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MULTI-SYSTEM

CANVAS affects the brain, nervous system, and inner ear, resulting in loss of coordination.

ARTICLE

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CANVAS Syndrome

By the Vestibular Disorders Association, reviewed by Dr. Diego Kaski & Prof. Adolpho Bronstein

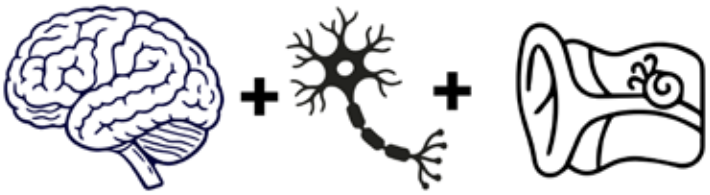
CANVAS—short for Cerebellar Ataxia, Neuropathy, and Vestibular Areflexia Syndrome—is a progressive neurological syndrome that disrupts three systems that normally work together to keep us upright, steady, and oriented: the cerebellum (one of whose roles is the coordination of eye, arm, trunk, and leg movements), sensory nerves (especially those carrying position and vibration sense), and the vestibular system (the inner-ear balance sensors that detect head movements). The result is a distinctive "triple-hit" to balance and movement that can feel like a confusing mix of unsteadiness, dizziness, oscillopsia (bouncing/blurring vision with head movement), numbness/tingling, and worsening gait instability over time.

Thanks to much research over the last decade, our understanding of this condition has transformed from a loosely sketched clinical pattern into a disease that can be recognized, tested, and genetically confirmed—which matters enormously for ending diagnostic uncertainty and guiding supportive care.

CANVAS usually begins later in life, with the average age of symptom onset in the early 50s. Many people have no family history of the condition, even though it is genetic. This is because the gene change is recessive, meaning a person needs two altered copies of the gene to develop CANVAS. It is a slowly progressive condition. Many people remain independent for years. About half of individuals use a walking stick after around 10 years, and a quarter require a wheelchair after 15 years. The rate of progression varies widely, and many people continue to live active, fulfilling lives with appropriate support and rehabilitation

WHAT "CANVAS" ACTUALLY MEANS

The name is a checklist of the three core features:



CEREBELLAR ATAXIA

The cerebellum that fine-tunes movement. When it's affected, people can develop:

- A wide-based, unsteady gait
- Poor coordination (limb "overshoot," clumsiness)
- Slurred speech (dysarthria) or swallowing issues (dysphagia) later on in some cases
- Oscillopsia (bouncy vision) due to downbeat nystagmus (vertical oscillation of the eyes)
- Neuropathy / neuronopathy
- In CANVAS, sensory nerves—particularly those relaying position sense—can be impaired, leading to:
- Numbness, tingling, pain, cramps
- Trouble knowing where feet/legs are in space (sensory ataxia), especially in the dark or on uneven ground
- Vestibular areflexia (bilateral loss of vestibular function)
- "Areflexia" here means the vestibular reflexes are reduced or absent. People may experience:
- Chronic imbalance and falls
- Oscillopsia with head movements: vision that blurs or bounces when walking or turning the head
- Difficulty with quick head movements, busy visual environments, or walking while looking around

Importantly, hearing is preserved, which helps distinguish CANVAS from other inner ear disorders.

Many people don't develop all three features at once. Symptoms can unfold gradually over years, which is one reason CANVAS has historically been under-recognized.

A big turning point: the RFC1 genetic discovery

For years, CANVAS was diagnosed clinically based on the symptom pattern and specialized vestibular/neurologic testing. A breakthrough came in 2019, when Cortese and colleagues identified a common genetic cause: a biallelic (two-copy) repeat expansion in the RFC1 gene—specifically an intronic AAGGG repeat expansion.

WHY THAT MATTERS:

- It showed CANVAS is often an inherited (recessive) genetic disorder, even when there's no obvious family history.
- It revealed RFC1 expansions are a frequent cause of late-onset ataxia, especially when sensory neuronopathy and bilateral vestibular loss co-exist.
- It helped define a broader clinical syndrome now often called the "RFC1 spectrum"—meaning some people have overlapping features (sensory neuropathy, chronic cough, ataxia, vestibular loss) that do not fit the classic triad at the start.

Importantly, finding biallelic RFC1 expansions in an individual does not automatically confirm a CANVAS diagnosis. This is because RFC1 expansions can present with a spectrum of findings and therefore clinicians still need to

match the genetic results to the patient's symptoms and examination findings.

The clinical "fingerprints" of CANVAS
Although genetics has clarified the cause of CANVAS in many patients, clinical identification remains essential, both to determine who should undergo testing and to guide management of real world symptoms. A widely cited and enduring clinical clue is an abnormal visually enhanced vestibulo ocular reflex (VOR), an eye movement signature that reflects the combined cerebellar and vestibular involvement and has been emphasized since the earliest clinical descriptions of CANVAS. Importantly, the phenotype extends beyond the classic triad: chronic cough, orthostatic symptoms, and neuropathic pain are common accompaniments, reinforcing the need to recognize CANVAS as a multisystem disorder rather than a narrowly defined vestibular syndrome.

CANVAS AS A MULTISYSTEM DISORDER RATHER THAN A NARROWLY DEFINED VESTIBULAR SYNDROME.

CANVAS is not only a disorder of balance. Large clinical series and reviews have consistently reported additional features, including chronic cough, orthostatic hypotension (with lightheadedness on standing), nerve pain, and swallowing difficulties, alongside a high risk of falls that becomes a major management priority. These additional features matter because they can appear early and help steer the diagnostic work up in the right direction, particularly in patients presenting with a puzzling combination of dizziness, gait unsteadiness, and sensory symptoms.

HOW CANVAS IS DIAGNOSED (AND WHY IT CAN BE MISSED)

Because CANVAS involves multiple systems, diagnosis often requires multidisciplinary testing. Common diagnostic components include:

1) Vestibular testing
To confirm bilateral vestibular hypofunction/areflexia, clinicians may use:

- vHIT (video head impulse test)
- Caloric testing
- Rotational chair testing
- Dynamic visual acuity testing (to quantify oscillopsia)

2) Nerve evaluation to evaluate sensory neuropathy/neuronopathy:

- Neurological examination (vibration sense, proprioception, reflex patterns)
- Nerve conduction studies/EMG (often showing sensory involvement)

3) Brain imaging
MRI can show cerebellar volume loss in many patients (though imaging may be subtle early on).

4) Genetic testing for RFC1 expansions

Standard genetic panels may miss repeat expansions unless the lab specifically tests for RFC1 repeat expansions. If CANVAS is suspected clinically, it's worth ensuring the correct test is ordered.
What causes symptoms: a quick, patient-friendly explanation
Your brain maintains balance by "triangulating" input from:

- Inner ears (vestibular)
- Eyes (visual)
- Feet/legs and joints (somatosensory/proprioception)

- Cerebellum (coordination and error correction)

CANVAS disrupts vestibular input and position sense while also affecting the cerebellum's ability to compensate. That combination makes compensation harder than in a single-system problem. It's why someone may feel dramatically worse:

- In darkness (less visual compensation)
- On uneven surfaces (requires proprioception)
- With head turns while walking (requires vestibular reflexes)

TREATMENT AND MANAGEMENT: WHAT ACTUALLY HELPS?

There is not yet a cure that reverses CANVAS, but there is a lot that can improve function, safety, and quality of life. Most care is supportive and symptom-targeted:

- Vestibular and balance rehabilitation
- Gait/balance therapy with fall-prevention focus
- Training to improve stability with head motion
- Use of assistive devices when needed (trekking poles, cane, walker)
- Home safety modifications (lighting, grab bars, tripping-hazard reduction)
- Foot care, proprioceptive strategies, appropriate footwear/orthotics
- Neuropathic pain treatments when present (medications, topical options, pain specialists)
- Managing "non-balance" features
- Orthostatic symptoms: hydration/salt strategies when appropriate, compression garments, medication guidance from clinicians (especially if autonomic dysfunction is confirmed)
- Swallowing issues: early referral to speech-language pathology (swallow evaluation), diet texture strategies if needed
- Chronic cough: evaluation to rule out common causes and consider CANVAS/RFC1 context if cough is longstanding
- When to suspect CANVAS (a practical checklist)

CANVAS BECOMES MORE LIKELY WHEN SOMEONE HAS:

- Gradually progressive imbalance and falls (especially later adult onset)
- Oscillopsia or marked unsteadiness with head movement
- Sensory symptoms (numbness/tingling) or loss of vibration/position sense
- Signs of cerebellar involvement (coordination issues, abnormal eye movements, speech changes)
- Chronic cough or orthostatic symptoms in the mix
- If this pattern fits, it's reasonable to ask a neurologist (often a movement disorders specialist or neurogeneticist) or a neuro-otologist about RFC1 testing and formal vestibular function testing.

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