Rett Syndrome
Assessment and Curricular Materials Modifications

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Rett Syndrome
A neuro-developmental, genetic disorder found mostly in girls

There are phases of degeneration, but overall it is Not a degenerative disease. Children do make progress and learn.

Apraxia is one of their biggest challenges

Apraxia is the inability to reliably connect thought to action.

Driving Analogy

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!

Apraxia in Rett Syndrome

Apraxia 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

• May have difficulty initiating movement
Motor skills may need to be developed or refined over many years before they become automatic.

Motor automaticity takes thousands of repetitions with Intent, purpose and variation.

Problems with Testing and Assessment

- Child may understand and have knowledge but not have the motor skill or communication skill to demonstrate it.
- Children may need to learn specific strategies first, in order to be able to demonstrate cognitive and language understanding.

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 labeled as “pre-intentional” or “low functioning” and not provided with an appropriate learning environment with Augmentative and Alternative Communication Supports.

Testing Requires an Automatic Motor Response

Apraxia Makes an Automatic Motor Response Very Challenging

Developing Automaticity

Working Memory Can Only Deal With a Limited Amount of Information at a Time
If something is not automatic yet, it will occupy the child’s working memory instead of operating in the background.

Automaticity is a Level of Skill Where You No Longer Have to Consciously Think About Performing that Skill.

Working Memory Can Only Deal With a Limited Amount of Information at a Time.

How much? How long?

What do I Think About?
What did she just ask me? Shift my weight, reach out, activate the switch... 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? Oh my stomach hurts?

Developing Automaticity takes practice: Thousands of Repetitions with

Developing Automaticity takes practice: Thousands of Repetitions with Intent.
Developing Automaticity takes practice: Thousands of Repetitions with **Intent**, **Purpose**, and **Variation**

Motivation Provides **Intent**

Natural Context Provides **Purpose** and **Variation**

Testing Provides None of the Needed Components that Facilitate Use of Automaticity

- **Intent**
- **Purpose**
- **Variation**

Teaching vs. Testing

- Imagine driving someplace in another town where you have gone for years

Teaching vs. Testing

- Now imagine taking a test on the directions to get there:
  - How many cross streets before your second left?
  - Name of all the streets
  - What is on all four corners of your 4th right hand turn
Teaching vs. Testing

• Being able to do something in context is different than taking a test about it

Dynamic Assessment

• Assessment is an ongoing interactive and dynamic process over time

• Integral part of teaching every day

Start with what the child can do successfully and build on that

Explore what Supports are Needed for Success in Each Situation

What Facilitates Function, and what Inhibits Function in each aspect of a task?

Easy to Prove What the Child Can Not Do

Look for Possibilities Not Limitations

Build rapport and gain the child’s trust

• Very Sensitive to Non-Verbal Communications and Attitude of Others

• Very tuned into what you are thinking and will reflect hidden emotions

Dynamic Assessment

• Assessment is an ongoing interactive and dynamic process over time

• Integral part of teaching every day

(Gayle Porter and Teresa Iacono, 2006)
Use Encouraging, Attentive 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

• Don’t keep “re-booting” the system

Separate Academics from Communication

4 to 1 Rule of thumb in Natural Contexts:

• 4 inputs: teaching, commenting, explaining, demonstrating, modeling

• 1 integrated test question related to that teaching

• 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?

Identify and regulate sensory readiness for learning

• Recognize a child’s readiness for learning

• Define specific strategies for sensory regulation for each child - up regulate or down regulate

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
Use peers for social motivation

• “I want to do what the others are doing!”

• Peers also make great models

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 – no clear task completion

• When using manipulatives, also make sure there is a product to show off to others - even if it just a paper with a note about what the child did

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

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

sometimes standing
sometimes sitting

“Light Tech” eye-pointing strategies and protocols

sometimes vertical

Writing With Alternative Pencils
Center for Literacy and Disability Studies UNC
Post-it Notes or Two White Boards
Plus: “I don’t know”

Touch Point Choices
No response or look down for “I don’t know”

“High Tech” eye-pointing and switch access
- Learning and practice required to develop automaticity with access
- Looking is different from eye-pointing
- Remember: You can not use an access strategy to test a child, until that access strategy has become automatic

Making a selection from 3 or more items
- Partner-assisted Listing
- Alternative to pointing
- 2 is not a list

Partner-Assisted Scanning
Learning Yes/No as an Alternative to Pointing - NOT for Responding to Random Questions

Time for Dessert
Would you like an apple?
Yes or No?

Would you like this?

Another Approach

Testing and Direct Questions
last item always: “I don’t know”
List Song Verses for Choices

- Clap hands
- Clap feet
- Wiggle
- None of those

Symbols System for Language to talk about choices

Imaginative Play
List of Choices within Play and Robust Language System for Giving Opinions and Receptive Language Input

Objects

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
Help the child focus attention on the important relevant components of a task

Window to Direct Visual Focus

Cut Words Apart to Build Sentence

Window to Direct Visual Focus

Dollar Store Picture Frames
Counting

10, 20, 30, 40, 50, 60, 70, 80, 90, 100,
I don’t know, start again….

That’s it!

Carpet Square and Velcro

I see 7.

Counting Manipulatives

Number Sentence / Part Part Map

3 and 4 is 7

Disclosure: Linda J. Burkhart

• Self-Employed
• Paid Consultant and Trainer
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• Work with children, families and teams privately
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Presume Competence!

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