Teaching Movements for Communication for Individuals who have Rett Syndrome

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Rett Syndrome

A neuro-developmental, genetic disorder found mostly in girls - There is a phase of degeneration, but over-all it is <u>Not a</u> <u>degenerative disease</u>. After that phase, <u>children do make progress and learn</u>

Dyspraxia and/or Apraxia?

<u>Apraxia</u> is the inability to reliably connect thought to action <u>Dyspraxia</u>: the signal gets through some of the time, but may be delayed or misdirected

Dyspraxia and 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



Getting from Intent to Action

Breathing and Alerting Abnormalities Affect Ability to Move as Intended

- Difficulties with autonomic nervous system controlled by the <u>brain stem</u>
- <u>Breathing</u> dysrhythmias
- Weak parasympathetic (automatic <u>calming</u>) response
- May get too much or too little <u>oxygen</u> and/or <u>carbon dioxide</u> due to breathing

Fatigue and Difficulty Regulating Autonomic Nervous System

Additional Challenges with Autonomic Nervous system

- Temperature regulation
- Circulation (sometimes to one extremity randomly)
- Sleep cycle disruptions
- Swallowing
- Gastro-intestinal movements
- Anxiety
- Agitation

Inability to Move 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

Skylar's Play Box



Find Skylar

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

 Often confused with sensory processing or cognitive challenges 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 / Rhythm
- Intention/Interest

Encouraging, Quiet Wait Time

• Don't keep "re-booting" the system

Joci wait time switch ed

Teach Movements For Communication

- Initiation
- multi-modal gestures
- selection of message (partner-assisted scanning, eye pointing, switch access, etc.)
- Access methods for independent and autonomous communication on an SGD

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

Developing Automaticity takes practice: Thousands of Repetitions with

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Developing Automaticity takes practice: Thousands of Repetitions with Intent, Purpose, and Variation



- Key movement issues are movement skills that develop spontaneously in typical development.
- Teach children with physical challenges how to move in a variety of positions – concepts of movement.
- Support children to generalize this skill in a range of environments and positions.
- Need to link concepts of movement to function



Key movement issues













 Children need to be in sustained symmetrical positions to prevent musculo- skeletal deformities

• Need to teach children to use both sides of their body.

 May need lateral supports on supportive seating/standing frames to maintain symmetry



Weight-bearing

- Children need to learn to weight-bear through aligned limbs.
- Weight-bearing assists in providing the child with stability. E.g. flat feet in standing, WB through hands to keep body stable

Weight-bearing ensures:

- Elongation of muscles
- Improves bone density
- Improves joint strength
- Reach, grasp, release for fine motor



Weight-bearing

 Many children have challenges with keeping their knees bent and feet flat in sitting.

 Ankle weights assist children to "feel" weight down through flat feet.



Disassociation

 Learning to disassociate movements, that is to move one part of the body while keeping the rest of the body still, enables the person to control movements and be more successful.





Lift feet in the air and maintain them there for the next section





- Stability enables a child to: weight-shift, weight-bear, disassociate and maintain symmetry.
- Stability is required in all positions to enable movement.

 Many components: using different body parts to make themselves stable e.g. using hands in sitting to stabilize self, ensuring feel are flat.

Core stability



Teaching concepts - stability

 Check your sitting - how is your body feeling...feeling tired?

- Could you sit like this all day?
- What effect would it have on your body?
- Pain
- Deformity
- Fatigue



Position checks to help children to learn to be stable prior to movement

E.g. sitting

My feet are flat My bottom is back My back is straight My arms are straight My hands are grasping My head is in the middle and my mouth is closed

Equipment and facilitations to assist children to learn stability

- Body splints/suits e.g. second skin, DMO, TSL etc.
- Arm and leg wraps
- Ankle weights
- Straps when, where and why?
- Weighted lap blankets
- Pelvic, shoulder and lateral supports
- Pushing down through straight arms and flat hands for stability.



Touch Points Co-Planned Sequenced Social Scripts

Emma script cropped

Why is Yes / No So Hard and Often Seen as Inconsistent?



Gayle Porter

Learning Yes/No as an <u>Alternative to Pointing</u> - NOT for Responding to Random Questions

Partner-Assisted Scanning

Partner-Assisted Scanning

Two movements to reject &

<u>accept</u> - differentiated "YES" / "NO" signals

- Dyspraxia Less skill required from the partner by eliminating the timing element
- Allows the child to control the speed of the communication

Two movements to reject &

<u>accept</u> - differentiated "YES" / "NO" signals

 Less skill required from the partner by eliminating the timing element

 Reduces partner influence and misreading of social responses within a scan

 Increased activity may cause physical fatigue for some children One movement to accept One signal to indicate "YES"

The child does nothing until the required option is indicated

 Partner needs to provide an appropriate pause time between each item familiar partners often feel more confident of the child's responses One movement to accept One signal to indicate "YES"

 The child needs to be able to reliably produce their "YES" movement within the identified pause time

 Experience suggests that less familiar partners often feel less confident of the child's responses

Problems with "look at me for yes"

- May work for a quick shared thought, but breaks down with longer autonomous communicative messages
- Eye contact and smiles are social connection and may get misinterpreted as "yes" when used to engage
- Not responding takes effort and child must inhibit looking during a scan or list
- Some children begin to look more autistic, because they actively inhibit eye-contact to prevent accidentally saying "yes"

Long term goal to use a natural gesture that will be readable by many communication partners down the road

- •Yes/no head movements
- Who will be able to read it without training?
- Doesn't require extra steps for the partner to hold up cards to look at for each scan
- Children are perceived as smarter if they use a more typical means of saying yes and no

Teaching movements for communication

Mr Yes/Mr No-(learn to accept/reject)

- Start with the child in a stable position enables disassociation of head from body.
- Provide the child with time to produce the motor movement.
- Verbally reference what child is doing to give feedback.
- Ensure success





Rhythmical intentions

"Rhythmical intention is another form of facilitation. It has two important factors, firstly to make activity voluntary and second, rhythm."

Hari & Akos, 1998, p. 209





 The use of rhythm to facilitate "activity " / movement

 Rhythm of movement

Kinetic melody

Intention

- To make activity voluntary
- To teach children how to use language (cognitive processes) to regulate their own behaviour / movement.

(internal speech / self talk)



Speech incorporates both rhythmical and semantic aspects.

I clap twice

Semantics = clap clap Rhythm = clap clap clap

> **Intention**: Execution:

I clap twice

I clap twice

clap clap

1, 2



Reported effects of rhythmical intention:

- improves attention and concentration
- anticipation
- energising
- motivation
- group cohesion point of focus between people
- provides rhythm of the movement
- allows time for the task to be carried out or maintained
- rhythm can influence muscle tone; can increase co-contraction of muscles.
- clarifies the intention



Reported effects of rhythmical intention:

- provides the process for achieving the intention
- •helps to do the movement
- information about the child's own body
- how to move own body
- what the movement means
- the components of the movement
- •affirms the achievement of the task
- teaches the child how to "regulate own movement" - "use self talk"
- *strengthens memory and learning
- facilitates endurance
- organizes



Learning to intend head movements in group using Rhythmical Intention



Teach Yes/No Head Movements with a Target

Hand Held - Talking yes/no switches used as a target and feedback for head movements (not mounted)

Fade the use of Switches and move to head movements for Yes/No

Kylie choose wrist band and hand

"High Tech" eye-pointing

Start with play

Build access skills

 Avoid high cognitive load when learning access

 Can not use for testing until access is automatic

Manipulate the Environment Pretend Play



Personally Relevant (topics, photos, videos)





Large Targets





Limited Number of Targets



Control to Choose, Change and Stop





Simple Powerful Pageset



Pause to Model

Speak Message Button





Physical strategies which enable a stable position in her trunk and upper limbs for communication:

- lateral supports on her supported seating
- ankle weights to assist with feet flat (disassociation and weight-bearing)
- knee strap to reduce external rotation
- second skin boned arm gators/arm wraps. This also supports healthy development of Fleur's hips.
- When using Tobii (high tech) grasp bar and hand cuffs (assist with disassociation and stability).



Challenges with self regulation have improved with;

- maturity
- increased strength and endurance
- socialisation at Kinder
- increased use of language to express herself; complain and share opinions.
- Increase partner use of operational procedures in PODD that increase autonomy, reduced frustration
 - does not need music as often to calm down.



Fleur looks towards her PODD communication book and vocalises to initiate communication.

Strategies which have supported Fleur's learning include:

- 'I want my communication book' on a 'step by step' communicator within reach at all times
- Verbally referencing our observations of movements which may indicate an initiation
 - Support partner recognition of subtle movements



Strategies to support use of PODD communication book:

- Correct procedures for Partner-assisted visual scanning
 - Not auditory plus visual
- providing un-interrupted pause time after a visual scan
- acknowledging and responding to subtle movements for yes/no
- all partners valuing and modeling language in her PODD book for a range of functions, across all environments



Strategies to support use of eye-gaze device currently with a simple, powerful PODD page set):

- ensuring a stable position to enable disassociation of head and eye movements
- commencing with daily practice of motor eye movements for leisure purposes
- exposure to modeling in high-tech PODD page set
 - simple language level
 - modeling all operational components
 - is now combining 3-4 words in both light tech and high tech.

Fleur seems to be having difficulties accessing all points on a screen, particularly top right corner.

• may be affecting 'speak' button

