

Getting Past the Barrier of Apraxia to Interactive Communication for Children with Rett Syndrome

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Rett Syndrome

A neuro-developmental, genetic disorder found mostly in girls - There are phases of degeneration, but over-all it is Not a degenerative disease.

Children do make progress and learn (after degenerative phase)

What is Dyspraxia or Apraxia?

Apraxia is the inability to reliably connect thought to action

Dyspraxia: the signal gets through some of the time, but may be delayed

Driving Analogy



- Imagine that your car is your body and you are your brain
- Sometimes when you step on the brake, the car speeds up. Sometimes it slows down.
- Sometimes when you want to turn right, the car turns left
- That is what apraxia/dyspraxia feels like

Getting from Thought to Action!

- The child may be able to perform one movement, such as picking up a cookie - because that has been practiced for years with intent, purpose and variation
- Performing a different movement, such as pressing a switch is an entirely different skill for these children. Each movement must be learned over thousands of repetitions with intent purpose and variation

Dyspraxia and Apraxia in Rett Syndrome

- Neurological connections are formed, but not as strongly as typically brains
- Similar to the neurological signals using the back roads instead of the main highway
- Getting from intent to action takes **more time!**

Neurological Stereotypies

- Neurologically caused - child does not intend to make these movements and may not be under her control
- Varies with day, stress, anxiety, pain, fatigue and other unexplained reasons
- Masks intelligence - because it appears that the child wants to put her hands in her mouth, like that of a much younger child - but that is not usually the case

The Child Must Over-Ride the Stereotypies to Perform a Motor Task for Communication

- Wait for a response beyond the stereotypy with patient anticipation and no pressure
- Try gently holding or splinting one of both arms to interrupt the stereotypy. This may be appreciated and desired by the child and she should be allowed to ask for the splints on or off
- Music can sometimes reduce stereotypies
- Intention/Interest may reduce stereotypies

Breathing and Alerting Abnormalities Affect Ability to Move as Intended

- Difficulties with autonomic nervous system controlled by the brain stem
- Weak parasympathetic (automatic calming) response - she may become anxious and have difficulty calming herself
- Breathing dysrhythmias
- May get too much or too little oxygen and/or carbon dioxide due to breathing irregularities

Encouraging, Quiet Wait Time

- Don't keep "re-booting" her system
- Wait quietly without pressure
- Limit or eliminate use of hand-over-hand assistance
- Wait to cheer until the child completes her intended movement
- Always give informative feedback related to the intent of the movement

Ability to Initiate Movement Decreases with Demand

- The harder the child tries, the harder it is for her to perform it on demand.
- She may need to move away before moving toward what she intends to do
- When the child gets stuck, gently move her to body slightly to initiate movement, then allow her to finish moving on her own

Dyspraxia/Apraxia Also Affects Movements that Control Speech

- Nonverbal and social signals and sustained or sequenced eye movements may be affected by the difficulty of moving as intended. (She may appear disinterested)
- Even though eye-gaze is one of their strengths, she may have difficulty maintaining eye gaze and moving her eyes efficiently

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
- Sequenced eye movements such as gaining your visual attention and then looking at what she desires
- Non-verbal signals

Lack of early communicative signals fails to naturally trigger a sense of communication readiness to the communication partner

Therefore, these children may get labeled as “low functioning” and not provided with an appropriate learning environment with Augmentative and Alternative Communication Supports

Begin with Receptive Input of an Aided Language System-
“Aided Language Stimulation”
“modeling language” in natural Contexts

Use a robust aided language system with a full range of communicative functions

Teach Movements for Communication

Teach Yes/No as an Alternative to Pointing
- NOT for Responding to Random Questions

Partner-Assisted Scanning

Work to teach typical movements of a head nod for “yes” and a head shake, or turning head away for “no” - whenever possible

- These movements will be more easily recognized by untrained communication partners - especially future partners
- Doesn't require partner to stop and hold up yes/no cards for each scanned item
- Use what the child can do now for yes/no - agree and disagree - Plan and teach more intelligible movements for the long term

Work to teach typical movements of a head nod for “yes” and a head shake, or turning head away for “no” - whenever possible

- Will allow the child to use intelligible head movements to agree or disagree during a conversational discourse when listening to a partner
- Try talking yes/no switches to provide a target for head movement and clear feedback. Activate switches when the head moves toward them - no need to make them actually press the switches

Work to teach typical movements of a head nod for “yes” and a head shake, or turning head away for “no” - whenever possible

- Fade the use of yes/no switches
- Move to using your two hands (one by her cheek and one under her chin), then just use two fingers
- Move switches back slightly after gently tapping them on her chin and cheek - saying yes or no? and waiting
- Eventual goal is for her to be able to move her head without the need to use the switches

Partner-Assisted Scanning With Objects in the Environment







Something else

or

None of those

Last item always needs to be “something else” or “none” of those

Testing and Direct Questions last item should always be: “I don’t know” or “Please get my communication book”

red 	yellow 	blue 	I don't know 
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Visual Partner-Assisted Scanning with PODD Communication Books



(Pragmatic Organization Dynamic Display)

Gayle Porter, Melbourne Australia

Visual Partner-Assisted Scanning with PODD Books

- If the child has good vision, visual scanning is faster and more efficient than auditory plus visual scanning
- Auditory plus visual adds verbal clutter and calls attentions to words that are not part of the message
- Learning the symbols - instead of relying on the auditory, means the child may move up more quickly to higher language levels, and also be able to more easily add the use of an Eye-Gaze SGD (speech generating device) to her repertoire of communication systems

understandinglu.blogspot.pt/2014/02/just-do-it.html?m=1

Blog about Lucy, who uses a PODD communication book and a PODD page set on a Tobii with Grid2 software. She chooses which one to use when

Just Do It!

Said with PODD Communication Book

- "I think it's, you, fun, not, school" (She is home schooled)
- "I want, want, hug, it's about now"



- "Let's go, in the car, you, me, can't, it's about now, I love you." She said this on a snowy day when we had been planning to go to the library, but it was too bad out!

Some things Lucy said with PODD on Tobii: Eye-Gaze (SGD)



Grid2 Software with PODD pageset

- "Let's go, in the car, visit, pappy"
- "I don't want to do it, let's do something else"
- "I want, our, garden, yummy"
- "Have, pain, my, tummy, tired, I don't like it"
- "Lucy, yours, play, music, loud, yours, loud, song" (Her favorite song is "I'm Yours")

Problems with Testing and Assessment

- Performing on demand is extremely challenging for these children
- They are often not able to repeat responses out of a meaningful context, so it is not possible to give them repeated consecutive trials outside of a context
- Tests are standardized based upon normal movement abilities, not on children who experience movement challenges

Direct Questions Increase Difficulty of Moving with Intention

Instead of Questions - Make Statements:

I wonder where the _____ is
That is a _____

(The child may then look at the indicated item or correct response without being directly asked to respond)

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) (collect data over days and weeks instead of during one testing situation)
- 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?** Is it worth the effort?

One experience that taught me:

Large number of children with Rett Syndrome at an International Rett Syndrome Conference - Evening Tribute Party

Many of them owned and could use high-tech Eye-gaze Speech Generating Systems. However, none brought them to the party.....

The only child I could have a conversation with at this event was the one who bought her PODD Communication book to the party

She told me, using her book, that she was cold, so we helped her put on her sweater. I wonder how many other children were thinking the same thing?

High Tech Eye-Gaze Communication Systems can be fantastic for these children! However, they can not be the child's only system, because communication needs to be able to happen at any time - even when device is not set up or not charged

Use Light Tech AND High Tech!

“Considerable experience in working with children with complex communication needs has convinced me that a successful AAC intervention program must have family members as the key participants. AAC is a way of life; it is not an exercise to be practiced twice a week during therapy sessions or even an hour a day in the classroom”

Judy Seligman-Wine

Goals in the implementation of a home program for children with Rett Syndrome

- Teach multimodal strategies for AAC
- Model the use of the communication tools in real contexts for the child and for families
- Work cooperatively with school teams
- make communication fun and meaningful both for the girl and for the people who are interacting with her through empowering them in the use of these tools

iPad Play Activities

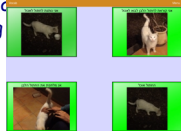


- Partner activates the iPad according to child's direction
 - Use light-tech eye-gaze
 - Or partner-assisted scanning

Create Personal Stories

- Photograph activity and spontaneous events and program them into a story on the iPad, with which the girl can tell other members of her family, neighbors, people at school, etc.

- Example: Every day Ori calls the white cat in her garden to come and eat using a Step by Step. Photos show her feeding the cat and learning to pat it (recorded as a story) (Story has a right to left sequence)



Supporting Parents to take the Lead

" The changes in my daughter's communication profile were largely facilitated through my involvement, resulting in a great increase in communication opportunities throughout the day - at home, at school, and at extended family gatherings. Now that the family sees her with her communication tools and know that she has a way of responding to them, they talk and relate to her differently. Once I saw and understood her communicative abilities, I was able to involve others in these experiences. I see this as a process that is on-going,; the more others relate to her communicative abilities, the more she actively relates to them...."

Assume Competence!

Keep Your Expectations OPEN!