

## Rett Syndrome: 'Light Tech' vs. 'High Tech': Should It Be Either / Or?

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## Rett Syndrome

A neuro-developmental, genetic disorder found mostly in girls

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

- Age of onset between 6 and 18 months
- Once thought it affected only female; it is now known that there are boys with RS.
- 95% of children with RS complex communication needs
- Numerous other medical issues (sleep, respiration, seizures, constipation, etc.)

- Thought that children with RS are severely intellectually limited
- Level of language comprehension is good; the missing link is their ability to demonstrate their level of understanding
- Now widely accepted: very severe apraxia = the main disability affecting both speech development and hand function in children with RS

DSM Classifies Rett Syndrome within Autism Spectrum Disorders

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

## What is Apraxia in Rett Syndrome?

Apraxia is the inability to reliably connect thought to action

### **The Child Must Over-Ride the Stereotypes to Perform a Motor Task for Communication**

- Neurologically caused and Variable
- Wait for a response beyond the stereotype with patient anticipation
- Splinting
- Music
- Intention/Interest

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

**Development of compensatory strategies for indicating what they want to say**

Very strong strategy – eye pointing/gaze

Have been taking advantage of this ability to enable access to “low tech” communication tools

**Now - as though the new technologies enabling computer access through eye gaze were developed specifically for this population**

**Using these technologies children with RS are demonstrating competencies that they were unable to show previously**

- Language comprehension
- Expressive communication – relating information, asking questions, etc.
- Cognition and learning ability
- Social interaction

**Eye gaze technology = the perfect panacea which will enable the RS population to communicate easily and meaningfully in all situations**

**BUT - not a magic bullet**

Child with RS must have a means to express what she wants to say, whenever and wherever she wants to say it.

AAC must provide a means for the child to express herself in any environment, whenever she has a message to communicate, not simply when she is set up with the equipment according to someone else's agenda.

The use of multiple modalities and a range of systems is more effective than a single communication system



Tendency – start with high tech (eye gaze) to drop everything else – “low tech” i.e. paper charts, simple technology – single and sequenced message devices

**DON'T!!**

Is not an either/or decision

The child must not be held back in terms of language development as her motor skills develop

Solution: Parallel Learning

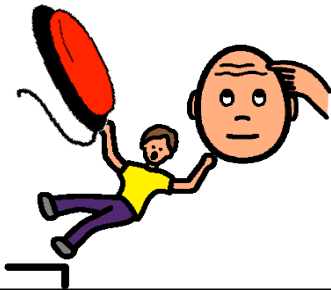
### Juggling Sensory, Motor, Language and Cognitive skills



Girls with Rett Syndrome May Have Lost or Never Reached a Level of Automaticity with Motor Skills

May Require a Great Deal of Cognitive Effort to Move

### Balance Cognitive and Motor Difficulty



### Juggling Explains Inconsistency of Performance



- 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 with variation and natural motivation

### Parallel Programming



### “Light Tech” Communication Book for Language

PODD Light Tech Communication Book

### A variety of ‘light-tech’ tools are available

- eye-pointing
- yes/no responses
- Partner assisted visual and auditory scanning
- Communication Books such as PODD
- Co-planned sequenced social scripts

**Switch Play to Develop Motor Skills**


**High Tech Play to Develop Eye-Pointing Motor Skills**

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

**But always keep "light tech" systems available**

**"Light Tech" Systems**

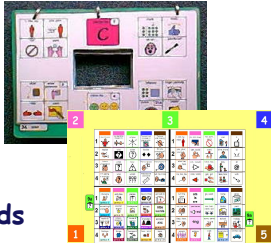
**Advantages**



- Portability / Usability
- Multiple environments
- Multiple positions

**"Light Tech" Systems**

**Advantages**



- Reduced motor demands
- The use of a 'smart partner' operating system

**"Light Tech" Systems**


**Disadvantages**

- size and weight of the system to provide a large vocabulary that is matched to the child's needs
- the need for partner-training for operating the system correctly

**"High Tech" Systems**

**Advantages**

- Speech-generated or pre-recorded voice that can be spoken out loud
- Initiate and communicate independently (when set up for use)



## “High Tech” Systems

### Advantages

- Independence in message generation
- Access to extensive vocabulary without adding weight as with a paper system



## “High Tech” Systems

### Disadvantages

- need for more refined motor access skills
- limited environments
- dependence on battery power

## “High Tech” Systems

### Disadvantages

- equipment failure
- Need for higher levels of language competencies – increased time needed to prepare messages which are morphologically correct

## Features for High Tech systems

- Consider the difference between looking and pointing
- Cover cameras while looking
- Teach child to pause while looking and then unpause – beginning with games
- Focus learning on child’s intent, not following directions

## Features for Light Tech partner-assisted systems

- Robust language system
- Designed for efficient use of alternative access method such as partner-assisted scanning
- Designed for conversation (both parts)
- Designed for pragmatic use at any time

Biggest disadvantage – COST

International Ramifications

In Israel, only one child with RS has her own eye gaze system

Not available in any educational institution

Not an option

**Child 1**

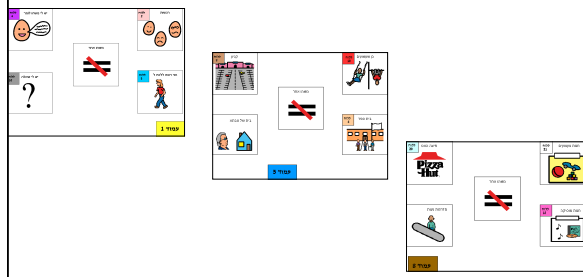
7 years old

First introduced to AAC strategies at age 2 years

Currently uses a combination of strategies to communicate:

- gestures, vocalizations, facial expressions
- Yes/no responses

- Communication chart – “navigational” chart – based on idea of PODD but 5 items per page

**Step by Step**

- Information and messages back and forth between school and home; content determined by her (communication chart)
- Stories composed on her communication chart – interactive singing
- Social Scripts
- Babysitting for baby brother – singing to him
- Games and interactions with other children – hide-and-seek

Likes to write stories and then tell them to others

Had opportunity to use My Tobii 2 years ago; on loan for three months, learned quickly, used it well, mother's excitement

Now - computer through use of scanning - not for communication - too slow

Also – simulation of eye gaze

- Grid 2 software (only Hebrew AAC speech output software)
- Charts same as her “navigational” charts
- She gazes at what she wants to say and her communication partner presses the touch screen to elicit the spoken message

**Child 2**

Now 15 ½ years old

- Attended in past a kindergarten for children with c.p., then an anthroposophic school for children on the spectrum

- For the past two years, school for children with learning disabilities
- Has a classroom assistant at all times at school who also acts as her communication assistant
- Exams through multiple choice questions

Her communication tools:

- a single message speech output device on her desk at all times "I have something to say"
- an erase board,
- a wallet with "Yes" and "No" in it,

- her eye gaze "navigation" chart



- a Step by Step used in many different ways
- Her communication assistant always has "yes" on her right hand and "no" on her left hand
- a MyTobii (desk top) - only at home - likes to use it to select music and songs, not to talk