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

ISAAC 2012
The Child Must Over-Ride the Stereotypies to Perform a Motor Task for Communication

- Neurologically caused and Variable
- Wait for a response beyond the stereotypy 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 so 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

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

- 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

Juggling Explains Inconsistency of Performance

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

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)

“Light Tech” Systems

Advantages

• Reduced motor demands
• The use of a ‘smart partner’ operating system

Disadvantages

Portability / Usability
Multiple environments
Multiple positions

Light Tech z Systems

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

• Independence in message generation

• Access to extensive vocabulary without adding weight as with a paper system

Disadvantages

• Need for more refined motor access skills

• Limited environments

• Dependence on battery power

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

**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