



CANVAS SYNDROME

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.



Key points

- A rare neurological syndrome affecting balance.
- Most common in adults over 50.
- Slowly gets worse over 10 or more years.
- 3 parts:
 - **cerebellar ataxia** (problem with muscle control managed by the cerebellum)
 - **neuropathy** (problem with nerves)
 - **vestibular areflexia** (vestibular system reflexes do not work properly)
- Balance problems may be worse when all 3 parts are active rather than only 1 or 2.
- Vertigo (spinning sensation) usually not a symptom.
- Treatment mainly involves managing symptoms and avoiding things that could make symptoms worse.
- Vestibular rehabilitation (an exercise-based therapy) can help the brain compensate for missing inner-ear signals.

What is CANVAS syndrome?

CANVAS is an acronym that stands for Cerebellar Ataxia (CA), Neuropathy (N) and Vestibular Areflexia (VA) Syndrome.

CANVAS syndrome is a neurological disorder, which is a problem with the brain, spinal cord and/or nerves. It usually starts in adults over 50 and slowly gets worse (progresses) over 10 years or more.

A syndrome is a collection of problems or symptoms that are often found together. This means that CANVAS syndrome has three components:

- The cerebellum (sair-a-BELL-um) is the part of the brain that controls muscle coordination. Ataxia (a-TAX-ee-a) is a problem with muscle control or coordination. So cerebellar ataxia is a problem with muscle control or coordination that is caused by a problem with the cerebellum.
- Neuropathy (new-ROP-a-thee) is a problem with the nerves. When the nerves that carry signals about touch, pressure, pain, and temperature are affected, it is known as sensory neuropathy. When the nerves that help control muscles and movement are affected, it is known as motor neuropathy. When the nerves that help control the functions of your internal organs are affected, it is known as autonomic neuropathy.
- The vestibular system in the inner ear is an important part of the balance system. Areflexia (ay-re-FLEX-ee-a) literally means a lack of reflexes. So vestibular areflexia means that the vestibular system's reflexes are not working normally. In particular, the vestibulo-ocular reflex (VOR) is affected. This reflex keeps your eyes steady even if your head is moving. For example, it enables you to focus on the words on this page even if you nod your head up and

down or turn it from side to side. And when you walk, the VOR keeps your vision stable while your head bobs up and down. Patients with CANVAS syndrome have bilateral vestibular loss, meaning that both inner ears are affected.

CANVAS syndrome was first described in 2004, and researchers are still learning about it. It is still unclear if CANVAS syndrome is a distinct disease or a collection of symptoms that sometimes occur together. CANVAS syndrome seems to be quite rare: possibly only 5 people in a million have it.

What are the causes?

Researchers are still studying the causes of CANVAS syndrome. People with CANVAS syndrome have atrophy (shrinkage) of the cerebellum, the part of the brain that controls muscle coordination. They also have damage to certain nerves.

Researchers have found some cases where CANVAS syndrome seems to run in families, and recently, a genetic change (mutation) has been found in a few families with CANVAS syndrome. But it is still not clear how this mutation causes CANVAS syndrome. It is also possible that more than one gene can cause CANVAS syndrome in these families.

What are the symptoms?

People with CANVAS syndrome have symptoms of all three of its components: cerebellar ataxia, neuropathy and vestibular areflexia. Sometimes, people may have symptoms of one or two components for many years before developing symptoms of all three.

Symptoms of cerebellar ataxia may include:

- problems with walking (gait); for many people, this is the first symptom of CANVAS syndrome
- imbalance
- difficulty with coordination, such as trouble picking up objects on the first try

- trouble doing things that need fine motor control, such as writing or eating
- slow, slurred speech (dysarthria)
- rapid, involuntary eye movements (nystagmus - nuh-STAG-muhs)
- trouble swallowing, or feeling as though things get stuck in your throat (dysphagia)

Symptoms of neuropathy may include:

- chronic cough
- muscle weakness, numbness or pain in the hands and feet (peripheral neuropathy)
- pain from things that would not normally cause pain (allodynia)
- abnormal feelings such as burning or electric shock from things that would not normally cause those feelings (dysesthesia - duh-suhs-THÉE-zhuh)
- trouble swallowing (dysphagia – duh-SFEI-juh)
- problems with balance
- *eeling dizzy or lightheaded when you sit up or stand up (orthostatic hypotension)
- feet that are cooler and sweat less than the rest of the body
- dry mouth or eyes
- constipation
- erectile dysfunction

Symptoms of vestibular areflexia may include:

- loss of balance (also called postural imbalance or loss of postural control)
- unsteady gait (walking or running)
- feeling unstable or dizzy
- in some people, blurry or jumpy vision when the head moves, including when they are walking or running (oscillopsia - oss-ill-OP-see-uh)

The balance and gait problems get worse when it is dark, or when you are on uneven or springy ground

or on a moving surface. These problems usually go away when you are sitting still or lying down.

People with CANVAS syndrome usually do not have feelings of spinning (vertigo).

Vestibular problems in CANVAS syndrome always affect both sides of the body.

Balance problems may be worse when someone has all three components of CANVAS syndrome (cerebellar ataxia, neuropathy and vestibular areflexia) than if they only have one or two of its components.

How is it diagnosed?

CANVAS syndrome may be diagnosed by a primary care doctor, but it is more often diagnosed by a specialist such as an otolaryngologist, an otologist, or a neurologist.

CANVAS syndrome is diagnosed when you have all three of its components: cerebellar ataxia, neuropathy and vestibular areflexia. Diagnosis is challenging when you have only one or two components. It may take more than 10 years before the final component shows up and you are finally diagnosed.

Your doctor will ask about your symptoms. Try to be as specific as possible about your symptoms and when they get better or worse.

Your doctor will also ask about your medical history, including any medications you are taking or recently stopped taking and any conditions you have been diagnosed with in the past. Your doctor will also do a thorough physical and neurological exam. Tests may include asking you to watch the doctor's nose while the doctor moves your head, or to watch your own thumbs while the doctor turns you in an office chair.

You will probably have some of the following diagnostic tests:

- balance tests
- vestibular function tests
- nerve conduction studies

- magnetic resonance imaging (MRI)

If you have trouble swallowing (dysphagia), your doctor may suggest an assessment by a speech-language pathologist. Speech-language pathologists specialize in assessing and treating problems with speech, communication, swallowing and feeding.

Your doctor may also suggest genetic testing to rule out other genetic mutations that cause ataxia, such as Friedrich ataxia and spinocerebellar ataxia (SCA) type 3.

How is it treated and managed?

Currently no treatments are available to reverse or slow the progression of CANVAS syndrome.

Treatment mainly involves managing the symptoms of CANVAS syndrome and avoiding things that could make the symptoms worse.

Managing drops in blood pressure

Some people with CANVAS syndrome have sudden drops in blood pressure when they sit up or stand up (orthostatic hypotension). This can cause dizziness, lightheadedness, blurred vision, and other problems. To prevent this, your doctor may recommend some or all of the following:

- avoiding certain medications that can make the problem worse
- drinking fluids and staying well hydrated, especially in warmer weather
- wearing compression stockings
- taking a steroid or medication that increases blood pressure

Managing swallowing problems

If you have problems with swallowing, a speech-language pathologist may be able to help. Often, a specific head position while eating can help with swallowing problems. In some cases, you may need to eat foods that have a particular texture or consistency, which are easier to swallow.

Managing balance problems

When the vestibular system is not working properly, it means that the brain's balance system is getting little or no information from the inner ear balance mechanism. You will feel unsteady and off balance.

If you have neuropathy, your imbalance will be even greater. The brain's balance system will get little or no information from both the vestibular and proprioceptive (skin, muscles, and joints) systems. You will need to rely on information from your eyes (visual system) to stay upright.

It is a challenge for your brain to make up for the missing information from the vestibular and proprioceptive systems. Vestibular rehabilitation may help.

Vestibular rehabilitation is a type of exercise therapy. Its goal for people with CANVAS Syndrome is to help the brain learn to compensate for signals that are missing. A vestibular therapist can help you set treatment goals and design an appropriate program.

Vestibular rehabilitation for CANVAS Syndrome may include:

- **habituation exercises** to help you get used to symptoms that happen when you move your head
- **gaze stability exercises** to help you focus on an object while your head is moving; for instance, you may begin by moving your head slowly and watching an object that is not moving, and work up to watching a moving object while your head is moving fast
- **balance exercises**, including strength training
- **substitution exercises**, which means learning to

use your visual and proprioceptive system more effectively for balance; if you have neuropathy, substitution exercises will focus on learning to rely more on your visual system

An assessment with an occupational therapist can help spot things in your home that might be a risk for tripping and falling. They can also suggest assistive devices and modifications like grab bars to help you stay steady.

Your doctor may recommend neurological physiotherapy. It is a type of exercise therapy for people with problems that affect the brain, spinal cord, and nerves. It can help improve walking, strength, endurance, and balance. A physiotherapist will work with you to design a program of exercises.

What to expect in the future

In future, as researchers learn more about CANVAS syndrome, it may be possible to diagnose it more easily. Treatments to prevent, reverse or slow CANVAS syndrome may also become available.

It is also possible that as we learn more about it, the definition of CANVAS syndrome may change. For example, the diagnosis of CANVAS syndrome could one day be limited to people with a specific genetic mutation and certain symptoms.

Visit our website

View this and other articles about vestibular disorders – www.balance&dizziness.org.

In addition, find information about how the balance system works, the journey from diagnosis to treatment, building a wellness toolkit, and more.

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