It Just Takes Time: Strategies for individuals with Rett Syndrome

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Rett Syndrome

A neuro-developmental, genetic disorder found mostly in girls - There is a phase of degeneration, but over-all it is Not a degenerative disease. After that phase, children do make progress and learn.

Dyspraxia and/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 or misdirected.

Dyspraxia and Apraxia in Rett Syndrome

- Neurological connections are formed, but not as many

- Compare to using the back roads instead of the main highway

- Getting from intent to action takes more time!
Efferent Kinetic Dyspraxia (motor)
They have the cognitive intention, but not the motor intention
- Neurological connections are formed, but not as strongly
- Difficulty with voluntarily initiating, sequencing and/or co-ordinating movements
- Difficulty finding the 'on' ramp to the freeway
- Difficulty with motor planning (moving from intention to action, despite a physical ability to produce that movement). Getting from intent to action takes more time!

Driving Analogy
Imagine: You are your brain, your car is your body. When you turn the steering wheel right, the car goes left, sometimes...

Getting From Thought to Action!
It takes more time!

Neurological stereotypies
- Caused neurologically - not intentionally (like blinking)
- May be movements that are also intentional (just like eye blinks can be voluntary or involuntary)
  - Hand wringing, washing
  - Hands in mouth or in hair
  - Other hand movements
  - Other body movements, kicking
  - walking in circles

Neurological stereotypies are different from sensory processing challenges
- Sometimes occur along with Rett Syndrome
- Problem is not as much getting the information into brain as it is for getting it back out
- Strategies that work for children who have sensory challenges (normalizing sensory input) often don’t work well for girls with Rett Syndrome
- Strategies that do help are usually related to helping with stability or calming
The Child Must Over-Ride the Stereotypies to Perform a Motor Task for Communication

• Wait for a response beyond the stereotypy with patient anticipation
• Arm wraps
• Music / Rhythm
• Intention/Interest

Dyspraxia/Apraxia Also Affects Movements that Control Speech

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Dyspraxia/Apraxia Also Affects Movements that Control Speech

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Breathing and Alerting Abnormalities Affect Ability to Move as Intended

• Difficulties with autonomic nervous system controlled by the brain stem
• Breathing dysrhythmias
• May get too much or too little oxygen and/or carbon dioxide due to breathing
• Impacts ability to move on demand

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Use Encouraging, Quiet Wait Time

Don’t keep “re-booting” the system

When the student initiates the intent and completes the movement, they strengthen the neurological motor loop

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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)

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Strategic Feedback and a little Assistance when Stuck

- Move her a little (shoulders/pelvis)
- Separate hands
- Only help once child shows intent
- Allow child to complete movement on her own

Additional Challenges with Autonomic Nervous system

- Temperature regulation
- Circulation (sometimes to one extremity randomly)
- Sleep cycle disruptions
- Swallowing
- Gastro-intestinal movements
- Anxiety
- Agitation

Weak parasympathetic (automatic calming) response

- More easily alerted
- More difficulty calming themselves down
- Easily become dysregulated
- Fatigue

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

Provide Individually Designed Mini-Breaks

- a quick nap
- a bite to eat
- short music break
- change of activity
- get up and move
- etc.

Regulate your own state first then “lend” the individual your state
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, Reflexive” or “low functioning” and not provided with appropriate language (AAC) tools, supports, and learning environments.

**NOTE:** We cannot see intention, so for children older than the age where typically developing children show intention we can **not** use the term “pre.”

It is Easy to Make the Wrong Assumptions about Cognitive and Language Potential for Children with Rett Syndrome

Some individuals with Rett are reading, doing math, and other academic work at grade level.

Assessment is a Challenge!

Think Dynamic Assessment Instead

- Assess within natural contexts
- Integrated within the day
- Collect data over time, instead of sitting down in one session

Direct Questions Increase Difficulty of Moving with Intention

Make Statements Instead:
I wonder where the ____ is?
5 to 1 Rule of thumb in Natural Contexts:

- 5-6 inputs: teaching, commenting, explaining, demonstrating, modeling AAC in contexts.
- 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?

Children with Rett Syndrome are Reading and Participating in Academic Learning

- Many do best in inclusive environments
- Modifications include adjustments for
  - Quantity
  - Time allowed
  - Working with peers in group projects

Tips for Literacy and Academic Learning

- Provide robust balanced literacy program like their peers are learning

Focus on Instruction and Experience vs Testing

Separate Academics from Communication

AAC is how we talk about work - not the work
Position of child in relationship to position of materials

Think Vertical

standing or sitting, move materials to keep them at eye level

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

Modify the amount and specific components of work required

Subtraction

9 - 4 =
7 - 5 =
8 - 7 =
9 - 2 =
6 - 5 =
8 - 1 =

• Teacher determines what is most important for each activity

• Focus on quality learning instead of quantity

Produce a product as a result of the child’s efforts

• We often make the child work until they fatigue or the time runs out instead of having clear finished work

• When using manipulatives, also make sure there is a product (paper, or snap a picture, etc.)

Motivated by Connection with Others:

• Be interactive - socially engaging

• Take turns, laugh, tease and share pleasure in little things

Motivated by Connection with Others:

• Most powerful motivation is social connection

• Dyspraxia increases during individual sessions away from peers. Movements are easier and participation increases in classrooms, kindergarten, with peers, and siblings

• Teach the child next to them
Teach Peers to be Friends, not “Helpers”

- Teachers and other adults often set the tone of the classroom
- Discourage “helping”
- Encourage “doing things together”
- Friendships are formed through common interests, not forced
- Naturally talk to other children around her, using her AAC
- Encourage peers to use her AAC by moving it toward them and responding to them
- Assist with operation of AAC without taking over the conversation between child and her peers

Limit or eliminate hand-over-hand assistance - try to support movement initiated by the child, instead of moving their hand for them

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

Entice, don’t tell them what to do

- Interact and then wait with attention, but not demand
- Sometimes look away to free their gaze
- Limit asking questions
- Make comments

Everyone Needs to See a Reason for Doing Something
Language Samples for data collection about communication

- Collect language samples in natural contexts
- Sample instead of writing down everything
- Note partners communication before and after
- Note cued, responded to a question, or initiated
- Note communicative function

Complex Communication Needs

Imagine:

- You are a teenager with Rett Syndrome
- Mom is dressing you and talking about what you are going to do this weekend
- Going to little brother’s soccer game
- Going 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?

Activity - Please find a partner

- First person - will have a secret message
  - In a minute, I’ll show you a secret message to communicate (don’t tell your partner)
  - hold your legs up to simulate instability
  - sit on hands (no gestures)
  - answer only with yes or no head nod - but count to 5 each time, before you move to nod or shake your head
- Second person
  - Ask yes/no questions to guess the message
  - Switch roles and repeat activity

Partner 1 - close eyes or turn away

Partner 2 - pick one and don’t tell anyone

- My foot itches
- My knee hurts
- I want Daddy to help me
Switch Roles

Partner 2 - close eyes or turn away

Partner 2 - pick one and don't tell anyone

• I want to show you something in my bag
• Let's go to the beach
• That is a nice scarf you are wearing

How did that feel?

All of those messages can be easily said using a yes/no access method for partner-assisted scanning in a robust system such as PODD

• Sample messages:
  • My foot itches
  • My knee hurts
  • I want Daddy to help me
  • I want to show you something in my bag
  • Let's go to the beach
  • That is a nice scarf you are wearing

Communication is the Priority!

• Select and model a robust communication system
• Focus on interaction and connection more than the aid (SGD, communication book)
• Teach access methods for both non-electronic and electronic communication
• Teach language and pragmatics through modeling in natural contexts for real reason

The Juggling Act and Working Memory

Sensory, Motor, Language and Cognitive skills
Juggling Explains Inconsistency of Performance

We Need Parallel Learning!

Team plans long term direction and works on skills in parallel

- If we wait for everything to develop in a coordinated fashion, we will be waiting forever
- Don’t hold the child back in one area because of deficits or difficulties in other areas
- Develop rich cognitive schemas and not just splinter skills

Team Planning and coordination is Critical
Everyone needs to be moving in the same direction - long term

- Everyone needs to know enough from other team members to:
  - Understand and use the child’s means of communication
  - Know how the child is learning to move and recognize helpful vs harmful positions and movements
  - Recognize and address dysregulation – know how to address sensory needs on the spot
  - Incorporate what bests facilitates learning for each child (processing differences, learning needs)

Focus on one component or skill within each activity, or part of activity

- Reduce motor load for difficult cognitive, language and academic tasks
- Reduce cognitive load for motor learning tasks
- Teach electronic access (such as eye-gaze) as a separate but parallel skill to language and academic learning

Begin with Receptive Input - Aided Language Stimulation

Use a robust aided language system
Selecting a Robust Aided-Language System

- Clear language organizational structure that builds to increasing levels of language sophistication
- Full range of communicative functions
- Can be always available - 'non-electronic' and 'electronic' options
- Designed for autonomous communication

AAC System Design - NEEDS (Goossens, Crain & Elder 1992)

- Sufficient vocabulary
- Similarities between individual systems
- Built for development
- Selection techniques which are not too physically taxing to promote meaningful communication
- Built for growth

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

Will she take it to a party?

- Gayle Porter

It is not possible to rely on an electronic device as the only option for communication

- All people who use aided AAC require a non-electronic (paper tech) option

Non-Electronic and Electronic

- Not
- Either One or the Other
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!

Said with PODD on Tobii: Eye-Gaze
• "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")

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

Partner-Assisted Scanning

Why is Yes / No So Hard and Often Seen as Inconsistent?

Gayle Porter

Time for a snack?

Would you like an apple?

Yes or No?
Would you like this?

Yes or No?

I guess you aren’t hungry…..

Another Approach

Model First

None of those
Testing and Direct Questions

“I don’t know” and Please get my communication book / device

Two movements to reject & accept - differentiated “YES” / “NO” signals

• Allows the child to control the speed of the communication

• Dyspraxia - Less skill required from the partner by eliminating the timing element

Two movements to reject & accept - differentiated “YES” / “NO” signals

• Reduces partner influence and misreading of social responses within a scan

• Increased activity may cause physical fatigue for some children

One movement to accept
One signal to indicate “YES”

• The child does nothing until the required option is indicated

• Partner needs to provide an appropriate pause time between each item familiar partners

• Possibly increased cognitive fatigue

One movement to accept
One signal to indicate “YES”

• 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
- Apraxia / Dyspraxia increase with any timing demand
- How long do you wait for “no response”?  
- Great variability in time required to initiate movement

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

Motor Cognitive Learning for Access to Partner-Assisted Scanning

Remember to Attend to Position and Teach Key Issues of Movement

- Stability
- Alignment
- Weight bearing
- Weight shift
- Disassociation
- Self-talk to intend movements
Learning to intend head movements in group with Rhythmical Intention

Teach Yes/No Head Movements with a Target - If Child is not in a motor learning program

Practice Yes/No in Fun Ways

Fade the use of Switches Goal is to use head movements for Yes/No

Objects, Books, Dry-Erase Boards

Partner-Assisted Scanning with iPad Apps

Pipe Cleaner Pointers
- Won't activate screen
- Clarifies what is being scanned
- Helps focus visual attention
Partner Assisted Scanning on Apps - Word Wizard

Eye-Gaze - Non-Electronic

- Dyspraxia makes it difficult for partners to read more than a few points accurately
- More tendency for partner influence
- Difficulty with sequencing movement for clear signals
- Electronic displays don’t have these issues

Eye-Gaze - Electronic

- Looking to locate and looking to select are different
- Need to develop motor skill for selecting with increasing accuracy

“Electronic” eye-pointing

- Start with play
- Build access skills
- Avoid high cognitive load when learning access
- Focus learning based on child’s intent, not following directions
- Can not use for testing until access is automatic

Provide opportunities to learn Eye-Pointing as access with reduced cognitive load
Simple Powerful Pageset - If Still Learning Access

Simultaneously used with non-electronic communication book that has similar patterns

Add Speak Button for speaking the message window

Pause to Model

Model Self-Talk and Operational Commands

- Speak message
- Pause
- Delete Word
- Clear
- Go back to Main

A way to ask for her “talker” in her book and a way to ask for book from her talker

“Electronic” eye-pointing and/or switch access

- Remember: You can not use an access strategy to test a child, until that access strategy has become automatic
Assume Potential!

Pause, wait, pause some more

Allow way more time!

Keep Your Expectations OPEN!

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