

Communication and Learning Strategies for individuals with Rett Syndrome

General Characteristics of Rett Syndrome that Impact Learning

- Rett Syndrome is a neuro-developmental, genetic disorder found mostly in girls - There is a phase of degeneration, but over-all it is not a degenerative disease. Children do make progress and learn.
- One of their greatest challenges is Apraxia/Dyspraxia.
 - Apraxia is the inability to reliably connect thought to action
 - Dyspraxia: the signal gets through some of the time, but may be delayed or misdirected
 - Neurological connections are formed, but not as strongly
 - Compare to using the back roads instead of the main highway
 - Getting from intent to action takes more time!
- Breathing and Alerting Abnormalities Affect Ability to Move as Intended
 - Difficulties with autonomic nervous system controlled by the brain stem
 - Breathing dysrhythmias
 - Weak parasympathetic (automatic calming) response
 - May get too much or too little oxygen and/or carbon dioxide due to breathing
- Difficulty Regulating the Autonomic Nervous system
 - Fatigue
 - Temperature regulation
 - Circulation (sometimes to one extremity randomly)
 - Sleep cycle disruptions
 - Swallowing
 - Gastro-intestinal movements
 - Anxiety
 - Agitation
- Observe for Sensory Regulation and Readiness for Learning
 - Identify signals of dysregulation and regulation for each child
 - Work with OT, family and others to make a list of strategies that help with regulation - Apply as needed
 - Take advantage of teachable moments when child is regulated
- Inability to Move Increases with Demand
 - The harder the child tries, the harder it is for her to perform it on demand.
 - May need to move away before moving toward what she intends

- Neurological Stereotypies
 - Neurologically caused - child does not intend to make these movements
 - Varies with day, stress, anxiety, pain, fatigue and other unexplained reasons
 - Masks intelligence
 - Often confused with sensory integration problems
 - The Child Must Over-Ride the Stereotypies to Perform a Motor Task for Communication
 - Wait for a response beyond the stereotypy with patient anticipation
 - Splinting
 - Music / Rhythm
 - Intention/Interest
- Dyspraxia/Apraxia Affects Movements that Control Speech
- Dyspraxia/Apraxia Affects Movements that Control non-verbal communication
 - Difficulty moving as intended affects other communication skills - non-verbal social signals and sustained eye contact (She may appear disinterested)
 - May make it difficult to maintain eye gaze and move eyes efficiently (even though eye-gaze may be a strength)
- These children often have trouble with typical skills that we classify as early communicative behaviors
 - Early communicative gestures
 - Directed or coordinated eye-gaze for joint attention
 - Non-verbal signals
- Therefore, they may get incorrectly labeled as "pre-intentional" or "low functioning" and not provided with appropriate Augmentative and Alternative Communication Systems, supports, and strategic learning environments
- We cannot see intention, so for children older than the age where typically developing children show intention we cannot use the term "pre"
- It is Easy to Make the Wrong Assumptions about Cognitive and Language Potential for Children with Rett Syndrome

Strategies to enhance learning

- Encouraging, Quiet Wait Time
 - Interact and then wait with attention, but not demand
 - Sometimes look away to free gaze
 - Don't keep "re-booting" the system
- Attentive / Respectful Wait Time
 - They know when someone is waiting for them or not

- They often learn which people will likely take the time to wait, so they can decide if it is worth the effort
- Note: When the child produces spontaneous movement, no one can see the time it took between initiation and movement
- Understand that these children are very sensitive to non-verbal communications and attitude of others
- Very tuned into what you are thinking and will reflect hidden emotions
- Will often have certain people that they work well with - people they can rely on to wait and respect them
- Sometimes, you can facilitate faster attention to you or materials:
Movement, Proximity and Moving Your Face into the Child's View
- Look for, and respond to, any subtle communicative signals the child uses
 - Communication is multi-modal
 - Any attempts at communication are accepted and valued
 - Work towards clearer and more reliable signals
 - Move towards more independence
 - Support the child's Intent
 - Provide Strategic Feedback, instead of prompting
- Provide a little Movement Assistance when Stuck
 - Move her a little (hip, trunk, shoulder)
 - Separate hands
 - Only help once child shows intent
 - Allow child to complete movement on her own
- Most of these children are significantly motivated by connection with Others:
 - Be interactive - socially engaging
 - Take turns, laugh, tease and share pleasure in little things
 - Teach the child next to them
 - Modeling by peers and/or adults can be a very effective strategy
- Due to effort required for initiating and carrying out movements, they need to see a good reason for doing something. Determine the purpose or goal of an activity from the adult's and child's perspective - Why am I doing this?
- When a child wants to do something, her brain actually acts more efficiently and that task becomes easier for the child
- Withdrawal or passivity may be the child's way of protecting themselves from further failure
- Prompting and helping too much can lead to Learned Helplessness
- Role of teacher, therapist, and para-professional is to facilitate independence, active engagement, and support for problem solving - Not just "get it right"
- Scaffold the environment to allow for problem solving and discovery learning

- Limit or eliminate hand-over-hand assistance - try to support movement initiated by the child, instead of moving her hand (or any part of the body) for her
- External rewards and reinforcers can reduce mastery motivation and shift child's attention away from task toward the reinforce
- Empty praise is NOT helpful
- Meaningful feedback vs. praise for performance
 - Less general "cheering"
 - She knows when she did something or when someone just put her through the motions (hand over hand)
 - Focus meaningful praise and feedback on what she does do
 - No one likes to be told what to do all the time. Focus on providing feedback for what she does
 - Utilize Strategic Feedback Instead of Prompting

Use a robust Aided-Language system to model general receptive input - Aided Language Stimulation - Select a robust language system that has:

- A clear language organizational structure that builds to increasing levels of language sophistication
 - Systems that build from early language through complex syntax - without changing patterns of vocabulary organization
 - Systems that maintain similarities across access methodologies
- Full range of communicative functions
- Can be always available - light and high tech options
- Designed for **autonomous** communication
 - Imagine: You are a teenager with Rett Syndrome
 - Mom is dressing you and talking about what you are going to do this weekend
 - Go to little brother's soccer game
 - Go to the park and watch the kids playing
 - You would like to go to the mall and buy new shoes - How do you say that?
 - Communication Autonomy (von Tetzchner & Grove, 2003)
 - Must be the child's message - Even if she needs help to communicate it
 - Not just a response to the options provided by others
 - Responsible for her own message
- System Design - NEEDS (Goossens, Crain & Elder 1992)
 - Sufficient vocabulary
 - Design considerations for aided-language stimulation
 - Similarities between individual systems
 - Built for development

- Selection techniques which are not too physically taxing to promote meaningful communication
- Built for growth - like you buy clothes for a toddler
- Will she take it to a party? (Gayle Porter)
 - PODD can be a Powerful Option to meet these specifications (Pragmatic Organization Dynamic Display) (Gayle Porter 2006)
 - Features to Support Autonomous Communication in PODD
 - Aided language development is supported -multiple page sets (communication books)
 - The range of page sets reflects a developmental process as reported in the literature on both typical and aided language development
 - Page sets are designed to facilitate the use of aided language stimulation
 - Similarities of patterns across access methods - 'light' and 'high tech'
 - Organization of vocabulary within PODD
 - According to the individual's communication function and discourse requirements
 - Combines the advantages of a wide variety of organizational systems: pragmatic intents, categories, activity specific, topic and anecdote organizations can all be used
 - Includes scaffolds for supporting aided language development
 - Efficiency to meet communication requirements is the overriding factor
 - Tested and refined through use in a range of natural contexts
 - Scaffolds for early aided-language acquisition and efficiency
 - Strategies for increasing use of syntax and utterance length for partner understanding
- No pre-requisites for aided-language input
 - Myth: Children must have a certain set of skills to be able to benefit from AAC (Ronski and Sevcik, 2005)
 - Typical children hear the language that they will use for a full year before the first expressive word
 - Children need to be exposed to a large number of language models in natural context to be able to learn language patterns
- It's about building Language and Interaction - Not just learning vocabulary or answering questions correctly
- Begin with Receptive Input - Aided Language Stimulation immediately

Teaching movements for Communication: Initiation, multi-modal gestures, selection of message (partner-assisted scanning, eye gaze, switch access, etc.) and access methods for independent and autonomous communication on an SGD

- What Does Research Say About Learning a Motor Task?
 - Initiation of intent must come from within the child
 - Problem solving with opportunities for trial and error are essential
 - Practice and repetition with a purpose
 - Thousands of repetitions with variation
 - Motor skills may need to be developed or refined over many years
- "Touch Points" - Dale Gardner-Fox M.S., RPT
 - 2 and 4 points
 - No response means
 - Co-planning Sequenced Social Scripts (Burkhart and Musselwhite)
- Learning Yes/No as an Alternative to Pointing - NOT for Responding to Random Questions!
- Why is Yes / No So Hard and Often Seen as Inconsistent?
- Indicating a selection from a list should always include options for "none of those" or "something else"
- Obligatory lists should always include option for "I don't know" and also
- Partner-assisted scanning: reject / accept
- Teach two movements to reject & accept - differentiated "YES" / "NO" signals
 - Dyspraxia prevents using any access methods that involve timing
 - Allows the child to maintain control of the speed of communication, enabling her to take as much time to process as needed
 - Less skill required from the partner by eliminating the timing element
 - Reduces partner influence and misreading of social responses within a scan
 - Increased activity may cause physical fatigue for some children
- One movement - for accept
 - The child does nothing until the required option is indicated
 - Partner needs to provide an appropriate pause time between each item familiar partners often feel more confident of the child's responses
 - The child needs to be able to reliably produce their "YES" movement within the identified pause time
 - Experience suggests that less familiar partners often feel less confident of the child's responses
- Problems with "look at me for yes"
 - May work for a quick shared thought, but breaks down with longer autonomous communicative messages

- Eye contact and smiles are social connection and may get misinterpreted as "yes" when used to engage
- Not responding takes effort and child must inhibit looking during a scan or list
- Some children begin to look more autistic, because they actively inhibit eye-contact to prevent accidentally saying "yes"
- Teach Yes/No Head Movements with a target
 - Long term goal to use a natural gesture that will be readable by many communication partners down the road
 - Yes/no head movements
 - Who will be able to read it without training?
 - Doesn't require extra steps for the partner to hold up cards to look at for each scan
 - Children are perceived as smarter if they use a more typical means of saying yes and no
 - May be worth the cost of learning
 - Hand held talking (yes/no) switches may be used as targets for head to move toward during the yes/no training process as well as providing clear feedback to the child (not mounted)
 - Goal to fade the use of switches and move to head movements for Yes/No as soon as possible. (Keep switches available for days when movement challenges are more severe)
- Partner-Assisted Scanning with objects, verbal lists, environmental gestures, and robust communication system
- Partner-Assisted Scanning with iPad Apps
 - Pipe Cleaner Pointers
 - Won't activate screen
 - Clarifies what is being scanned
 - Helps focus visual attention
- Rhythmical Intention and Self-Talk
 - Learning to intend movements
 - Attention to alignment, weight bearing, stability and weight shift
 - Rhythm and development of self talk
 - Practice
 - Still allowing for wait time

Partner-Assisted Scanning with robust communication system such as PODD

- Begin with receptive input
- Reduce the use of questions - make statements and describe what is going on in a variety of contexts
- Model what the child might be thinking and what her behavior might be saying

- Recognize when she might have something to say
- Offer her "do you have something to say" when it seems likely she does - be fine if she says no
- Clear your mind
- Scanning is not a series of questions
- Use separate voices for operational scan (monotone, rhythmical) voice and interactive social communication
- Whatever the child says/babbles is correct
- Assume intention, even at the babbling stage and use the communication system to respond and talk to the child: expand, recast, continue the conversation
- Consider specific customizations for the design of the robust communication system for use with partner-assisted scanning
 - Visual scanning as best option if possible
 - Visual plus auditory slows down the process and adds verbal clutter to the working memory
 - Visual scanning teaches picture symbols which makes transitioning to more advanced levels of language easier
 - Scanning is not a series of questions
 - Keep rhythm even and monotone: this column, this one, etc.
 - Navigation for control of utterance length and more complete messages
- Light Tech vs High Tech
 - "Light Tech" Systems Advantages
 - Portability / Usability
 - Multiple environments
 - Multiple positions
 - Reduced motor demands
 - Face to face connection with communication partner during the whole process
 - The use of a 'smart partner' operating system
 - "Light Tech" Systems Disadvantages
 - size and weight of the system to provide a large vocabulary that is matched to the child's needs
 - the need for partner-training for operating the system correctly
 - "High Tech" Systems Advantages
 - Speech-generated or pre-recorded voice that can be spoken out loud
 - Initiate and communicate independently (when set up for use)
 - Independence in message generation

- Access to extensive vocabulary without adding weight as with a paper system
 - High tech: eye-gaze systems better at reading eye-gaze for communication than another person
- "High Tech" Systems Disadvantages
 - need for more refined motor access skills
 - limited environments
 - dependence on battery power
 - equipment failure
 - Need for higher levels of language competencies - increased time needed to prepare messages which are morphologically correct
 - May block visual range and face to face communication
- Features for Communication Systems light and high tech
 - Robust language system
 - Designed for efficient use of alternative access method such as partner-assisted scanning or eye-pointing
 - Designed for conversation (both parts)
 - Designed for pragmatic use at any time
- Strategies for High Tech systems
 - Consider the difference between looking and pointing
 - Cover cameras while child is looking and not pointing
 - Teach child to pause while looking and then un-pause
 - Begin with highly motivating activities and games
 - Focus learning on child's intent, not following directions
- Eye-Gaze Communication system
 - Light tech: use eye-gaze for choices of 2 - 4 - not generally for communication, because of the limited number of items on a page for language development
 - Individuals with Rett Syndrome are capable of using partner-assisted scanning for light tech communication access and eye-gaze for high tech communication access
 - All children who use AAC require a paper system, even if they have a high tech system, so that communication can happen in any context and position throughout the day - not just when the technology is set up and working
 - Light tech and high tech systems must have similarities so that use is transparent (symbols, organization of vocabulary, etc.
 - Light tech and high tech systems must have differences to take advantages of the particular efficiencies of each platform (message window selection, editing and operational

controls, screen/book shape and size, smart partner operating system vs. computer operating system, etc.)

- Eye-gaze is a motor skill that needs to be learned
 - Different from just looking
 - Involves holding gaze and looking around screen for vocabulary,
 - Begin with simple games and motivating activities that reduce pressure for correctness when learning
 - Manipulate the Environment
 - Pretend Play
 - Personally Relevant (topics, photos, videos)
 - Large Targets
 - Limited Number of Targets
 - Control to Choose, Change and Stop
 - Pausing for looking away for thinking and processing
 - Pause for modeling
 - Speak message window - separate so child can read and edit message before speaking
 - Consider using a reduced set of vocabulary (Simple Powerful page set) when introducing communication system on the eye-gaze device, if access is still challenging. Make sure to keep vocabulary symbols and organization consistent with light tech system and also next level to move to on eye-gaze

Developing Automaticity

- What do I Think About?
- Activate the device? What did she just ask me? Hold up my head? Who just walked in the door? What was that noise? What do I know about this? How could I answer that? Why does my stomach hurt?
- Challenges of Working Memory
 - What do I Think About?
 - Activate the device? What did she just ask me? Hold up my head? Who just walked in the door? What was that noise? What do I know about this? How could I answer that? Why does my stomach hurt?
- Developing automaticity takes practice
 - Thousands of Repetitions with
 - Intent
 - Purpose
 - and Variation
- Motivation Provides Intent
- Natural Context Provides Purpose and Variation

Testing vs. Teaching

- Being able to do something in context is different than taking a test about it
- Children Learn by Doing
- Emphasize Experience - Not Drills
- Problems with Testing and Assessment
 - Pressure to perform on cue - increases dyspraxia
 - No intent, purpose or variation
- Use Dynamic Assessment integrated into the day, instead of single session, multiple questions assessment
- Direct Questions Increase Difficulty of Moving with Intention
 - Make Statements Instead:
 - I wonder where the _____ is
 - That is a _____
- 4 to 1 Rule of thumb in Natural Contexts:
 - 4 inputs: teaching, commenting, explaining, demonstrating, modeling (may need to be 5 or 6 to 1 at first)
 - 1 integrated test question related to that teaching (stated indirectly if possible)
 - Repeat (data collected over time not in one sitting)
- Plan and Look for Teachable Moments
 - Follow the child's interests - Relate information to the child's life experiences
 - Child needs to understand: Why am I doing this?
- "High Tech" eye-pointing and switch access
 - Remember: You can not use an access strategy to test a child, until that access strategy has become automatic
- The Juggling Act and Working Memory
 - Sensory
 - Motor
 - Language
 - Cognitive skills
- Girls with Rett Syndrome May Have Lost or Never Reached a Level of Automaticity with Motor Skills
 - May Require a Great Deal of Cognitive Effort to Move
- Balance Cognitive and Motor Difficulty
- Juggling Explains Inconsistency of Performance
 - Need to take successes and move on, as opposed to requiring repetition of the task over a given number of trials
 - Provide opportunities for repetition/practice within natural contexts with variation and natural motivation
- Parallel Programming instead of sequential
 - "Light Tech" Communication Book for Language

- Switch Play to Develop Motor Skills
- Play to Develop Eye-Gaze Motor Skills
- Eventually: Combine Motor and Language Skills to Operate a Communication Device

Separate Academics from Communication

- Produce a product as a result of the child's efforts
- Modify the amount and specific components of work required
 - Teacher determines what is most important for each activity
 - Focus on quality learning instead of quantity
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- Changing the amount of time allowed to complete work
 - May need to break up assignments into shorter time periods across multiple days
 - Writing a simple story may take a week or two
- Position of child in relationship to position of materials
 - Think vertical
 - Dollar store picture frames
 - PVC eye-gaze frame
- "Light Tech" eye-pointing strategies and protocols
- Help the child focus attention on the important relevant components of a task
 - Post-it Notes
 - Two White Boards
 - Objects
 - Window to Direct Visual Focus
 - Cut Words Apart to Build Sentence
- Working with manipulatives that the child may not be able to directly interact with
 - Counting Manipulative
 - Magnetic
 - Velcro on carpet squares
 - Number Sentence / Part Part Map
- Assume Competence!
- Keep Your Expectations OPEN!