Rett Syndrome
Apraxia and Communication

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Rett Syndrome

A neurodevelopmental, genetic disorder found mostly in girls

Affects approximately 1 in every 9,000 - 10,000 live female births

Apraxia

Apraxia is the inability to reliably connect thought to action

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!

Driving Analogy
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.

Apraxia Also Affects Movements that Control Speech

• Apraxia affects other communication skills - non-verbal social signals and sustained eye contact (She may appear disinterested).

• May make it difficult to maintain eye gaze and move eyes efficiently.

Neurological Stereotypies

• Neurologically caused - child does not intend to make these movements.

• Varies with day, stress, anxiety, pain, fatigue and other unexplained reasons.

• Masks intelligence.

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

• Wait for a response beyond the stereotypy with patient anticipation.

• Splinting.

• Music.

• Intention/Interest.
Apraxia affects 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

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

- Motor skills may need to be developed or refined over many years

Where Do You Start?

- Accept and Celebrate that Language is multi-modal - Any Attempts at Communication are Accepted as Valid
- Look for, and respond to, any subtle communicative signals the child uses

Work Towards Clearer and More Reliable Signals

- Move towards more independence
- Support the child’s Intent
- Provide Strategic Feedback, instead of prompting
Try Talking “Yes”/“No” Switches with the PODD
Clear Concrete Feedback

Partner-Assisted Scanning with Objects in the Environment and with a PODD

Attentive Wait Time
Ask and then wait with attention
Don't keep “re-booting” the system

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

Try Different Body Parts for Access
• Head
• Cheek
• Chin
• Etc.

Parallel Programming
"Light Tech" Communication Book for Language

Present Vocabulary Systematically and Predictably
*PODD Communication books Gayle Porter (Melbourne Australia)

Switch Play to Develop Motor Skills
Play Activities to Develop Eye-Gaze Motor Skills

Eventually: Combine Motor and Language Skills to Operate a Communication Device

Rett Syndrome Point of Access: Eye Gaze
It is often very difficult for girls with RS to indicate what they want to say by pointing to an object or picture with their hand

Response time using their hand is very slow:
combination of motor issues
apraxia
low muscle tone
stereotypic hand wringing movements
Often use their eyes to indicate their choices; fast, efficient, and accurate

Even if they do finally get there with their hands, they may point to a different response than what their eyes had indicated.

It is usually their eyes that have indicated what they really want to say.

“Tell me with your eyes what you want to say”

When they try to indicate their choice by pointing with their hand,

their eyes go to what they want to say long before they get there with their hands.

Eye gaze as her means of access for communication:

the girl with RS does not waste time and physical energy in trying to organize herself motorically while everyone else is waiting patiently (or impatiently) to hear what she wants to say.

Eye pointing is the generally accepted way in which girls with RS make a selection from between a number of items for the purpose of communication
Today eye gaze technology is also being used to enable them to indicate what they want to select on a computer screen.

Need to work on hand function:

now generally accepted that we do not try to work on two difficult goals within the one activity.

Many other activities throughout the day to work on hand function:

- self-feeding
- Artwork
- Gardening
- Powered-mobility
- Vocational work
- etc.

For some girls even eye gaze is too limiting.

Large vocabulary

How to access?

Partner-assisted auditory scanning

Chana:

New Communication Chart
240 + pages
Scanned verbally by her communication assistant/communication partner
Fast and efficient access to a very large vocabulary
What is a Communication Assistant??

A person whose job it is to facilitate communication for a person who communicates using AAC.

Communication Assistants for Girls with Rett Syndrome
By serving as her mouthpiece
- interpreting her unaided communication attempts (gestures, eye gaze, etc).
- verbalizing the messages which she indicates on her communication display
- validating an utterance produced on the SGD/VOCA when necessary

Preparation of AAC materials – to meet ongoing communication needs in all environments
- Advocacy
- Enhanced communication opportunities
- To serve as a model for others wanting to communicate with the AAC communicator

A Communication Assistant MUST be trained
- what is Rett Syndrome
- how does this specific girl communicate
- use of multimodal systems
- recognizing and providing opportunities for communication
- construction of messages for girls with reduced vocabularies

- personal relationship with and respect for the girl whom he/she is assisting
- enabling autonomy
- advocacy
- learning to WAIT!
- etc.

(see Collier, McGhee-Richmond & Self, 2010)

A Communication Assistant is NOT a personal caregiver

The Story of Merav
Merav: 21 year old young woman with RS. One of seven siblings in a bilingual family. Good age appropriate language comprehension skills in Hebrew and English. Active participant in all family activities including homework, dance sessions, music festivities, camping trips, and all holiday celebrations.

Up to age 11 attended a program with a strong emphasis on communication
Used a communication chart and a VOCA to interact at home and at school
Global reading
Self-feeding
Happy

School transfer .................

Age 15 - funding to develop a personalized communication program
At that time Merav spent her time in class either sleeping or screaming:
no communication system,
being fed with no expectation of self-feeding,
appeared sad with no joie de vivre

In the class were seven special needs teenagers, five of whom were non-speaking. All required physical care - toileting, transfers, assistance at mealtime, etc.

The most critical element in implementing a communication program with Merav within her present educational environment was the reduction of the pressure felt by the classroom staff as a result of having Merav in their group.

Maximum of three staff in the class - the teacher, the teacher's assistant, and a volunteer
No background knowledge about Rett syndrome
All felt that the combination of Merav's educational, therapeutic, and physical needs placed too much of a demand on them, with the result that only her physical needs were attended to
First step in the implementation of her communication program - communication assistant, in order to reduce the burden felt by the classroom staff

Plan welcomed by the school
Agreed that it would not be part of the job description to take over responsibility for Merav’s physical care

Implementation - initially for 3 hours per day.
Development of a PODD and use of VOCA
Dramatic reduction of screaming episodes and sleeping
End of the school year - program positively evaluated
During the following school year - communication assistants accompanied her for five and a half hours a day

Changes in Merav’s behavior in all areas, including communication, were noticeable
Even more remarkable were the changes in the attitude of the classroom staff
Program expanded - now has three communication assistants who are with her throughout the entire school day six days a week, and also accompany her to her after school program

She has a multi-page communication chart which she uses at home and at school
She participates in all class activities including geography, citizenship and biology lessons, personal grooming, vocational work, ceramics, gardening, shopping at the school kiosk, weekly outings to the museum, and class discussions

Communication Assistants

Training
Supervision
Evolving job description

- communication facilitator: her mouthpiece – translation, not interpretation
- preparation of communication materials to enable full participation
- advocating for her rights as a full participant within the class and within the school

Invitation to spend weekend away from home with her after-school group (without her communication assistant).

The staff of the program felt sufficiently confident to include Merav, due to the modeling of the use of her communication system by her communication assistant.

A great success and a highlight for Merav: she later wrote a story on the computer telling of this experience (the first of many stories to follow).