

## 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

DSM Classifies Rett Syndrome  
within Autism Spectrum Disorders

Clinically, this is not so; Some of  
their strongest motivators are  
often peers and social interaction

## Apraxia

Apraxia is the inability  
to reliably connect  
thought to action

## Apraxia in Rett Syndrome

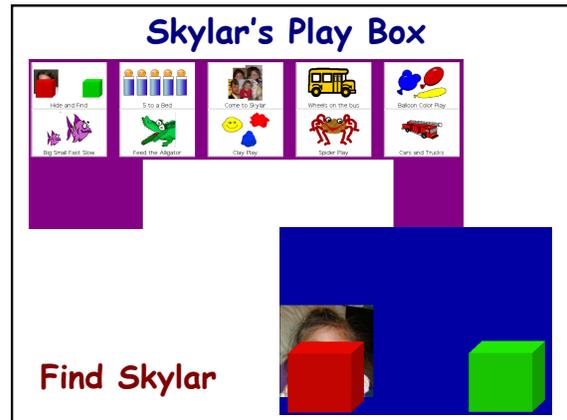
- Neurological connections are  
formed, but not as many
- Compare to using the back  
roads instead of the main  
highway
- Getting from intent to action  
takes **more time!**

## 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**

Gabriella exp different

**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

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Response time using their hand is  
very slow:

combination of motor issues

apraxia  
low muscle tone  
stereotypic hand wringing  
movements

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**Often use their eyes to indicate their choices; fast, efficient, and accurate**

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

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.

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.

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

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**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**

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**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

א1	א1	עבור ל 2	אמירות מהירות
		עבור ל 5	משהו לא בסדר
			I want אני רוצה
		עבור ל א1	I don't want אני לא רוצה
		עבור ל 3	I feel אני מרגישה
		עבור ל 4	I think אני חושבת
		עבור ל 7	I'm doing אני מספרת משהו
		עבור ל א1	אחר

אב	אב	אופס, טעיתי	oops	חזר לדבר ששיתנו בו קודם
		אני שואלת	I am asking	עבור ל 8
		אני רוצה להראות לך משהו	I want to show you something	עבור ל 37
		אני רוצה להגיד משהו למישהו אחר	I want to tell something to someone else	עבור ל 9
		אני רוצה להראות משהו למישהו אחר	I want to show something to someone else	עבור ל 28
		יש לי רעיון	I have an idea	עבור ל 10א
		תסבירו לי	explain to me	עבור ל 11
		תנסו לומר מה אני רוצה לומר ב-10 שניות	try to say what I want to say in 10 seconds	

אג	אג	אופס, טעיתי	oops	חזר לדבר ששיתנו בו קודם
		להמשיך	to continue	עבור ל 10א
		לדבר	to speak	עבור ל 37
		מה שאמרתי (נאמר)	what I said	
		להיות פה	to be here	
		מה שקורה עכשיו	what is happening now	
		לעשות משהו	to do something	עבור ל 10א
		משהו אחר	something else	עבור ל 37

ד1	ד1	אופס, טעיתי	oops	חזר לדבר ששיתנו בו קודם
		מה שנאמר עכשיו	what is said now	עבור ל 37
		מה שקרה עכשיו	what is happening now	עבור ל 37
		מה שעומד לקרות	what is going to happen	עבור ל 23
		מה שחשוב בגודל	what is important in size	עבור ל 37
		למה?	why?	עבור ל 8ה
		משהו אחר	something else	עבור ל 8א

זכ	זכ	אופס, טעיתי	oops	חזר לדבר ששיתנו בו קודם
		אני אתן לך רמז	I will give you a hint	עבור ל 37
		זה קשור ל...	It's related to...	עבור ל 37
		אני רוצה להראות לך משהו	I want to show you something	עבור ל 23
		אני רוצה להראות לך משהו	I want to show you something	עבור ל 37
		משהו אחר	something else	עבור ל 37
		זה חלק מ...	It's part of...	עבור ל 37
		די, תעזבו	Enough, forget	עבור ל 37

**Communication Assistants for Girls with Rett Syndrome**

**What is a Communication Assistant??**  
 A person whose job it is to facilitate communication for a person who communicates using AAC

#### 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 on going 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

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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.

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

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.

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

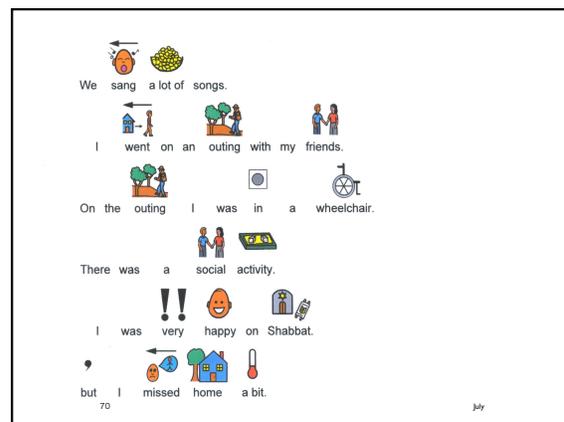
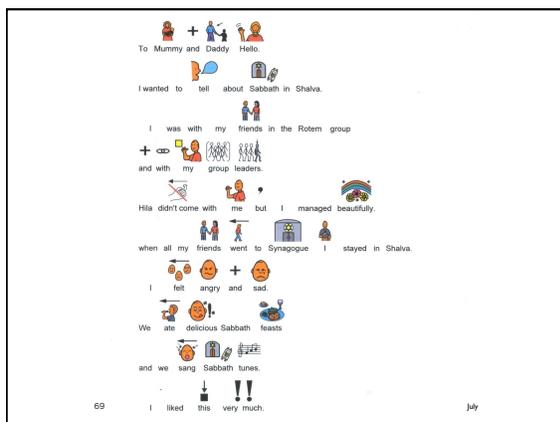
**Evolving job description**

- 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)



This resource can be purchased from  
**The Israel Rett Syndrome Foundation**  
 P.O. Box 1031  
 Ramat Gan 52110  
 Israel  
[www.rett.org.il](http://www.rett.org.il)  
[sigaltirosh@gmail.com](mailto:sigaltirosh@gmail.com)  
[judywine@gmail.com](mailto:judywine@gmail.com)

Price:  
 \$20. U.S.  
 16 Euro

Resource for the Development of a  
**Communication Program**  
 For Girls with Rett Syndrome  
 Judy Wine, Ed.D.  
 with Appendix by Linda J. Burkhart

SILENT ANGELS

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