Introduction

- Ataxia is derived from Greek word ‘a’-not, ‘taxis’-orderly, (not orderly/ not in order)
- Ataxia is the inability to make smooth, accurate and coordinated movements
- Ataxia is a symptom, not a specific disease or diagnosis.
- The term ataxia is most often used to describe walking that is uncoordinated and unsteady.
- Ataxia can affect coordination of fingers, hands, arms, speech (dysarthria) and eye movements (nystagmus).

Ataxia can arise from disorders of:

- Cerebellum (most common)
- Sensory pathways (Sensory Ataxia)
- Posterior columns, dorsal root ganglia, peripheral nerves
- Frontal lobe lesions via fronto-cerebellar associative fibers
- Extra pyramidal system
- Vestibular system

The clinical approach to patients with ataxia involves differentiating ataxia from other sources of imbalance and incoordination.

Distinguishing cerebellar from sensory ataxia, and designing an evaluation based on knowledge regarding various causes of ataxia and cerebellar disorders.

Sensory Ataxia

- The ataxia of severe sensory neuropathy and of posterior column disease of the spinal cord (sensory ataxia) simulates cerebellar ataxia; presumably this is a result of involvement of the large peripheral spinocerebellar afferent fibers.
  1. loss of distal joint, position sense,
  2. absence of associated cerebellar signs such as dysarthria or nystagmus,
  3. loss of tendon reflexes, and
  4. the corrective effects of vision on sensory ataxia.
  5. Romberg sign:
- This sign is not found in lesions of the cerebellar hemispheres except that the patient may initially sway with eyes open and a bit more with eyes closed.
Caveats

- Friederick’s ataxia, Multiple sclerosis…
- Overlap of clinical features to be expected in clinical practice

Cortical Ataxias

- **FRONTAL LOBE ATAXIA** refers to disturbed coordination due to dysfunction of the contralateral frontal lobe;
- Results from disease involving the frontopontocerebellar fibers.
- Hyperreflexia, hypertonia and Release reflexes while cerebellar lesion can cause diminished or pendular reflexes and typical hypotonia
- A lesion of the “SUPERIOR PARIETAL LOBULE” (areas 5 and 7 of Brodmann) may rarely result in ataxia of the contralateral limbs

Frontal lobe ataxia/ Bruns’ ataxia

- Frontal lobe ataxia refers to disturbed coordination due to dysfunction of the contralateral frontal lobe; it may resemble the deficits due to abnormalities of the ipsilateral cerebellar hemisphere.
- Pressure on the brainstem by a cerebellar mass lesion may cause corticospinal tract findings that can confuse the picture.
- Bruns’ ataxia refers to a gait disturbance seen primarily in frontal lobe lesions.

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<thead>
<tr>
<th>Table 24-2 Features of Cerebellar Ataxia, Sensory Ataxia, and Frontal Gait Disorders</th>
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<td><strong>Base of support</strong></td>
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<td><strong>Velocity</strong></td>
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<td><strong>Stnde</strong></td>
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<td><strong>Romberg</strong></td>
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<td><strong>Heel → shin</strong></td>
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<td><strong>Initiation</strong></td>
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<tr>
<td><strong>Turns</strong></td>
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<td><strong>Postural instability</strong></td>
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<td><strong>Falls</strong></td>
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</table>

Nystagmus, ocular dysmetria, and other eye movement abnormalities
Steppage gait
Reeling, ataxic gait
Decreased reflexes
Other signs of cerebellar disease (dyssynergia, dysmetria, dysdiadochokinesia, hypotonia, rebound, impaired check response)
Thalamic Ataxias

- Transient ataxia affecting contralateral limbs after lesion of anterior thalamus
- may see associated motor (pyramidal tract) signs from involvement of internal capsule
- also can result in asterixis in contralateral limbs (hemiasterixis)

Vertiginous ataxia

- Vertiginous ataxia is primarily an ataxia of gait and is distinguished by the obvious complaint of vertigo and listing to one side, past pointing, and rotary nystagmus.

Nonvertiginous ataxia

- The nonvertiginous ataxia of gait caused by vestibular paresis (e.g., streptomycin toxicity) has special qualities

Vertiginous and cerbellar ataxia

- Vertigo and cerebellar ataxia may be concurrent, as in some patients with a paraneoplastic disease and in those with infarction of the lateral medulla and inferior cerebellum.

VESTIBULAR – ataxia associated with vestibular nerve or labyrinthine disease results in a disorder of gait associated with a significant degree of dizziness, light-headedness, or the perception of movement

PSEUDO ATAXIA- Mild Pyramidal weakness & Extrapyramidal disorders, weakness of proximal leg muscles mimics cerebellar disease

PSYCHOGENIC – Extremely anxious patients

Classification

Based on onset and progression

- acute
- sub acute
- chronic

Based on site of presentation

- unilateral
- bilateral
- limb ataxia
- truncal ataxia

Ataxia

- “errors in the RATE, RANGE, FORCE & DIRECTION of movement”
- Gait ataxia
- Truncal ataxia
- Limb ataxia
Classic features and tests

- **Dyssynergia:** results in jerky decomposed movements (heel-knee-shin test)
- **Dysmetria:** due to delayed activation of antagonists
  - often correction to target by series of jerky corrections (finger nose test)
  - may lead to intention tremor in limbs with finger-to-nose or foot-to-target testing as rhythmic oscillation emerges close to target
- **Dysdiadochokinesis:** irregularities of force, speed, and rhythm

Other features

- **Hypotonia:** decrease in resistance to passive movement of muscles related to depression of gamma motor neuron activity (usually seen transiently in acute phase of cerebellar lesions), pendular knee jerk
- **Rebound phenomenon:** related to poor tone and weak check response, so when tap or displace limb, wider range of movement in return to static position, incl. Holmes phenomenon when suddenly release flexed arm held against resistance - unable to stop flexion and arm strike self (delay in activation of antagonist triceps muscle)
- **Dysarthria:** often scanning type with irregularities in tone, with words broken into syllables; often slow with occasional rapid portions ("explosive speech")
- **Ocular Motor Abnormalities:**
  - usually if vestibular connections or flocculonodular lobe affected
  - pursuit movements no longer smooth, but saccadic
  - may over- or under-shoot target with attempts at fixation (ocular dysmetria)
  - in primary position may see saccadic intrusions (such as macro square-wave jerks) or primary nystagmus (incl. vertical, esp. up-beat nystagmus) or periodic alternating nystagmus
  - rebound nystagmus can occur with contralateral-beating nystagmus on return of eyes to primary position after eccentric gaze evoked nystagmus to one side
- **Writing abnormalities**
- **Positional projectile vomiting** (posterior fossa lesions)

Approach to Cerebellar Ataxia in Adults

The “four” questions

- **History:**
  - Mode of onset?
  - Progression?
- **Examination:**
  - Focal/Symmetric involvement?
  - Localisation of the cerebellar lesion

MODE OF ONSET

- **ACUTE-** hours to days
- **SUB ACUTE-** days to weeks
- **CHRONIC-** months to years
Approach to Ataxia

Ataxia

- **Unilateral/Focal** (acute, sub-acute, chronic)
- **Symmetrical** (acute, sub-acute, chronic)

<table>
<thead>
<tr>
<th>Unilateral/focal ataxia</th>
<th>Symmetrical ataxia</th>
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<tr>
<td><strong>Acute</strong></td>
<td><strong>Acute</strong></td>
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<tr>
<td>vascular</td>
<td>intoxication</td>
</tr>
<tr>
<td>infection</td>
<td>acute viral cerebellitis</td>
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<tr>
<td>demyelination</td>
<td>post-infectious syndrome</td>
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<tr>
<td><strong>Sub-acute</strong></td>
<td><strong>Sub-acute</strong></td>
</tr>
<tr>
<td>neoplastic</td>
<td>drugs &amp; toxin</td>
</tr>
<tr>
<td>demyelination</td>
<td>alcohol &amp; nutritional</td>
</tr>
<tr>
<td>infection-AIDS related</td>
<td>lyme disease</td>
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<tr>
<td><strong>Chronic</strong></td>
<td><strong>Chronic</strong></td>
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<tr>
<td>stable gliosis</td>
<td>inherited: phenytoin</td>
</tr>
<tr>
<td>congenital</td>
<td>paraneoplastic: anti-gliadin Ab</td>
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<td>hypothyroidism: tabes dorsalis</td>
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Acquired vs. Genetic causes of ataxia

ACQUIRED CAUSES OF ATAXIA

**Vascular:** Ischemic stroke, hemorrhagic stroke, AV malformations  
**Infections:** Acute cerebellitis, postinfectious encephalomyelitis, cerebellar abscess, HIV  
**Toxic:** Alcohol, anticonvulsants, mercury, 5-FU, cytosine arabinoside, lithium  
**Neoplastic/compressive:** Gliomas, ependymomas, meningiomas, basal meningeal carcinomatosis, craniovertebral junction abnormalities  
**Immune:** Multiple sclerosis, paraneoplastic syndromes, anti-GAD, gluten ataxia  
**Deficiency:** Hypothyroidism, vitamin B₁ and B₁₂, vitamin E

GENETIC CAUSES OF ATAXIA

**Autosomal recessive:** FA, AT, AVED, AOA 1, AOA 2, other inborn errors of metabolism  
**Autosomal dominant:** SCA types 1 through 2
Progression

1. Progressive
2. Static
3. Intermittent symptoms
4. Reversible Ataxias

1) Progressive ataxia

Classifications of Greenfield and of Harding:

1. spinocerebellar ataxias, with unmistakable involvement of the spinal cord (Romberg sign, sensory loss, diminished tendon reflexes, Babinski signs).
2. pure cerebellar ataxias, with no other associated neurologic disorders.
3. complicated cerebellar ataxias, with a variety of pyramidal, extrapyramidal, retinal, optic nerve, Oculomotor, auditory, peripheral nerve, and cerebrocortical accompaniments.

2) Static ataxias

- Vascular causes

3) Intermittent symptoms

- Episodic ataxias (inherited etiology)

4) Reversible ataxias

- Infectious causes – postviral
- Thyroid
- Drugs
- Toxins