stimulation and exercise control
  - ADHD – a problem with regulating sensory and behavioral stimulation and focusing on a goal
  - Tic disorder – a stress response to lack of control over environment
  - Autistic-like behavior – the failure of regulation strategies, and the adoption of dysregulated behavior

Definition

The primarily voluntary regulation of cognition, behavior, emotion, and physiological states for the purpose of goal-directed actions



After Blair & Raver, 2012

Adversity

- Fragile health
  - Breathing problems
  - Multiple hospitalizations
  - Multiple surgeries with anesthesia
  - Multi-sensory impairment
  - Defects in major organs
- Nervous parents
- Sources of stress
  - Social relationships
  - School
  - Family
  - Abuse

Quality of Services and Support

- Lack of medical or specialist knowledge
- Needs multi-disciplinary medical and educational teams
- Parent-Professional relationships
- Lack of social support
- Parent and family resilience

CHD7 Gene

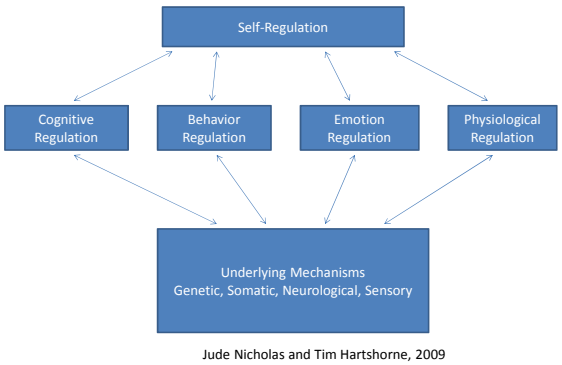
- Regulatory gene
  - Neural crest
  - Placode cells
- Multisensory impairment
- Major organs may be affected
- Vestibular functioning impaired

Stress

- Endocrine regulatory system
- Perception of adversity
- Availability of resources
- Response of professionals
- Response of family

Neural Connectivity

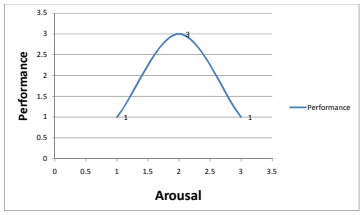
- Prefrontal cortex and executive function
  - Reactive forms of learning and behavior
  - Reflective forms of learning and behavior
- Neuropsychological control over behavioural schemas
  - Routine control
  - Supervisory attentional system



Jude Nicholas and Tim Hartshorne, 2009

Dunn Conceptual Model

Arousal of thoughts, behavior, feelings, sensations	PASSIVE Self-regulation Strategies	ACTIVE Self-regulation Strategies
Habituation	Non-reactive Tune it out	Sensation Seeking
Sensitization	Reactive to Stimuli	Sensation Avoiding



Self-regulation is used to manage arousal levels

With a regulatory disorder, child is challenged to manage
- Cognitive – unfocused vs. obsessive
- Behavior – hyperactive vs. hypoactive
- Emotion – reactive vs. passive
- Physiological – overwhelmed vs. underwhelmed

Self-regulation begins with a goal

– What do you want to have happen?
– What must you do to make it happen?

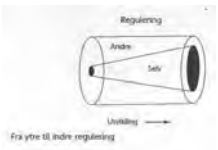
Study for an exam

- Cognitive
- Behavioral
- Emotion
- Physiological

Strategies?

Supporting self-regulation

- Because self-regulation skills are hard for children with significant disabilities to develop
- We have to provide the external support for what will become an internal self-regulatory process



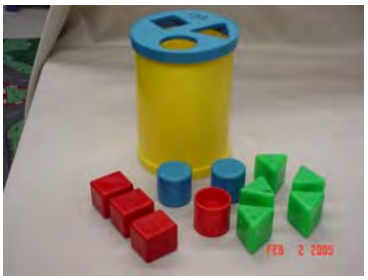
Scaffolding

- The process of planning and organizing the activity of children so that they can execute a task that is beyond their current level of ability.

Components of Scaffolding

1. Identification of the problem to be solved
2. Focus activities on outcomes and goals
3. Frustration control
4. Reducing the complexity of the task
5. Marking critical relevant features
6. Modeling

The Shape Sorter



1. Problem Identification
2. Focus on outcomes
3. Frustration control
4. Reducing complexity
5. Marking features
6. Modeling

Examples

- Cognitive self-regulation
  - Break down larger goals into shorter (pie)
- Behavioral self-regulation
  - Feedback on reactions from others (consequences)
- Emotional self-regulation
  - Creating an environment for self-soothing
- Physiological self-regulation
  - Squeeze technique; hand on arm or leg

Summary

- Children with disabilities often have poorly regulated systems
- This is centrally related to stress, deriving from adversity, quality of supports, and genetics
- The child's attempts to self-regulate manifest as peculiar behavior, often labeled as challenging
- They will do better socially and academically if they can learn to self-regulate
- They can only develop self-regulation skills slowly while they experience a lot of scaffolding from the adults in their lives

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