

# Catching a Glimpse and Fueling the Flames of the Hidden Torch Within

## Adopt these Beliefs:

- Everyone Communicates
- Communication Begins with Intent
- Getting from Intent to Action is What is Difficult for Girls with Rett Syndrome
- The result of the effort, must be worth the effort

## Apraxia

- Girls with Rett Syndrome's most profound disability is Apraxia
- Apraxia is the inability to efficiently 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
- 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. 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
- May make it 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
- Intrinsic motivation can help reduce the effects of apraxia
- Peers can be powerful motivators for girls with Rett Syndrome

## Neurological Stereotypies

- Hand wringing, Mouthing, etc.
- 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 cognitive disability - which is rarely the case
- 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 turning her head to activate switches or using an eye-gaze system
- 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 you 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
- All testing shows us - is what the child has or hasn't done in the past given the language learning environment that has been made available to her
- 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

### **Teaching vs. Testing**

- 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

- 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

## The Juggling Act

- Girls with Rett Syndrome may not have reached, or may have lost, automaticity with many skills, especially those with a motor response, and thus need to focus attention on each component to be successful
- To be able to communicate effectively, many individual components must be coordinated. For example:
  - **Sensory-motor demands:** motivation, strength, motor planning, muscle tone, endurance, motor automaticity, auditory filtering, auditory processing, tactile processing, proprioceptive processing, reaction time, visual discrimination, visual scanning/memory, visual tracking, integrating multiple sensory inputs.
  - **Cognitive demands:** motivation, cause/effect, initiating, discriminating purpose and function, developing cognitive schemas, making active choices, trial and error, problem solving, memory
  - **Language components:** motivation, processing of language in activity, relationship to and monitoring of the communication partner, pragmatics, processing of questions, auditory filtering, processing of symbol set, syntax/grammar, attention to task, memory
- Juggling means that the child may only have some of the 'balls in the air' at any given time, and having all the 'balls in the air' will be rare. **This explains why performance is so inconsistent and can not always be predictably repeated**
- 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 without pressure. Use variation and natural motivation



## Parallel Programming:

- Coordinating all components of a task on an automatic level is extremely challenging for children with Rett Syndrome

- Child can become easily frustrated
- If we wait for everything to develop in a coordinated fashion, we will be waiting forever
- We don't want to hold the child back in one area because of deficits or difficulties in other areas
- We need to be careful not to get stuck trying to 'prove' mastery
- The answer to this problem is to work on individual components in parallel.
- Use functional and natural contexts to give the child opportunities to develop skills, where only one component is cognitively challenging her at a time. For ex: motor, cognitive, language, etc.
- Child needs to experience success with at least one component at a time - engineer activities so this is possible
- The task should not be so difficult that the child doesn't experience some sense of success, nor too boring as to not be worth the effort
- Provide a variety of these types of activities throughout the day - to challenge the child in all areas of development, but mainly just one at a time.
- Plan as a team to make sure individual skills are moving in a coordinated direction and will eventually be able to be integrated into meaningful tasks
- Help the child make associations see the relationships between skills that she is developing
- Model component skills as well as integrated skills, so the child can see how things will eventually work together

### **Dynamic Assessment** (Gayle Porter and Teresa Iacono, 2006)

- Assessment is an ongoing interactive and dynamic team process that happens over time
- Integral part of teaching every day
- Start with what the child can do successfully and build on that
- Explore what you want to find out and what might help you find that out
- Intervene - Observe - Adjust Intervention - Observe (repeat)
- Provide moderate changes within a natural context and observe results
- Explore what supports are needed for success in each situation
  - We wouldn't expect a person who is blind to read without learning braille
  - We wouldn't expect a person who is deaf to understand a lecture without a sign language interpreter or captioning
  - We wouldn't expect a student to take a test without a paper and pencil
  - What Tools, Strategies, and Supports are Needed For Each Child who has Significant Physical and Multiple Challenges?
- What facilitates function, and what inhibits function in each aspect of a task, such as: motor skills, vision, hearing, attention, language processing, initiation, expressive

communication, interaction, motivation, active engagement, problem solving, state, and sensory processing

- Adult provides supports and scaffolds to determine possibilities
- Pay close attention to child's sense of competence and feelings of control and choice within the activity
- It is easy to prove what a child can NOT do
- Look for Possibilities Not Limitations
  - Discovery Process - Be a Detective
  - Not Just a Puzzle - Book of Puzzles
  - Challenging and Enjoyable Process!
- We need to find the spark, and fan the flames - Discover what the child can do!
- Remember the difference between testing and teaching