**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 overall 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
  o Neurologically caused - child does not intend to make these movements
  o Varies with day, stress, anxiety, pain, fatigue and other unexplained reasons
  o Masks intelligence
  o Often confused with sensory integration problems
  o The Child Must Over-Ride the Stereotypies to Perform a Motor Task for Communication
  o Wait for a response beyond the stereotypy with patient anticipation
  o Splinting
  o Music / Rhythm
  o Intention/Interest

• Dyspraxia/Apraxia Affects Movements that Control Speech
• Dyspraxia/Apraxia Affects Movements that Control non-verbal communication
  o Difficulty moving as intended affects other communication skills - non-verbal social signals and sustained eye contact (She may appear disinterested)
  o 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
  o Early communicative gestures
  o Directed or coordinated eye-gaze for joint attention
  o 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
  o Interact and then wait with attention, but not demand
  o Sometimes look away to free gaze
  o Don’t keep “re-booting” the system
• Attentive / Respectful Wait Time
  o 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
  o Less general “cheering”
  o She knows when she did something or when someone just put her through the motions (hand over hand)
  o Focus meaningful praise and feedback on what she does do
  o No one likes to be told what to do all the time. Focus on providing feedback for what she does
  o 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
  o Systems that build from early language through complex syntax - without changing patterns of vocabulary organization
  o 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
  o 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?
  o 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)
  o Sufficient vocabulary
  o Design considerations for aided-language stimulation
  o Similarities between individual systems
  o 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 (Romski 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
  o Visual scanning as best option if possible
  o Visual plus auditory slows down the process and adds verbal clutter to the working memory
  o Visual scanning teaches picture symbols which makes transitioning to more advanced levels of language easier
  o Scanning is not a series of questions
  o Keep rhythm even and monotone: this column, this one, etc.
  o Navigation for control of utterance length and more complete messages
• Light Tech vs High Tech
  o "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
  o "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
  o "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 then 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!