Physical Therapy Evaluation & Treatment of Cerebellar Ataxia

Session Time: 13:30-15:30, Saturday, October 8, 2021

Amy Wedge, PT, DPT
Board Certified Clinical Specialist in Neurologic Physical Therapy
Certified Brain Injury Specialist
LSVT BIG® & PWR!Moves® Certified Therapist
Department of Physical Therapy
East Carolina University

Andrew Newnam, PT, DPT
PWR!Moves® Certified Therapist
Department of Rehabilitation Services
Vidant Medical Center

Objectives
1. Understand cerebellar ataxia linking symptoms to neuroanatomy.
2. Discuss specific physical therapy evaluation measures for cerebellar ataxia based on current research.
3. Provide examples of specific clinical interventions for cerebellar ataxia based on available evidence.
4. Utilize a case study to further improve understanding of translating cerebellar ataxia research into clinical practice.

What is Ataxia?
➢ “Incoordination of movement following damage of the sensory or cerebellum system.”—Bastian AJ, 1997
➢ Greek — “disorderly” or “without order”
➢ Commonly associated with disease pathology of the cerebellum, but it can stem from other areas

Role of Cerebellum
➢ Main Functions
   ➢ Coordination
   ➢ Movement quality

➢ Other Functions
   ➢ Motor learning
   ➢ Motor control
   ➢ Automaticity
   ➢ Posture
   ➢ Balance
   ➢ Cognition

Motor Control and Motor Learning

Motor Control
➢ Ability to regulate mechanisms essential for movement
➢ Involves initiating, directing, and grading movement
➢ Process by which a new motor task is acquired

Motor Learning
➢ Requires practice
➢ Cerebellum plays a critical role
➢ Levels of mastery (automaticity)
➢ Optimization facilitates outcomes

Disclosures
➢ None, no conflicts of interest

What is Ataxia?
➢ “Incoordination of movement following damage of the sensory or cerebellum system.”—Bastian AJ, 1997
➢ Greek — “disorderly” or “without order”
➢ Commonly associated with disease pathology of the cerebellum, but it can stem from other areas
Motor Learning after Cerebellar Lesion

Vestibulocerebellum:
- Balance
- Postural adjustments
- Coordination of eye movements

Spinocerebellum:
- Muscle tone
- Coordination

Cerebrocerebellum:
- Motor planning
- Learning
- Memory

Cerebellar Region Function Motor Pathways Influenced
Lateral hemispheres Motor Planning for extremities Lateral corticospinal tract
Intermediate hemispheres Distal limb coordination Lateral corticospinal tract, rubrospinal tract
Vermis and Flocculonodular lobe Proximal limb and trunk coordination Anterior corticospinal tract, reticulospinal tract, vestibulospinal tract, tectospinal tract
Balance and vestibulo-ocular reflexes Medial longitudinal fasciculus

Principle Signs of Cerebellar Dysfunction
- Ataxia
- Appendicular
- Truncal
- Hypotonia
- Tremor
- Ocular motor abnormalities
- Coordination impairments
- Dysarthria
- Dysesthesia
- Dysynergia
- Dysdiadochokinesia
- Loss of check (rebound)
### Vestibulocerebellar Functions
- Enhances balance by coordinating postural adjustments (anticipatory and compensatory)
- Controls eye movements via VOR's
- Dysfunction:
  - Imbalance
  - Dis-equilibrium
  - Nystagmus
  - Truncal ataxia
  - Significant swaying during standing, staggering during gait
  - Fall

### Spino cerebellum
- Coordination of synergistic movement patterns needed for specific task
- Particularly movements of proximal limb and trunk
- Detects and aids in adjustment of movements relative to desired vs actual
- Dysfunctions:
  - Dysnergia
  - Dysmetria
  - Lock of movement check (rebound)
  - Ataxia of gait and stance

### Cerebrocerebellum
- Planning, coordination, and temporal sequencing of movement of the distal extremities (hand)
- Dysfunction:
  - Disrupts motor planning and prolongs reaction time
  - Decomposition of movement
  - Dysdiadochokinesia
  - Hypotonia
  - Dysarthria

### Types of Ataxia
- Genetic Ataxia
- Acquired Ataxia
- Idiopathic Ataxia
- Sensory Ataxia
- Episodic Ataxias

### Genetic Ataxia
- Autosomal Dominant
  - Spinal Cerebellar Ataxia (SCA)
  - Dysfunction of cerebellum & brainstem
  - Signs / Symptoms:
    - Ataxia
    - Extrapyramidal features
    - Parkinsonism features
    - UMN signs
    - Cognitive Impairments
  - Prognosis:
    - SCA1, SCA2, SCA3 = mid 50's
    - Other SCA's = normal lifespan

- Autosomal Recessive
  - Friedreich Ataxia
  - Degeneration of peripheral nerves and nerve fibers in SC
  - Signs and symptoms:
    - Ataxia
    - Neuropathy
    - Constitutional symptoms
    - Vision loss
    - UMN & LMN signs
    - Cardiovascular features
    - Cognitive features
  - Prognosis:
    - 40-50 years of age
Acquired Ataxia

- Damage directly to cerebellum or its pathways
- Stroke
- Tumor
- Trauma

Vascular Areas

- PICA territory
- AICA territory
- SCA territory

Key

Clinical Significance

- Lateral hemisphere lesion
  - Symptoms ipsilaterally to UE / LE
  - Delayed initiation of movement
- Vermis lesions
  - Incoordination to the head and trunk
- Occlusion of PICA
  - Wallenberg syndrome (Lateral Medullary Syndrome)
  - Ipsilateral Horner syndrome

Idiopathic Ataxia

- General Diagnostic Criteria
  - Sporadic
  - Insidious onset
  - Can be progressive
  - Cerebellar atrophy

- Subtypes*
  - Multiple Systems Atrophy – C
  - Idiopathic Late Onset Cerebellar Ataxia
  - Toxin-induced cerebellar ataxia
  - Multiple Sclerosis
  - Vitamin E deficiency

* Not an all-inclusive list

Sensory Ataxia

- Damage to dorsal columns and dorsal root ganglia
- Signs & symptoms:
  - Impaired proprioception
  - Impaired vibratory sense
  - +Romberg sign with EC
  - Stomping gait

Romberg's Test

Population | Eyes Open | Eyes Close
---|---|---
Healthy individuals | Stable | Stable
Sensory Ataxia | Stable | Unstable
Cerebellar Ataxia | Unstable | Unstable

Diagnosis

- Genetic testing
- Brain Imaging
  - CT
  - MRI

Prevalence

- Rare disease
- Cerebellar Ataxia: 26/100,000
- SCA has a prevalence of 2.7/100,000 worldwide
- 47 SCA subtypes have been identified, and the number of genes implicated in SCAs is continually increasing
Clinical Significance Summary

➢ Ataxia is ipsilateral to the side of the cerebellar lesion
➢ Midline lesions of the vermis or flocculonodular lobes cause truncal ataxia and oculomotor abnormalities
➢ Lesions of the intermediate part of the cerebellar hemisphere cause appendicular ataxia
➢ Ataxia is often caused by lesions of cerebellar circuitry in the brainstem or other locations rather than the cerebellum
➢ Because of connections between the cerebellum and vestibular system, cerebellar lesions are often associated with vertigo, nausea, vomiting, and nystagmus

PT Assessment & Evaluation

Cerebellar Ataxia

Body Structure / Body Function
- Cerebellar degeneration
- Coordination impairments
- Dyssynergia
- Dyssynergia
- Decomposition of movement
- Oculomotor abnormalities (nystagmus)
- Dysarthria
- Fatigue
- Loss of automaticity
- Decreased gait speed
- Gait ataxia
- Impaired motor control / motor learning
- Decreased postural control (postural instability / imbalance)
- Hypotonia
- Tremor

Activities
- Bed mobility
- Walking on various terrains
- Stepping over obstacles
- Managing an assistive device
- Difficulty maintaining sitting and standing balance during functional tasks
- Transfers
- Fine motor tasks

Participation
- Domestic life
- Interpersonal relationships
- Community access
- Work capacity
- Leisure activities
- Quality of life

Falls in Cerebellar Ataxia

➢ 73-93% of patients with SCA reported at least 1 fall during a 1-year period
➢ Increased gait variability associated with history of falls
➢ Can occur in any direction
➢ Fear of falling is seen in 42% of patients with SCA
➢ Increased severity & + pyramidal signs = increased rate of falls

Balance Disorders in Cerebellar Ataxia

➢ Postural Sway
➢ Reactive Postural Adjustments
➢ Anticipatory Postural Adjustments
➢ Multisensory Changes

Balance

Postural Control

➢ Regulation of the body’s orientation to gravity
➢ Product of proper sensory integration and motor control
➢ Cerebellum plays a crucial role in postural control
➢ Impaired postural control increases fall risk
➢ Attentional needs in postural control
Gait Disorders in Cerebellar Ataxia

➢ Decreased gait speed & step length
➢ Increased base of support
➢ Increased AP and lateral trunk displacement
➢ Decreased knee flexion
➢ High variability
➢ “Protective walking”

Oculomotor Dysfunction with Gait

➢ Oculomotor control allows gaze stabilization
➢ Saccadic eye movement is required for accurate foot placement
➢ Downbeat nystagmus impair vertical gaze stabilization
➢ Decreased gait velocity & stride length
➢ Increased BOI, double-support time
➢ Unilateral cerebellar lesions
➢ Ocular tilt reaction
➢ Changes in subjective visual vertical

Core Outcome Measures

➢ Berg Balance Scale
➢ Functional Gait Assessment
➢ Activities-Specific Balance Confidence Scale
➢ 10 Meter Walk Test
➢ 6 Minute Walk Test
➢ 5 Times Sit to Stand

Standardized Clinical Measures

International Cooperative Ataxia Rating Scale (ICARS)
➢ Assesses: Posture and gait disturbances, kinetic functions, speech disorders, oculomotor disorders
➢ Higher scores = increased impairment
➢ Psychometrics: excellent test-retest reliability, inter- and intra-rater reliability, adequate validity and floor effects

Scale for the Assessment and Rating of Ataxia (SARA)
➢ Assesses: Gait, stance, sitting, speech disturbance, finger-chase, nose-finger test, dysdiadochokinesia
➢ Higher scores = increased impairment
➢ Psychometrics: excellent test-retest reliability, inter- and intra-rater reliability, internal consistency and validity, adequate floor effects

Medical Interventions

➢ There is currently no cure for SCA
➢ Currently no pharmacologic therapies to stop disease progression
➢ Physical therapy is the current cornerstone of treatment for this condition
➢ Appropriate referrals to other interdisciplinary team members
What Does the research tell Us?

➢ Research is limited
➢ Currently relies heavily on case studies
➢ PT intervention has proven to be effective
➢ Specific interventions have proven effective in similar populations (stroke and Parkinson’s)

Principles of Neuroplasticity

1. Use it or Lose it
2. Use it and Improve it
3. Specificity
4. Repetition Matters
5. Intensity Matters
6. Time Matters
7. Salience Matters
8. Age Matters
9. Transfer or Generalization
10. Interference

Motor Control & Motor Learning

Principles to consider:
➢ Guided vs Independent practice
➢ Type of feedback
➢ Type of practice
➢ Internal vs external focus
➢ Progression
➢ Promoting autonomy
➢ Transfer of learning
➢ Aerobic priming

Aerobic Priming

➢ Facilitates motor learning
➢ Enhanced cognitive flexibility
➢ Increased motor cortex excitability
➢ Specific benefits demonstrated in cerebellar ataxia population
➢ Examples: Cycling, HIIT, Treadmill training

Training Intensity/Challenge

➢ May slow neurodegeneration
➢ Increases brain neurotrophic proteins (BDNF)
➢ Intensive therapy for 2 hours/day for 5 days/week reaps functional benefits and decreased ataxia
➢ MUST Challenge Patients
➢ HIIT training proven beneficial in post-stroke and Parkinson’s populations
➢ Higher intensity improves task motor performance

Automaticity

➢ High practice dosage needed for automaticity
➢ Cerebellum is critical in transferring attentionally demanding task to automatic task
➢ Task automaticity allows for less attention and more transferability
➢ Decreased interference with other tasks
➢ Poor sustained attention increases fall risk
➢ Increased ability to dual task
Task Specificity

- Has been linked to successful and long-lasting functional recovery in stroke
- Task specific training increases automaticity
- Can transfer to similar tasks
- May lead to more cortical reorganization vs high intensity
- Shown to enhance: sensory retraining, gait training, motor training of the upper extremities

External Focus

- Attentional focus is known to influence both performance and learning
- Benefits well documented in healthy population (Wulf)
- External cueing increases body position/ awareness
- Reduced sway and falls in small Parkinson's population vs no cueing or internally focused
- One study in stroke population indicates internal focus may be detrimental to learning

Variability

- Task variability improves information retention and translation
- PT approaches that incorporate a variety in approaches have been proven effective

Aerobic Exercise

- May alter disease progression in Alzheimer's and Parkinson's
- May improve postural stability, balance, and tremor in Parkinson’s Disease
- Decreased ataxia severity, improved balance, improved cardiovascular fitness, and improved gait in degenerative cerebellar disease

Postural Control Interventions

There is now moderate level evidence that rehabilitation is efficient to improve postural capacities of patients with cerebellar ataxia – particularly in patients with degenerative ataxia

- Virtual reality
- Exergame training
- Proprioceptive training
- Tai Chi
- Whole body vibration
- Torso weighting

Postural Strengthening

- Contraction of trunk muscles provide stable foundation
- Sequencing: transverse abdominis is first muscle activated prior to hip movement
- Persons with cerebellar ataxia can demonstrate truncal ataxia
- Postural strengthening can decrease UE support and reduce trunk flexion during activity
- Postural strengthening shown to improve dynamic sitting balance
- Possible carryover into improvements in standing balance, gait and ADLs
Core Strengthening for Cerebellar Ataxia

- Individuals with progressive cerebellar ataxia had significantly improved BESTest scores
- Reduction in LBP
- Increased contraction of pelvic floor muscles
- Should be used in adjunct with a traditional balance program

Balance Interventions

- Should focus on both static AND dynamic
- Can decrease postural sway and reduce fall risk
- A home balance program improved gait speed, stride length, double support phase and DGI in 14 cerebellar ataxia patients

Perturbations

- Corrective motor behaviors have not been expected to be affected by cerebellar damage
- 2019 study by Aprigliano F., et al. assessed corrective balance reactions in cerebellar ataxia population vs healthy controls
- Revealed a difference in corrective reactions to perturbations
- Cerebellar ataxia group also demonstrated capacity to modify gait towards safer behavior

Gait Interventions

- Gait training using partial BWS significantly increased gait performance
- Auditory cueing decreases gait variability in temporal and spatial gait parameters
- Improved BBS, FAC scores, and 10 MWT when combined with trunk stabilization with cerebellar ataxia
- Challenging speeds lead to maximized gait speed
- Both over ground gait training with therapist assistance and robotic assisted gait training were found to be equally as effective in improving gait in adults with ataxia.

Coordination training

- Benefits of coordination training have been linked to cerebellar ataxia
  - Improved motor performance
  - Reduction in ataxia symptoms
  - Improved gait velocity
  - Decreased lateral sway in gait
- Frenkel Exercises proven program designed for treatment of ataxia
  - Reduce fall risk in elderly
- Continuous training can lead to long term benefits

Frenkel Exercises

- Series of motions with increasing difficulty designed for patients with ataxia, specifically cerebellar ataxia, to improve coordination
- Performed in supine, sitting, standing, and walking
- For those with sensory ataxia, may be good to try these exercises with eyes closed
- Does not help with strengthening
- Patient must be able to complete an exercise correctly without excessive fatigue to be able to progress to the next exercise
- Perform each exercise at a slow, even tempo
**Resistance Training**
- Improved muscular strength and power
- Muscular power is often a focus (MS and Parkinson’s Disease)
- Can cause changes within the nervous system
- Effects on gait are unclear
- Training should be progressive
- Should be incorporated into functional exercises to improve performance

**Oculomotor Interventions**
- Gaze stabilization exercises can modulate eye movements with respect to head movements
- Significant improvements in stance time and walking speed in those with PSP
- Vestibular habituation training in combination with strengthening, stretching, and gait re-education lead to improvements in disability scales for those with cerebellar ataxia

**Weighting and Ataxia**
- Benefits of weighted vests in the treatment of ataxia is disputed
- Balance-based tono-weighting shows some promise in increasing stability
  - Weight placed in the opposite direction as the patient’s typical LOB
- Use of ankle weights have also showed promise
  - Improved functional mobility, gait and independence

**Balance Based Weighting**
- Sway area generally decreased
- Possible method for increasing static stability in cerebellar ataxia
- No carry over demonstrated in gait

**Trunk stabilization and Locomotor Training (Case study)**
- Single subject:
  - 23 yo male
  - Sustained severe TBI
  - Severe truncal and extremity ataxia
- Intervention:
  - 28 sessions (10 weeks) focused on Trunk stabilization and Locomotor training
  - LT performed on Litegait and overground
  - Trunk stabilization performed in a variety of positions and during functional activities

**Outcome**

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Gait, balance, and self-reported functional outcome measures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Outcome measures</td>
<td>PSE-1</td>
</tr>
<tr>
<td>10 Meter Walk Time (sec)</td>
<td>Unable</td>
</tr>
<tr>
<td>Functional Ambulation Category</td>
<td>2</td>
</tr>
<tr>
<td>Berg Balance Scale</td>
<td>570</td>
</tr>
<tr>
<td>Timed Up &amp; Go Score (sec)</td>
<td>3</td>
</tr>
<tr>
<td>OPTIMAL—Difficult</td>
<td>62</td>
</tr>
<tr>
<td>OPTIMAL—Confidence</td>
<td>48</td>
</tr>
</tbody>
</table>

PSE-1=Pre-Intervention, 2=Post-Intervention week 2, 3=Post-Intervention week 3, PSE-1=Pre-Intervention, NC=not completed.
*PSE-1 weeks 2,3,9 mean (sd): 5.33 (1.86), **Mean±SD=2 standard deviations from PSE-1 mean.
Key Points

➢ Relative contributions of study interventions and outpatient PT cannot statistically be determined

➢ The subject made no improvements in his pre-l measures when he was receiving only outpatient PT

➢ Trunk stabilization exercises used in conjunction with LT may be effective in improving gait and balance in a subject with severe ataxia.

Case Study

Case Introduction

Demographics:
➢ 73-year-old male
➢ Past medical history:
  ➢ Pituitary adenoma
  ➢ Nondrinker
  ➢ Nonsmoker

Brain MRI taken on 7/30/2018, 6/18/2019 revealed progressive cerebellar vermis atrophy

➢ Did not receive genetic testing.

Diagnosed with cerebellar ataxia in 2019

Most likely genetic cause given lack of parkinsonism and image findings.

Initial Evaluation (Subjective)

➢ Chief complaint: Impaired balance that has progressed over the last year

➢ Multiple falls

➢ Noted difficulty with dressing (buttons) and eating due to UE discoordination

➢ Arm tremor when reaching

➢ Oscillating tremor upon standing

➢ Caregiver: Wife

Initial Evaluation (Objective)

➢ Coordination: Noted dysmetria L>R

➢ 5/5 gross strength (with exception of hip Ext: 3/5 B)

➢ Requires unilateral UE support when completing dynamic seated task

➢ Increased sway in static standing

➢ SLS: Unable B

➢ Bed mobility: independent (slowed)

➢ Transfers: Heavy UE use, supervision

➢ Stairs (3, 4” steps): Step to pattern with B UE support and supervision

➢ Gait: Hypometric, variable, unstable, uses rollator

Tracking disease progression

➢ “The Posture and Gait (PG) sub-component of the International Cooperative Ataxia Rating Scale (ICARS) demonstrates the most robust psychometric properties with acceptable clinical utility.”

<table>
<thead>
<tr>
<th>Subscale</th>
<th>Items</th>
<th>Weight</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postural and gait disturbances</td>
<td>7</td>
<td>34 (12 for gain, 22 for stance)</td>
</tr>
<tr>
<td>Limb ataxia</td>
<td>7</td>
<td>52</td>
</tr>
<tr>
<td>Dysarthria</td>
<td>2</td>
<td>8</td>
</tr>
<tr>
<td>Oculomotor disorders</td>
<td>3</td>
<td>6</td>
</tr>
</tbody>
</table>
Outcome Measures Assessed (Initial)

- 5xSST: 21.95 sec (12.6 norm for age)
- Gait speed: P: 0.42 m/s, F: 0.5 m/s fast with rollator
- TUG: 24.22 sec
- TUG(cog): 28.47 sec
- Dual task Cost: 17.5%
- ABCs: 33.75%
- International Cooperative Ataxia Scale: 32/100
- BERG: 18/56 (High fall Risk)
- mCTSIB
  - Firm: EO: 30 sec, EC: 30 sec
  - Foam: EO: 3 sec, EC: unable

Treatments were designed to...

- Challenge/improve limits of stability
- Challenge anticipatory postural control
- Strengthen postural stabilizers
- Increase walking automaticity
- Challenge coordination/control in upper and lower extremity
- Increase stability in gait

Interventions for Rudy

- High intensity gait training on Treadmill with Litegait™
- Overground gait training
- Static stability & dynamic balance training
- Proximal strengthening (core and scapular stabilizers)
- Functional LE strengthening
- Bed mobility/Transfer training

Motor Control Principles Used

- Based on the progressive nature of the disease we may expect function to decline
- Most outcome measures indicate an improvement in function:
  - Decrease in risk for fall
  - Increased functional strength
  - Increased functional mobility

Compensatory Strategies

- Allows patients to regain some PLOF if they are not able to return to normal/prior movement patterns
- Widening gait stance
- Assistive device
- U-Step Walker
- Slow down movements
- Decrease the degrees of freedom
- Remove distractions
The Need for Further Research

- Growing body of evidence supporting the efficacy of neurorehabilitation in the treatment of genetic degenerative ataxia.
- High quality research necessary to determine optimal:
  - Intensity
  - Duration
  - Type

References


The Need for Further Research

- Growing body of evidence supporting the efficacy of neurorehabilitation in the treatment of genetic degenerative ataxia.
- High quality research necessary to determine optimal:
  - Intensity
  - Duration
  - Type

References
