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Examination and Treatment of Ataxia Following Cerebellar Damage

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Objectives

1. Describe the cerebellum's role in coordination, motor learning, postural control, balance reactions, and gait.
2. Identify the etiologies and mechanisms of ataxia.
3. Identify valid and reliable outcome measures for ataxia.
4. Identify the best practice interventions for ataxia
5. Outline an appropriate, evidence-based plan of care for patients with ataxia.
Ataxia Defined

- From Greek word for “disorderly”

- Incoordination of movement following damage of the sensory or cerebellar system

- Disease affecting the cerebellum typically cause ataxia, coupled with dysmetria and tremor

Poll Question 1

- When a person has ataxia, we would expect significant weakness in the muscles involved.
  - A. True
  - B. False
Poll Question 2

- In a person with ataxia, we would expect which of the following?
  - A. Hypertonicity
  - B. Hypotonicity
  - C. Normal muscle tone

Tremor

- Rhythmic, involuntary, oscillating movement of the body part

- 2 classifications
  - Resting tremor: when limb is supported and not voluntarily activated
  - Action tremor: produced during movement of body part
    - Intention
    - Postural
    - Kinetic
    - Isometric
Dysmetria

- Improper measuring of distance in muscular acts
- Hypermetria
- Hypometria

Anatomy Associated with Ataxia

- Most common is damage to cerebellum
  - Limb ataxia or ataxic gait
  - High amplitude tremor that accompanies movement
  - Ipsilateral ataxia due to damage to one cerebellar hemisphere
- Ataxia can result from damage to any of pathways providing cerebellar input or output:
  - Dorsal and ventral spinocerebellar pathways
  - Pontine nuclei
  - Three cerebellar peduncles
- Damage to structures that receive cerebellar input such as thalamus
- Demyelinating diseases in cerebellum or related pathways
Ataxia Classifications

Cerebellar Ataxia

- Damage to cerebellum or connections to cerebellum
- Could include upper limb incoordination, disturbance of posture and walking, dysmetria, dysdiadochokinesia, nystagmus or dysarthria.
- Often worse at faster speeds
Sensory Ataxia

- Ataxia following disruption of proprioceptive input from periphery
  - Damage to afferent component of peripheral nerves
  - Dorsal nerve roots entering spinal cord
  - Dorsal column of spinal cord
  - Medial lemnisci of brain stem
  - Sensory-receiving regions of thalamus or parietal cortex

Ataxia Differential Diagnosis

- Cerebellar damage versus sensory loss
  - Vision is key
  - Sensory ataxia – marked worsening with eyes closed
  - Cerebellar ataxia – minimal worsening with eyes closed
Vestibular Ataxia

- Result of damage to vestibulocochlear nerve or its central connections
- Results from cerebellar damage
- Symptoms: vertigo, nausea, loss of balance, nystagmus

Episodic Ataxia

- Autosomal dominant disorder with sporadic bouts of ataxia with or without myokymia
- 7 types
- Provoked by stress, startle, heavy exertion
- Can first appear at infancy
- Ataxia is most common symptom and caused by misfiring of Purkinje cells in cerebellum
  - Improper regulation of these cells – EA1
  - Malfunction of these cells – EA2
Common Cause – Damage to Cerebellum

Cerebellum Vasculature
Cerebellar Stroke

- Infarct of SCA caused greater impairment than patients with infarctions of PICA
- No significant differences in comparison between right and left sided infarctions or with or without involvement of cerebellar nuclei
- Recovery of motor deficits within first 3 months after stroke was high
- Worse ataxia with lesions in lobules IV-VI

- Bultmann U et al, Gait Posture, 2014

Role of Cerebellum in Motor Control

- Compares movement to intended output
- Predictive/anticipatory modifications in preparation for movement
- Motor learning
- Learns, memorizes and stores motor programs
- Adaptation
Other Roles of Cerebellum

- Balance and equilibrium
- Control of muscle tone
- Accurate direction, extent, force and timing
- Movement composition
- Role in speech production
- Control of eye movement and gaze

S/S Cerebellar Infarct
Cerebellar Dysfunction

- Disorganization of movement, especially rapid movements
- Impaired balance
- Impaired postural control
- Hypotonicity
- Dysmetria
- Decomposition

Ataxia
Dysdiadochokinesia
Tremor
Asthenia
Dysarthria
Ocular dysmetria, nystagmus, etc..

Mechanism of Cerebellar Damage

- Acquired
  - Stroke
  - Tumor
  - Structural (Chiari malformation, agenesis, hypoplasia)
  - Toxicity (alcohol, heavy metals, drugs, solvents)
  - Immune-mediated (MS, gluten ataxia)
  - Trauma
  - Infection (cerebellitis)
  - Endocrine (hypothyroidism)
Mechanisms of Cerebellar Damage

- Degenerative Non-Hereditary
  - Multiple system atrophy
  - Idopathic late-onset cerebellar ataxia

- Hereditary
  - Autosomal dominant disorders (episodic ataxias, spinocerebellar ataxias)
  - Autosomal recessive disorders (Friedreich ataxia, early onset cerebellar ataxia)
  - X-linked disorders (mitochondrial disease, fragile X-associated tremor)

Many Types of Ataxia
Friedreich’s Ataxia

- Autosomal recessive inherited disease; progressive
- Degeneration of nervous tissue in spinal cord
- Initial symptoms: gait disturbance
- Other symptoms: UE and LE muscle weakness, loss of coordination, vision impairment, hearing impairment, dysarthria, pes cavus, scoliosis, heart disease, diabetes
- Symptoms present between 5-15 years of age, but can be late onset

Spinocerebellar Ataxia

- Progressive, degenerative, genetic disease
- Often fatal
- No known effective treatment or cure
- Onset at any age
- Slowly progressive gait ataxia, as well as incoordination of hands, speech, and eye movements
- Many subtypes identified
MS related Ataxia

- 4 out of 5 people with MS will have ataxia to some degree at some point
- Ataxia and tremor are common
- Greater levels of ataxia correlate with higher levels of respiratory symptoms
- Severe tremor correlates with presence of dysarthria
- May experience sensory, vestibular or cerebellar ataxia (or combinations)

Pharmacological Management

- Strong evidence of benefit
  - 4-Aminopyridine – EA2
  - Riluzole – Ataxia with mixed etiology
- Weak evidence of benefit
  - Valproic acid – SCA type 3
  - Thyrotropin-releasing hormone – Spinocerebellar degeneration
  - Zesiewixz TA et al, Neurology, 2018
Ataxia

- Without order or incoordination
- Slurred speech, stumbling, falling, incoordination
- Trouble eating and swallowing
- Eye movement abnormalities
- Tremors
- Cardiac issues
Effect on Postural Control

- Poor upright stance stability
- Broad BOS
- Arms may be in high guard
- Stepping patterns are irregular in direction and distance
- Initiation of forward progression of LEs may start slow, then be flung rapidly and forcefully forward
- Unsteady, staggering with deviation from intended forward line of progression
- Veering, swaying, pitching

Note: The images illustrate the differences in postural control between a healthy subject and a patient with advanced cerebellar ataxia.
Poll Question #3

- What outcome measures do you use in persons with ataxia?
  - A. Ataxia specific (SARA, ICARS)
  - B. Functional Independence Measure (FIM)
  - C. Balance measures (Berg, TUG, etc)
  - D. Gait measures (10 meter, 6 minute)
  - E. More than one above

Outcome Measures

- Traditional function/activity measures
  - FIM
  - Balance/Postural control measures
  - Gait measures

- Ataxia specific
  - International Cooperative Ataxia Rating Scale (ICARS)
    - [https://www.sralab.org/rehabilitation-measures/international-cooperative-ataxia-rating-scale](https://www.sralab.org/rehabilitation-measures/international-cooperative-ataxia-rating-scale)
  - Scale for the Assessment and Rating of Ataxia (SARA)
ICARS

- Posture and Gait Disturbance
- Kinetic Functions
- Speech Disorders
- Oculomotor Disorders

SARA

- Gait
- Stance
- Sitting
- Speech disturbance
- Finger chase
- Nose-finger test
- Fast alternating hand movements
- Heel-shin slide
Interventions

Treatment

- Not lots of evidence
- Treat the symptoms
- Postural stability
- Gait
- Balance
- Accuracy of limb movements
Interventions

- Pressure splints – no additional benefit

- PT and OT – improvements in ataxia and functional abilities

- Stochastic vibration therapy – insufficient evidence
  - Zesiewicz TA, *Neurology*, 2018

Treatment of postural disorders in cerebellar ataxia

- Moderate level evidence that rehab is efficient to improve postural capacities
- Particularly effective in those with degenerative ataxia or MS

- **Intensive rehab** programs with balance and coordination exercises are key
Prevention of Falls

- 84% of patients fell at least once during a 1 year observation
  - Fonteyn et al, Eur Neurol, 2013
- Fear of falling needs to be assessed as well
- Living environment may need to be modified
- Address those factors that increase fall risks like visual impairments, bowel and bladder disorders, footwear, etc…
- Use of fall detection systems may be warranted

Partial Body Weight-Supported Treadmill Training

- In sample of persons with spinocerebellar ataxia
- PBWSTT protocol
  - Began with 30% BWS
  - First stage: gait and conditioning
    - Progressive reduction of BWS to zero
    - Progressive increase in velocity to maximal
  - Second stage: dynamic balance training
    - Main goal was challenging balance during gait
    - No UE support
    - Throwing/catching ball while walking
PBWSTT results

- Feasible and tolerated well
- Significant increase in gait performance, treadmill inclination, duration of exercise and cardiopulmonary capacity
- After second stage, balance improvements also demonstrated
  - Santos d Oliveira et al, *Rehabil Res Practice*, 2018

Treatment

- Intensive long term motor training
- Supervised as well as home exercise programs
- Intensive coordination training
- HEP focused on static and dynamic balance activities
  - Sitting and standing
  - Effect on walking
Treatment

- Use of biofeedback and/or bandwith feedback
- Decreasing degrees of freedom
- Activities that focus on stability, co-contraction, midrange control
- Use of resisted movements

Examples of balance exercises.

<table>
<thead>
<tr>
<th>Exercise category</th>
<th>Description</th>
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| Seated, stable surface | Track unassisted.  
- Side-to-side weight transfer.  
- Stand bending.  
- Arm exercise.  
- Foot circles.  
- Take outward.  |
| Seated, dynamic surface | Track on Physio-roll.  
- Oddly leaves.  
- Stand free, reach forward.  
- Reaching (hands on or free).  
- Side-to-side roll, back and forth roll.  
- Foot circles (hands on or free).  |
| Standing, stable surface progressing to loose surface. | Foot apart progressing to feet together.  
- Weight shift, arm and side-to-side.  
- Arm moves.  
- Stand rotation.  
- Up on toes.  
- Up on heels.  
- Eyes closed.  |
| Stepping and stepping | Single limber stance.  
- Lunge.  
- Toward stepping.  
- Step one to three step.  
- Floor version.  
- Arm and non-affected leg lift.  |
Treatment for Friedreich’s Ataxia

- Low-intensity strengthening exercise; need sufficient rest and avoid fatigue
- Flexibility/stretching exercises for LE
  - Stretching of spine, foot (pes cavus), hamstrings especially
- Coordination exercises
  - Emphasize watching movement for visual feedback
- Use of appropriate assistive device
- Balance exercises
- Conditioning
  - Moderate exercises

Gibson et al.

OBJECTIVE: The use of external body weights, although controversial, is occasionally employed to improve balance or mobility in patients with ataxia or tremor. This case report describes the effect of torso-weighting to counteract directional balance loss in a woman with relapsing-remitting multiple sclerosis.

CASE DESCRIPTION: Clinical examination of a 40-year-old woman after multiple sclerosis exacerbation revealed loss of balance in the posterior direction during quiet standing as well as loss of dynamic balance in the posterior and lateral directions. The patient's standing posture was with her trunk posterior to her pelvis. She exhibited decreased strength in both extremities and trunk, diminished sensation in the right lower extremity and pals, and an unstable ataxic gait. Difficulty with walking and severe fatigue and dizziness were also reported. Stabilizing balance and alignment were examined during (1) quiet standing with eyes open and eyes closed, (2) translational movements, and (3) multidirectional trunk perturbations. The patient demonstrated a loss of balance and alignment in the posterior direction in all tests.

INTERVENTION: Based on balance examination results, the patient was fitted with a 0.5-lb vest containing 1.5 lb of additional weight placed anteriorly on the torso at the level of the umbilicus. Progressive balance, gait, and functional activities were repeated both with and without weighting the torso over six weeks.

OUTCOME: Immediately on weighting, the patient demonstrated less sway in quiet standing, increased stability when perturbed, improved body alignment, and less ataxia during gait. The patient was able to accomplish more challenging activities with better balance while weighted. Functional improvement in walking and improved control during balance activities were demonstrated in later treatment sessions without weighting.

CONCLUSION: Placing small amounts of weight asymmetrically on the torso, based on directional loss of balance and alignment, seemed to assist this patient in maintaining balance during static and dynamic activities. Additional research may help determine whether this intervention is applicable to others with directional losses of balance, ataxia, or multiple sclerosis to improve balance control.

Randomized Clinical Trial of Balance-Based Torso Weighting for Improving Upright Mobility in People with Multiple Sclerosis

Gail L. Widener, PhD, PT, Diane D. Allen, PhD, PT, and Cynthia Gibson-Horn, PT

Background: Torso weighting has sometimes been effective for improving upright mobility in people with multiple sclerosis, but parameters for weighting have been inconsistent. Objective: To determine whether balance-based torso weighting (BBTW) has immediate effects on upright mobility in people with multiple sclerosis. Methods: This was a 2-phase randomized clinical trial. In phase 1, 35 participants were randomly assigned to experimental and control groups. In phase 2, the control group was subsequently randomized into 2 groups with alternate weight-placements. Tests of upright mobility included: timed up and go (TUG), sharpness Romberg, 360-degree turn, 25-foot walk, and computerized platform posturography. Participants were tested at baseline and again with weights placed according to group membership. In both phases, a physical therapist assessed balance for the BBTW group and then placed weights to decrease balance loss. In phase 1, the control group had no weights placed. In phase 2, the alternate treatment group received standard weight placement of 1.5% body weight. Results: People with BBTW showed a significant improvement in the 25-foot walk (P < 0.01) over those with no weight, and the TUG (P = 0.01) over those with standard weight placement. BBTW participants received an average of 0.5 kg, less than 1.5% of any participant's body weight. Conclusions: BBTW can have immediate advantages over a nonweighted condition for gait velocity and over a standardized weighted condition for a functional activity in people with multiple sclerosis (MS) who are ambulatory but have balance and mobility abnormalities.
Compensatory Strategies

- Slow down movements
- Reduce number of segments moving at any given time
- Widen BOS
- Minimal environmental distractions
- Weighting (axial v limb)
- Assistive devices
- Orthotics

Other Ideas?
Questions?

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