

Motricity in Rett syndrome



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Prof. Meir Lotan, France, 2023

First thing first



- To the French association for inviting me here
- The children and families with RS
- My Co-workers

What are the topics for today

- Proprioceptive system at the core of Rett syndrome difficulties.
- Fear of movement
- Core muscles activation

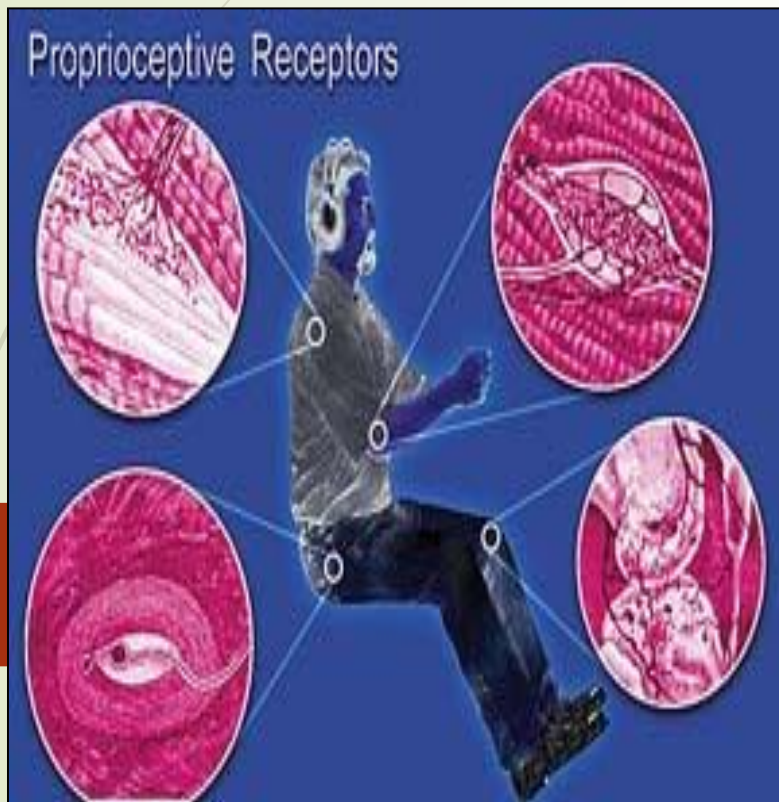
Preface

- One of the many characteristics of RS is fear of movement (FOM), especially when movement is initiated by an external facilitator.
- This fear of movement was mostly associated with the vestibular system (Lindberg, 2006).

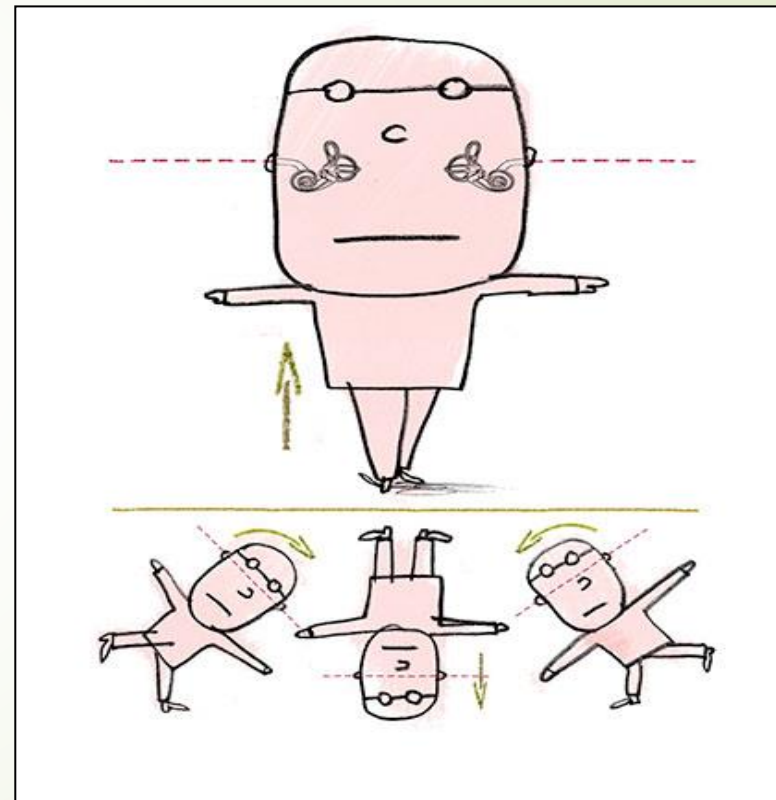


Fear of movement in RTT

Vestibular or Proprioceptive?



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Preface

- One of the many characteristics of RTT is fear of movement, especially when initiated by an external facilitator.
- This fear of movement was mostly associated with the vestibular system (Lindberg, 2006).
- Yet, we suspected that fear of movement in individuals with RTT originates from a dysfunctional proprioceptive system



Fear of movement

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- Was initially referred and defined by Jane Ayres at 1979 as:
- “An inability to appropriately react against gravity “
- “FOM initiates due to lack of sensory integration between the Vestibular and Limbic system (Ayres. 1979)

Vestibular vs.

Proprioceptive



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The present intervention:

- 10 children and women with RTT
- Ages: 3-30 (mean: 17.1 ± 10)
- Community home – 4; Home – 6
- Walking – 5 wheelchair – 5

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Vestibular -

Proprioceptive



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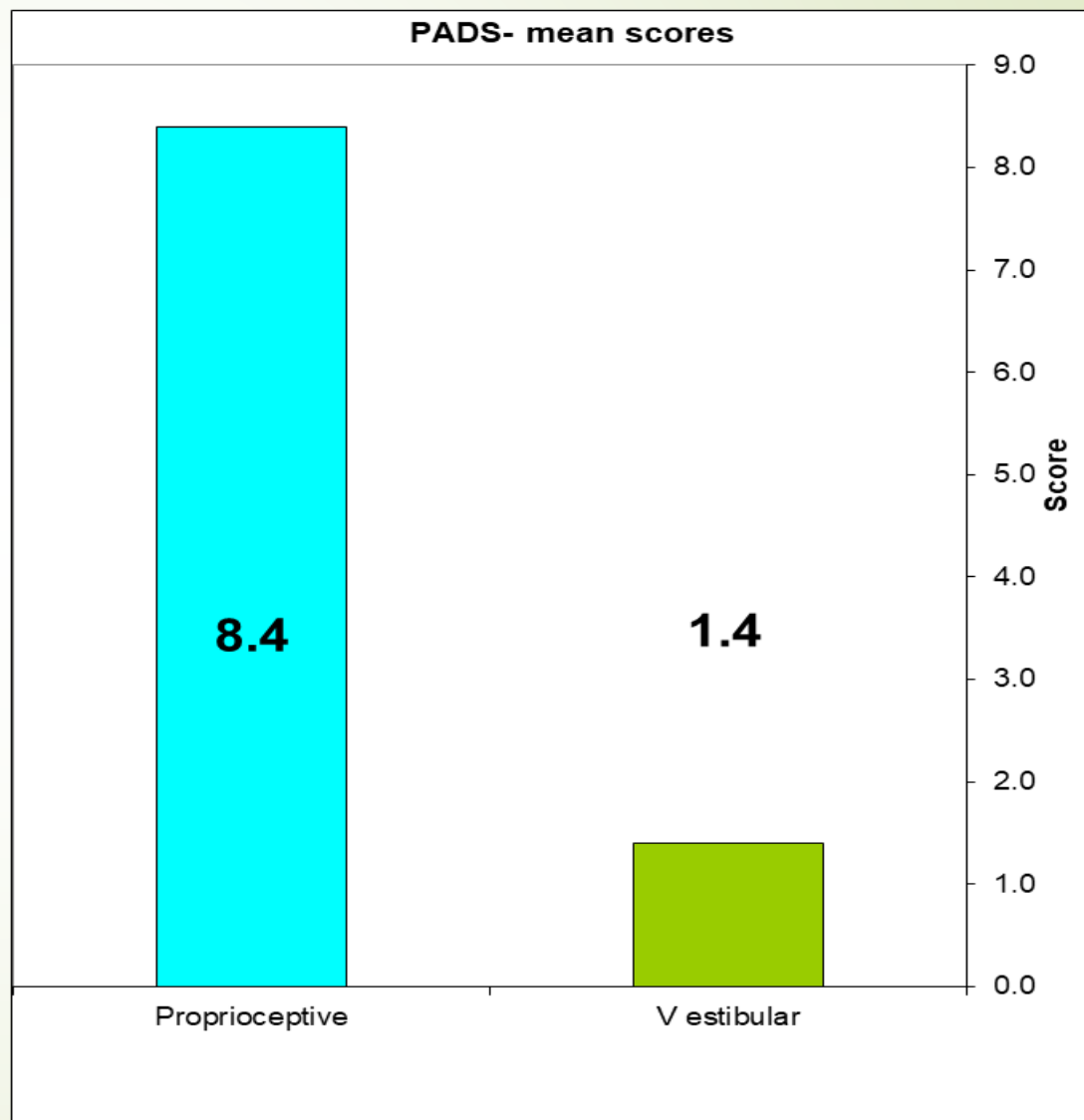
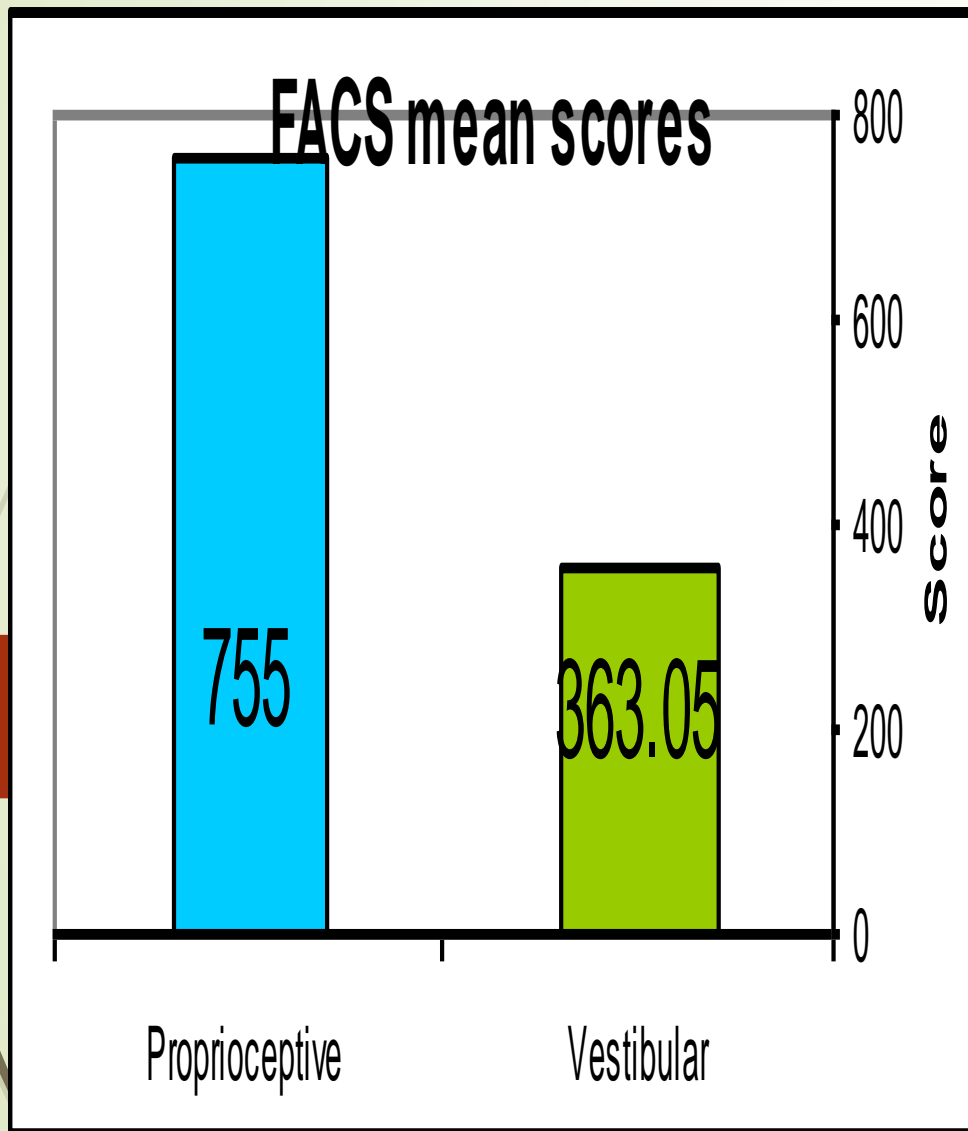
Fear of movement in individuals with RS- Measures

- **PADS -**
Pain and Discomfort Scale
- **FACS –**
Facial Action Coding System

Fear of movement in individuals with RTT

- Results

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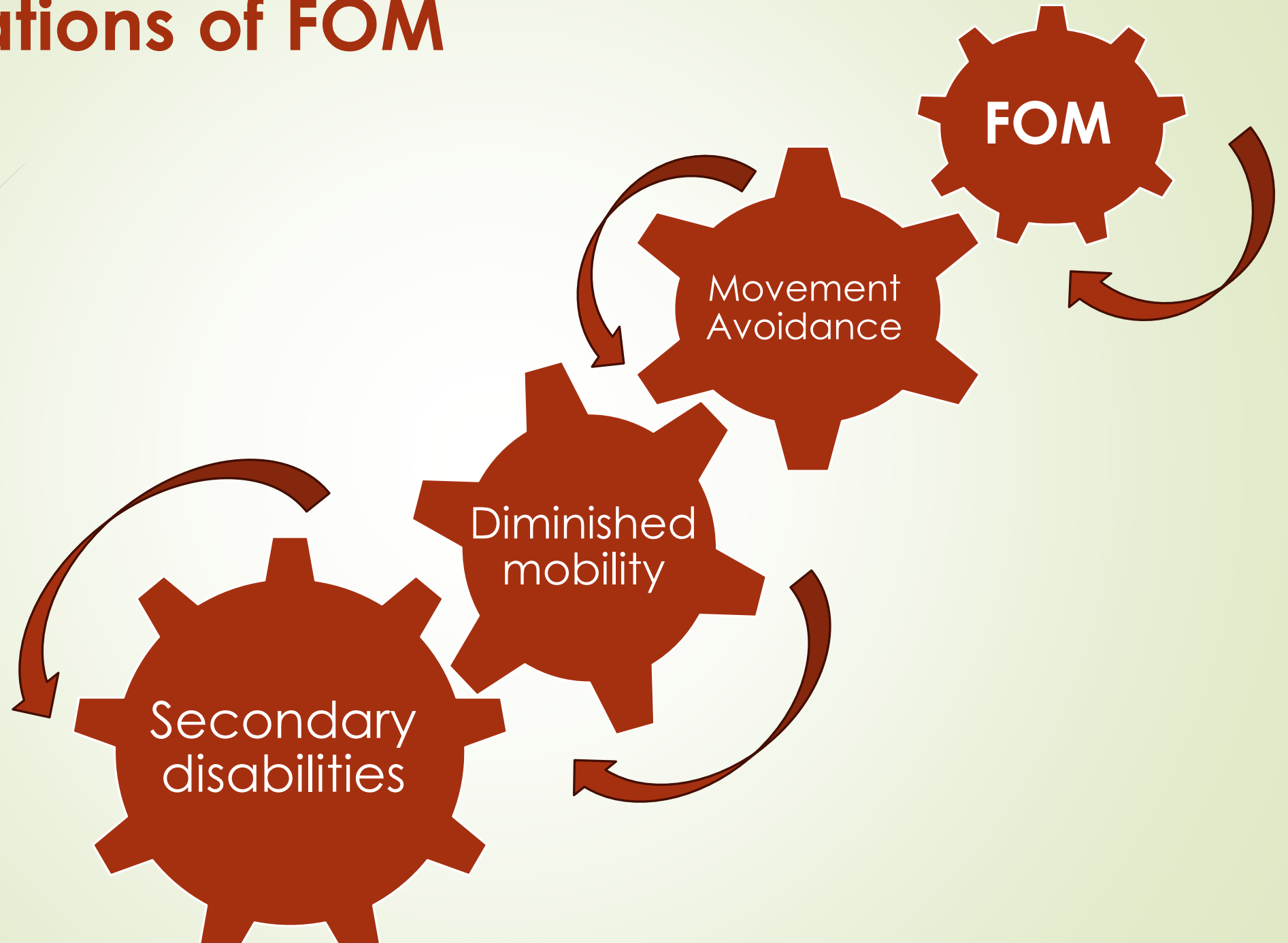
Conclusions:

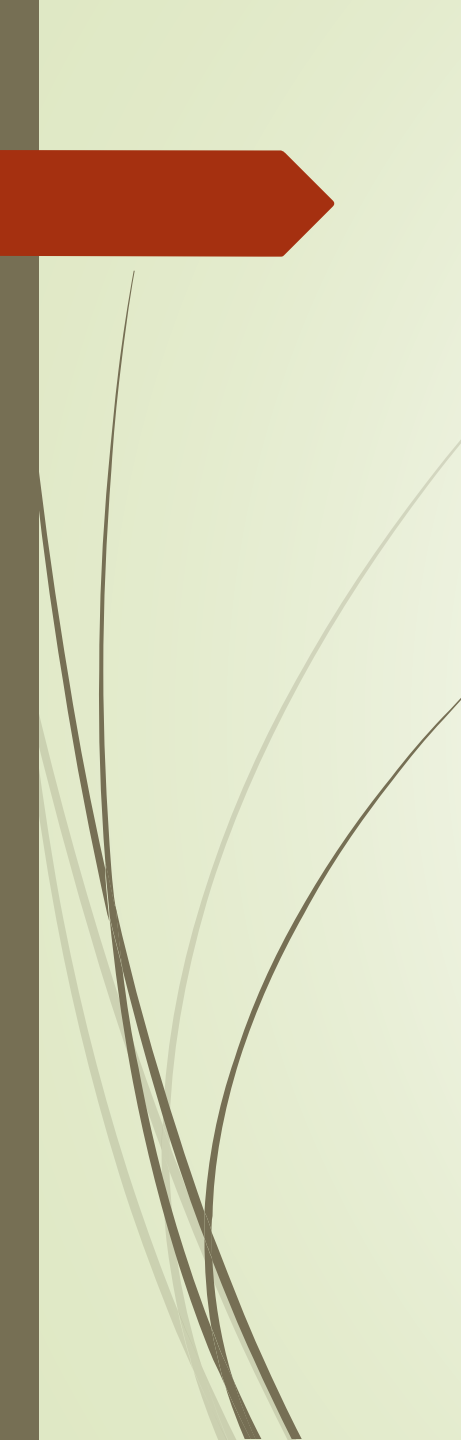
- The results suggest that fear of movement in individuals with RTT stems from misinterpretation of messages from the proprioceptive system –
- Therefore, activation of body and limbs is required from a young age.




Ramifications of FOM

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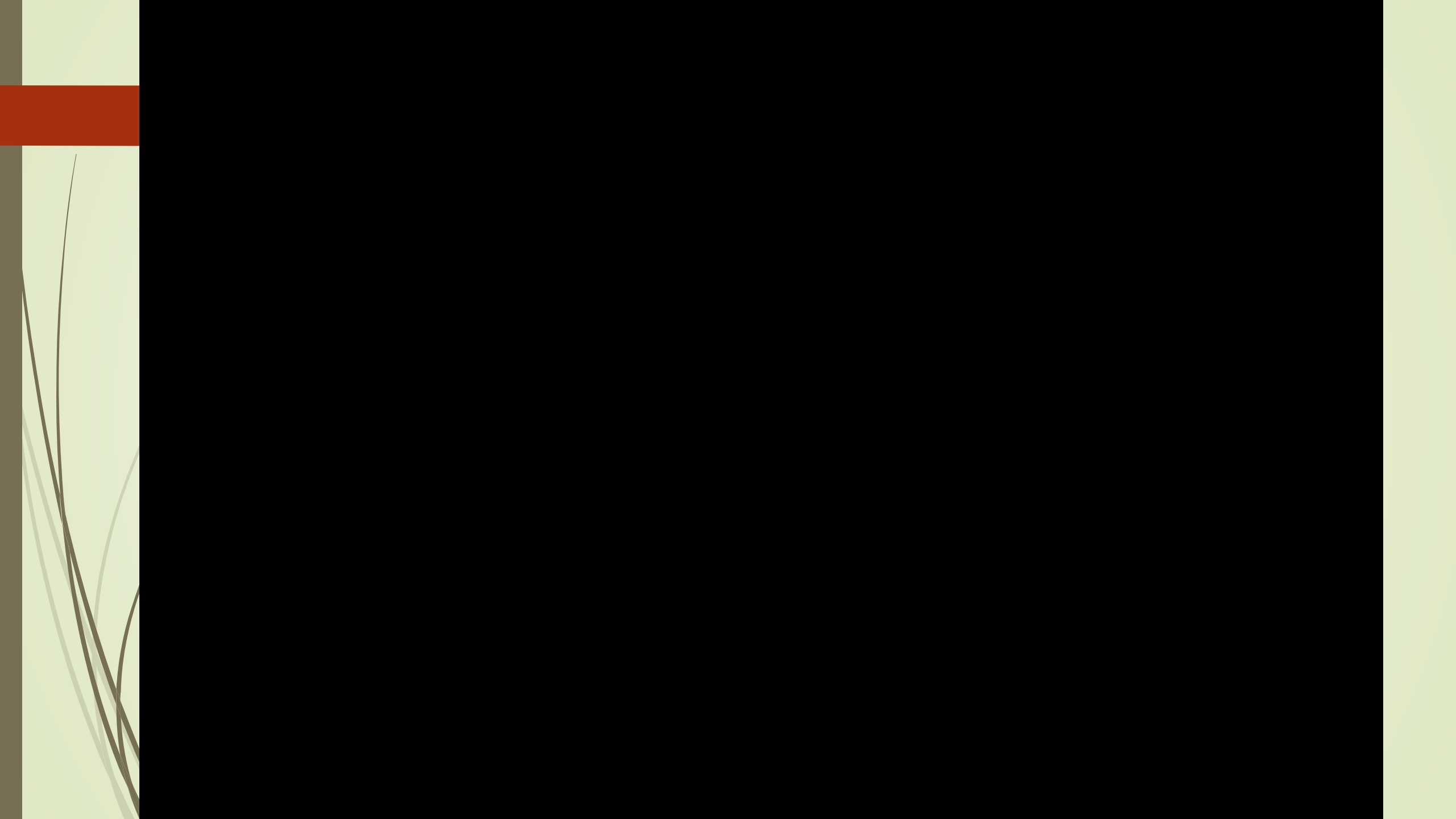
Clinical suggestions to enhance motricity in Rett syndrome

Get used to movement

- ➡ Make a lot of extremities movements,

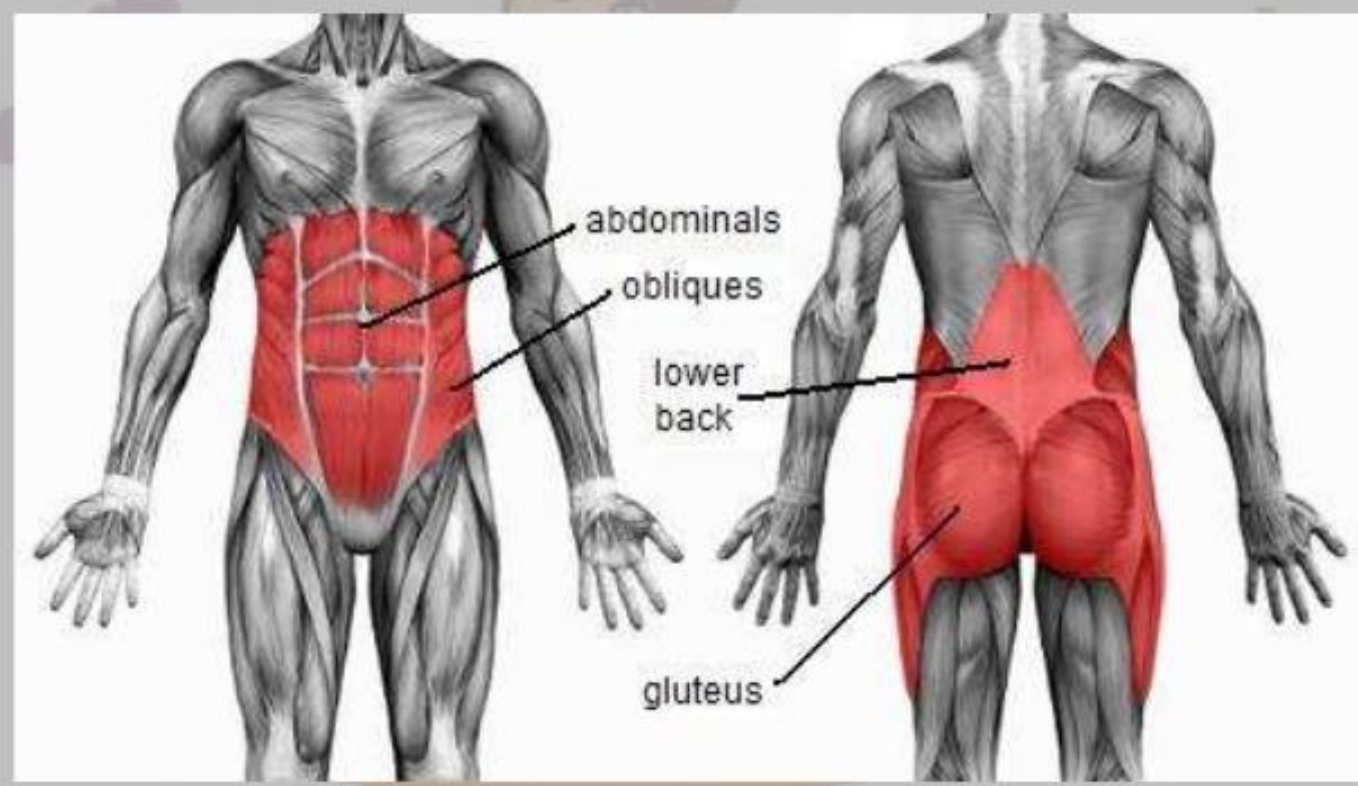
Make movement a joyful experience I do with love

- ➡ Dance with your child on your hands, on your shoulders,
- ➡ Dance with your daughter standing in front of the TV, with songs and video she likes
- ➡ Use the UpSee



Core muscles and their importance in Rett syndrome

Core Muscles



New

New

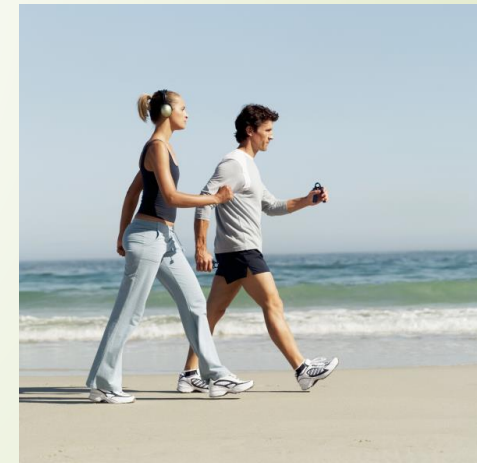
How do we move/Function?

We stabilize using core muscles!!!!

It's termed "Dynamic stabilization"

How long does it take us to master dynamic stabilization?

- Children gain walking ability around one year of age
- They reach adult-like control of walking at age 7 years

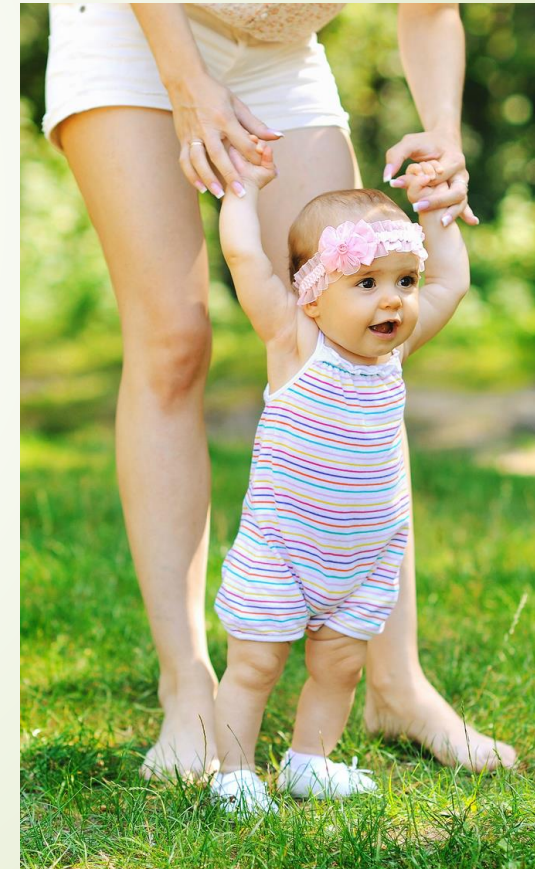


(Sutherland et al., 1980)

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What will you do when you want to move but have?

- Fear of movement
- Poor proprioceptive reception
- Anxiety
- Over-active sympathetic system
- Abnormal muscle tone
- Not enough training
- External point of support



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You fixate (instead of dynamically stabilize)



What is the solution?

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- Good control of trunk and pelvic movements is necessary for well controlled movements, required to perform activities of daily living .
- Children with a disability (Rett syndrome) have reduced ability to modulate muscle coupling.

The good thing about it

- Movement patterns of the pelvis can be improved by training
- (Barton, et ,al,. 2013)

What is the solution?

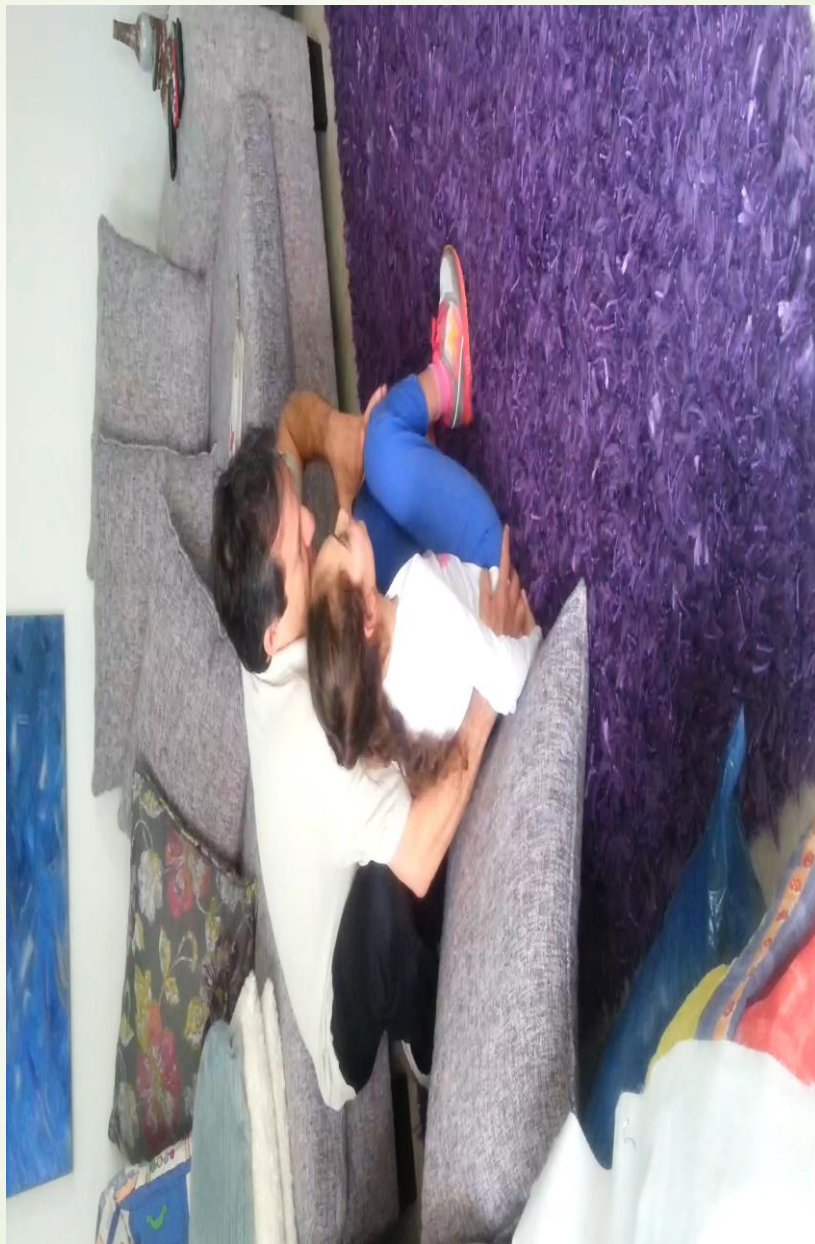
- Stability exercises, are recommended to improve proprioceptive capabilities
- Performing exercises on unstable surfaces, are recommended to improve reactive capabilities, which may reduce injuries (Willardson, 2007)
- Changes are maintained after the intervention ceases !!
(Shurtleff, Standeven, & Engsborg, 2009)



The solution:

**Functional Alignment
Core Training (FACT)**

L.I. Before Training



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L.I. After Training



Walking before and after intervention

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Parameters	Before	After
Stability	Not stable	More stable
Base	Too Narrow	Wider
Leg movements	Mild	Moderate
Step through/to	Step to	Step through
Walking speed	Slow/ difficulty initiating	Faster

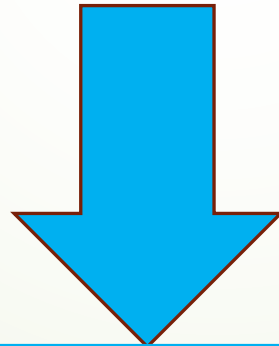
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L.I. During Training



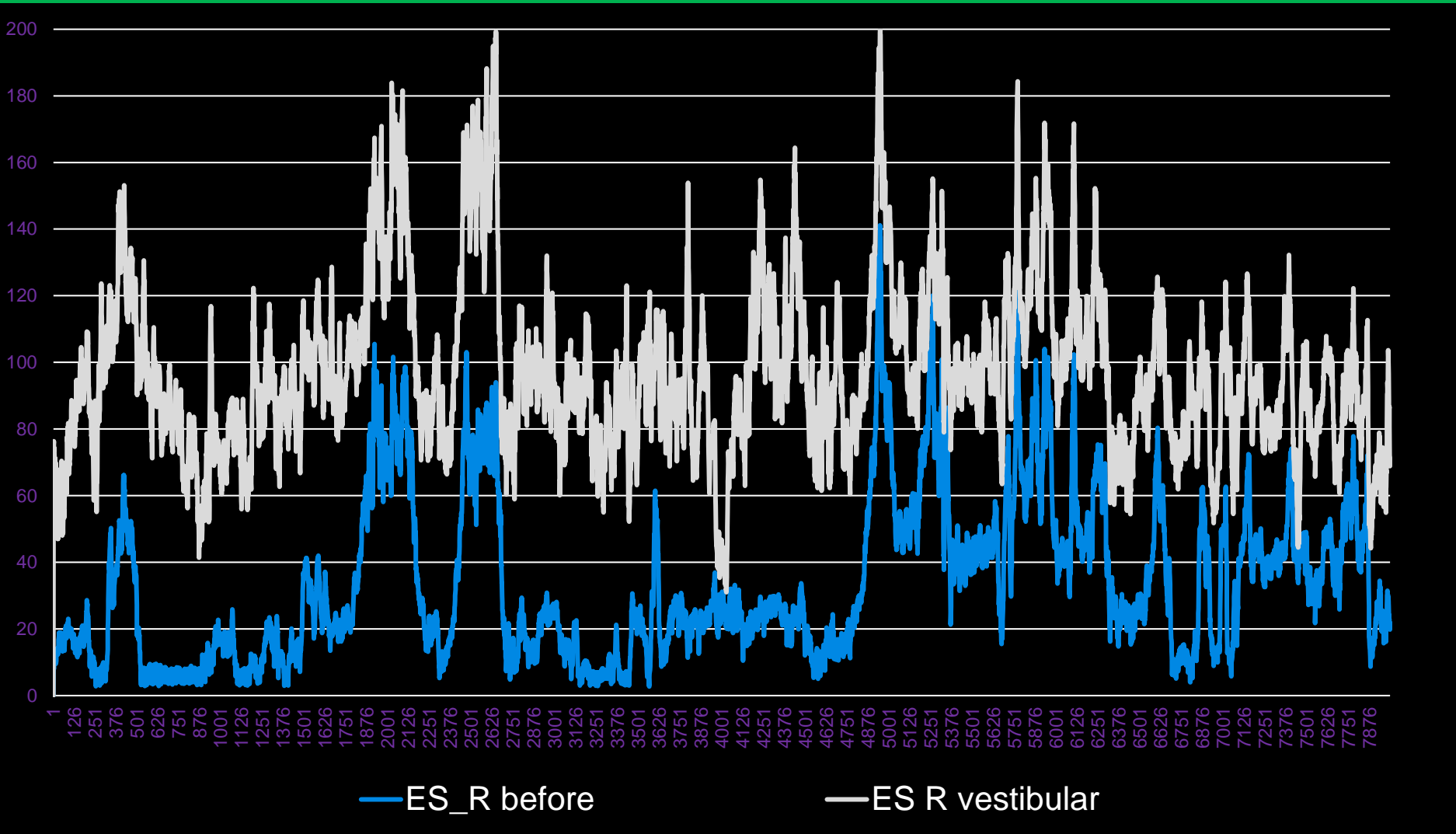
What changed?

- Stability?
- Muscle coordination?
- Core muscles activity?
- Concentration ?



EMG measurements

Back extensors before and during activity in VP



Mean Erector spine activity :

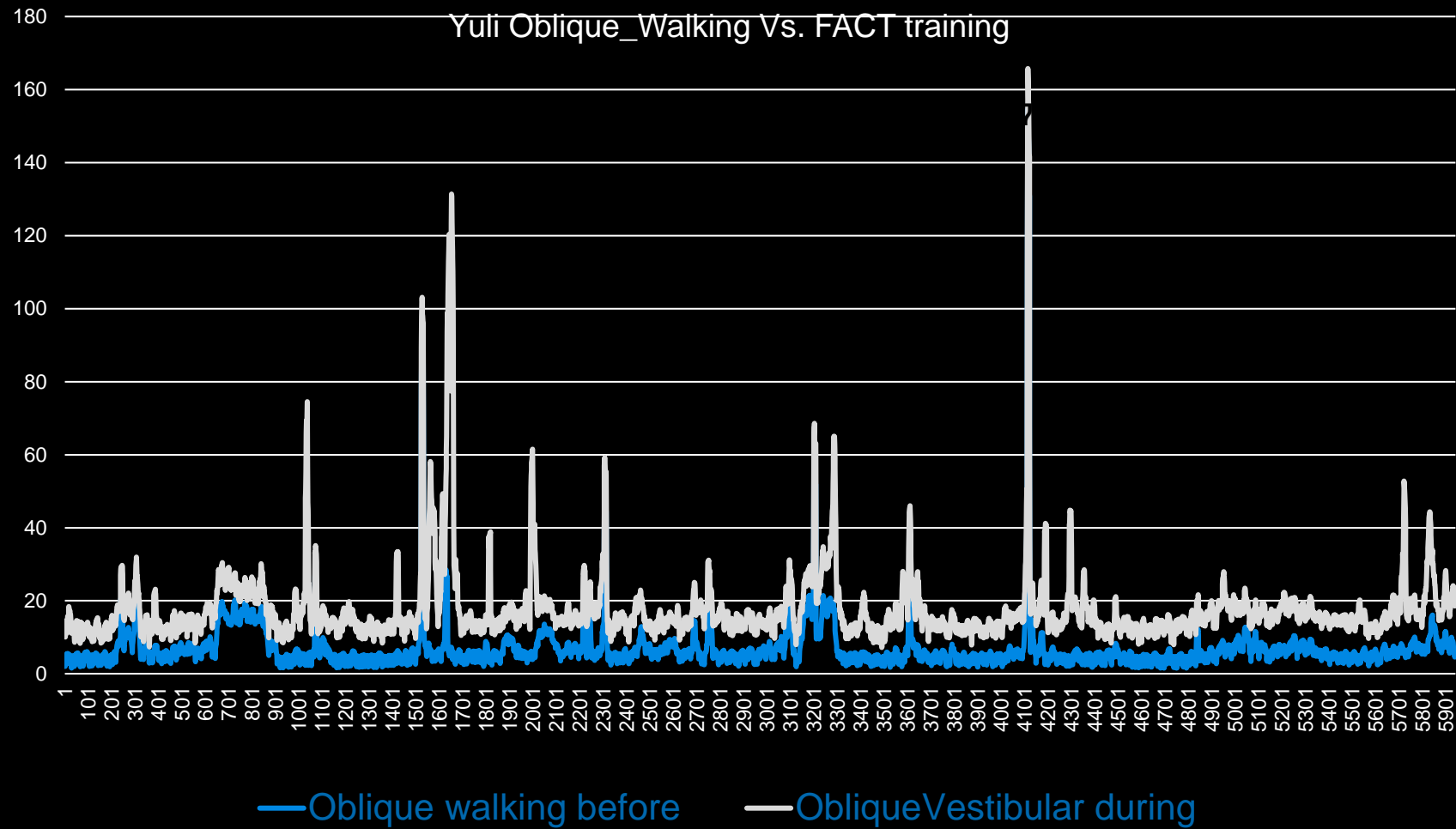
Before training : 33.68

During training: 63.2

P value ≤ 0.0000



Oblique muscles activity before and during VP



Mean oblique muscles activity :

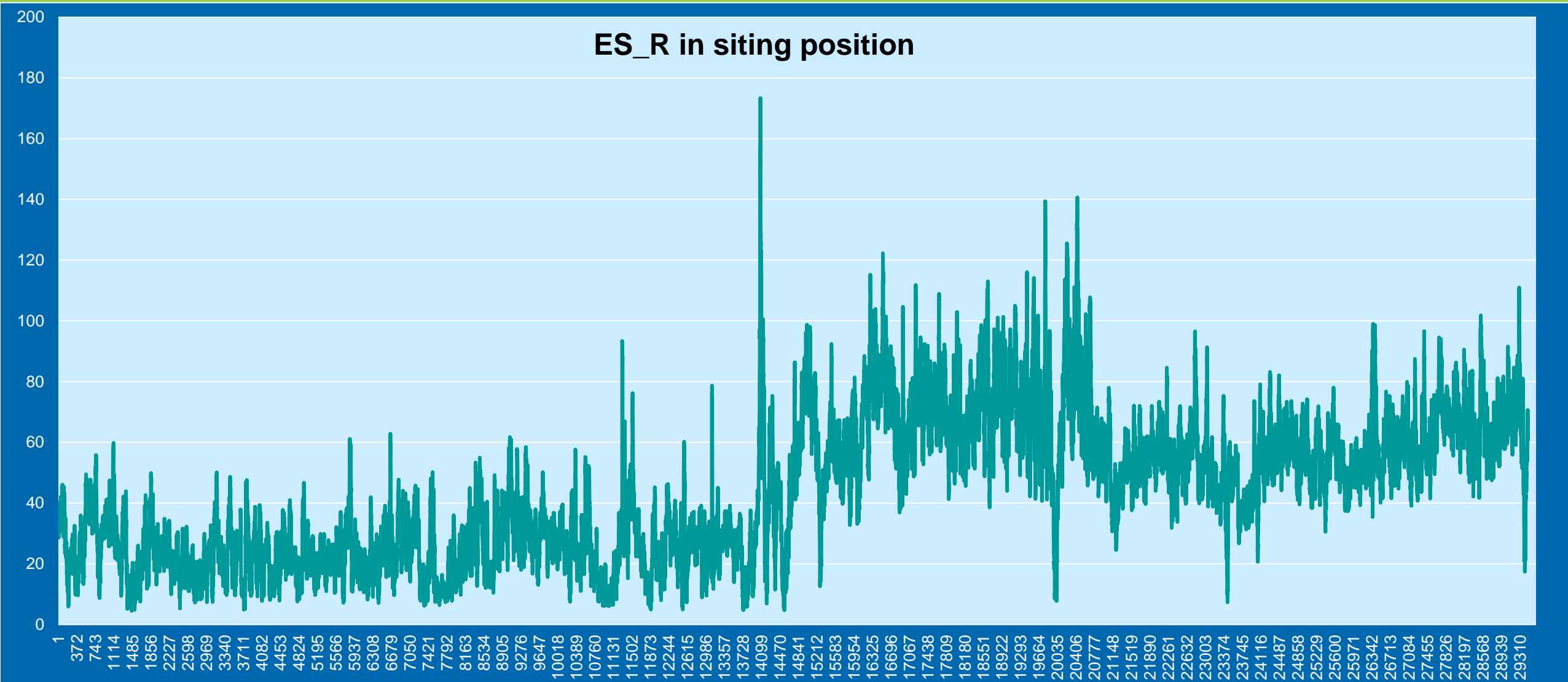
Before training : 7.44

During training: 10.7

P value ≤ 0.0000



Muscle activity during intervention inside VP



Average activity level of core muscles before and during exercise

Muscles	Before	During	T test Significance
Erector spine R	33.68	63.2	$P \leq 0.0000$
Erector spine L	20.9	29.9	$P \leq 0.0000$
Oblique	7.4	10.7	$P \leq 0.0000$
Gluteus mx R	20.6	26.6	$P \leq 0.0000$
Gluteus mx L	4.34	5.27	$P \leq 0.0000$

Results

- Improvement in quality, speed, and coordination was found when walking after practice within the VP.
- Activity on a vestibular plate activates core muscles ***automatically***, in comparison to rest situation
- The activity of the core muscles is enhancing with practice
- Improvement in bowl control was found as well

This method is available to....

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This method is available to....



On all fours

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This method is available to....

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The solution: **F**unctional **A**lignment **C**ore Training (FACT)

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Summation

- Individuals with RS have poor proprioceptive input
- This leads to obscure body scheme causing apraxia
- It also leads to fear of movement (and movement avoidance)
- This leads to abnormal fixating pattern of movement instead of “normal” (more functional) forms of stabilization

Therefore:

- A core training stability movement program should be implemented from a young age to prevent fear of movement and fixations from developing

Summation

- UpSee is an option for small girls who are unable to walk
- It can help in experiencing movement as a positive experience by reducing fear of movement
- This device can be used as part of physical therapy sessions

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Take home message

- Physical activity is essential for individuals with Rett syndrome from a young age
- It is important to construct an activity program as preventive medicine for individuals with RTT.
- It has to be adjusted to the child/family's needs and abilities
- Fear of movement due to misinterpretation of incoming proprioceptive sensory signals is typical of RTT

Take home message

- This difficulty in understanding own body messages leads to:
- Reduced movement,
- Enhanced dependency on others,
- Body parts fixation,
- Reduced activity level and
- Reduced participation.

Take home message

- Core muscle training is:
- Easy to perform at **all functional levels**
- Can be performed **everywhere**
- **Unexpensive**
- Reduces **fear of movement**,
- **Prevents fixations of body parts**
- Prevents the development of **pathological movement patters (Scoliosis)**

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**Thank you for
listening**

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