Rett Syndrome
Apraxia and Communication

What is Apraxia?

- Apraxia is the inability to carry out a cognitive intent - The child intends to move a particular way, but the neurological signal doesn't reliably get to the right muscles to move them consistently
- Analogy to losing control when driving a car, if the pedals and steering signals get crossed

Apraxia in Rett Syndrome

- Girls with Rett Syndrome's most profound disability is Apraxia or Dyspraxia
- 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 increases with demand
- Some motor skills remain intact - but only when triggered with an automatic event that doesn't require forethought or cortical intent. This can be confusing and may be interpreted by others as stubbornness, because it seems that the child can perform a task some of the time. (Example of child picking up a cookie, but not being able to activate a large talking switch with the same hand) However, the harder the child tries, the harder it is for her to perform it on demand.
- Sometimes the child seems to need to move away before moving toward what she intends. If you don't wait for the child to finish the intent, it may appear that she is rejecting or responding incorrectly, when in reality she hasn't finished her movement yet
- Delayed processing from intent to movement is typical for these girls
- Apraxia also affects muscles that control speech
- Apraxia affects other communication skills - including ability to use some non-verbal social signals and sustained eye contact
- Even though eye-gaze is often a strength of children with Rett Syndrome, It may be difficult to maintain eye gaze and move eyes between a desired object and a person who might be able to retrieve the object
- Waiting for a response with patient anticipation is critical to success. The child learns which people will likely take the time to wait for her, so she can decide if it is worth her effort
- Sometimes talking the girl through the motor movements and /or modeling them can be helpful.
- Peers can be powerful motivators for girls with Rett Syndrome

Neurological Stereotypies

- Hand wringing, Mouthing
- Neurologically caused - child does not intend to make these movements
- Child may have to "fight" or "over-ride" these movements to move with intent
- Inconsistency is the norm - Varies day to day, and within a day
- Varies with stress, anxiety, pain, fatigue and many other unexplained reasons
- Masks intelligence
- May be interpreted by others as severe retardation
- Music may reduce stereotypies for some girls
- Splinting or gently holding her non-dominant arm may lesson and improve function of her other hand
- Splinting both arms may work better for some girls
- Even though using hands looks more "normal" and seems to work some of the time, the child may be able to move a different body part more reliably to indicate intent for communication purposes or switch activation - for example her head
- On some days and at certain times, being able to break out of the stereotypy to use hands may be very difficult and may be frustrating for the child and other options should be provided
- Waiting for a response beyond the stereotypy with patient anticipation is critical to success. The child learns which people will likely take the time to wait for her, so she can decide if it is worth her effort to comply or initiate
- Facilitate attention through movement, proximity, and/or moving your face into the child's view

Problems with Testing and Assessment

- "Catch 22" -
    - How do you test language skills before teaching language skills? You need to put a language learning environment in place first
    - How do we test what an individual knows when she hasn't been given the appropriate supports needed to show us what she knows?
- Child may understand and have knowledge but not have the motor or communication skill to demonstrate it
- Children with severe, multiple and complex challenges may need to learn specific strategies first, in order to be able to demonstrate cognitive and language understanding
- We can't make assumptions about cognitive potential for children with complex sensory motor challenges
- 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, these children may get labeled as "pre-intentional" or "low functioning" and not provided with an appropriate learning environment with Augmentative and Alternative Communication Supports
- Communication strategies may need to be taught before testing - Aided Communication Intervention Before Assessment: A Case Study of a Child with Cerebral Palsy (Carol Goossens',1989)
- We Need to Create an Appropriate Language Learning Environment
- Motor skills may need to be developed or refined over time
- Children with complex sensory-motor challenges will need to learn, over many years, the sensory-motor control required to produce intelligible gross / fine motor movements for communication

Developing Automaticity

- Working Memory can only deal with a limited amount of information at a time
- Cognitive attention is needed to focus on anything that is not automatic
- Problem of available working memory - what to focus on? - Activate the switch? 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?
- Memory is stored as patterns not individual details and must contain some variety in order to be generalized to a broad number of situations
- Once a pattern is learned it becomes automatic and operates subconsciously, until there is a need to use it or change it
- If something is not automatic yet, it will occupy the child's working memory instead of operating in the background
- For many children who have severe multiple challenges, motor control requires cognitive attention and effort
- We need to be helping children to get to a point where cognitive efforts can be redirected from the motor skill to the content of the task
- Children need practice in natural contexts, utilizing repetition with moderate differences, intent and purpose

Where do you start?

- Look for, and respond to, any subtle communicative signals the child uses
- Language is multi-modal - Any Attempts at Communication are Accepted as Valid
- Attentive Wait Time
    - They often understand nonverbal communication
    - 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 her effort
- Try different body parts for access (head, shoulders)

- Present vocabulary systematically and predictably
- PODD Communication books Gayle Porter (Melbourne Australia)
- Try Talking "Yes"/"No" Switches with the PODD for Clear Concrete Feedback
- Try Partner-Assisted Scanning With Objects in the Environment and with a PODD
- Work Towards Clearer and More Reliable Signals
- Increase Clarity of Feedback for the child and for the Communication Partner
    - Reduces guessing
    - Provides more success for the child
    - Support the child's Intent
- Provide Strategic Feedback, instead of prompting

Automaticity

- Automaticity is being able to do something without conscious attention to the task
- Much of what we do is automatic - we don't have to think about walking, reaching, brushing our teeth, etc.
- Developing Automaticity takes practice: Thousands of Repetitions with Intent, Purpose, and Variation
    - Motivation Provides Intent - without motivation, intent is external and requires more cognitive effort to perform
    - Natural Context Provides Purpose and Variation - this type of practice facilitates the development of automaticity
    - The key is to connect intent with a rote motor pattern that we don't have to think about
    - Testing Provides None of the Needed Components that Facilitate Use of Automaticity: Intent, purpose and Variation
- Imagine driving someplace in another town where you have gone for years and know the route well
- 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
- Being able to do something in context is different then taking a test about it
- Children Learn by Doing
- Emphasize Experience - Not Drills

Respond with Natural Consequences:

- Respond to all attempts to communicate regardless of form (voice, gesture, picture symbol, eye gaze, etc.)
- Respond with natural communicative response (provide item requested, "oh you want some popcorn - here you go")
- Avoid artificial rewards for talking such as unrelated treats or "good talking"

Work towards Mastery Motivation:

- Mastery Motivation is intrinsic motivation
- Children will be motivated to only do what they know they can do or assume they can do successfully – This is often interpreted by others to mean: "She only does what she wants to do."
- Withdrawal or passivity may be the child's way of protecting themselves from further failure
- Being too helpful or rewarding dependency behaviors can reduce mastery motivation and lead to learned helplessness
- Role of teachers, therapists and para-professionals is to facilitate independence, active engagement, and support for problem solving
- External rewards and reinforcers can reduce mastery motivation and shift child's attention away from task toward the reinforcer. It is more effective for the activity to have relevance and intrinsic motivation for the child
- Reducing the pressure of testing situation and using a play experience instead, can help the child make use of more skills and be more likely to practice them on his own
- Recognize the difference between Testing and Teaching
- Children learn by doing
- Emphasize Experience - not drills