



Muscular Dystrophy  
New Zealand

## **CANVAS - CEREBELLAR ATAXIA, NEUROPATHY AND VESTIBULAR AREFLEXIA SYNDROME**

CANVAS is an adult-onset slowly progressive neurologic disorder that has an average age of onset of 50-60 years. Patients with CANVAS combine cerebellar ataxia (i.e. coordination problems, CA) peripheral nerve damage (neuropathy - N), and loss of vestibular function (vestibular areflexia -- the VA). The vestibular system is a collection of structures in your inner ear that provides you with your sense of balance and an awareness of your spatial orientation (meaning a sense of whether you are right-side up or upside-down). Your brain then integrates that information with other sensory information from your body to coordinate smooth and well-timed body movements. The combination of these three conditions above causes major disturbances to balance and gait which is progressive i.e.: can worsen with time and age. CANVAS is a very rare disorder, most of the cases reported with the condition do not have family history and thought to be inherited in an autosomal recessive pattern. This means two mutated genes are inherited, one from each parent. These disorders are usually passed on by two carriers. Their health is rarely affected, but they have one mutated gene (recessive gene) and one normal gene (dominant gene) for the condition.

Common features of this condition include "dolls head reflex" upon examination or impaired ability of the eye velocity to match head velocity when turning the head from side to side. Involuntary eye movements can occur in some individuals with the condition as well as difficulty articulating speech. Individuals with the condition may have problems standing and will show a positive Romberg test which means they experience loss of balance when the eyes are closed. Loss of coordination of the limbs can also occur. Other presenting features can include a persistent cough, dysesthesia (which is an abnormal unpleasant sensation felt when touched, caused by damage to peripheral nerves), oscillopsia (a visual disturbance in which objects in the visual field appear to oscillate), dizziness, and falls. Brain MRI of individuals with the condition will show cerebellar atrophy.



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### **Diagnosis:**

Physical examination, vestibular function testing, videooculography, nerve conduction studies and MRI of the brain are normally conducted to detect signs and symptoms of this condition.

### **Treatment:**

There is no known cure for CANVAS and treatments are generally to manage symptoms such as pain and dizziness. CANVAS is progressive and a person with this the condition may eventually require the use of a mobility assistance device and may need assistance to perform daily tasks. Modification of the home with things such as grab bars, raised toilet seats, and ramps may be beneficial.

Speech therapy and communication devices such as writing pads and computer-based devices may benefit those affected with affected speech. Weighted eating utensils and dressing hooks can help maintain independence. Weight control is important because obesity can exacerbate difficulties with ambulation and mobility. Individuals experiencing swallowing difficulties (dysphagia) may suffer significant weight loss and will benefit from seeing a speech language therapist and dietician.

People with CANVAS should be followed up by a neurologist annually with visits to physiotherapists, occupational therapists and other specialists as needed.

### **Employment:**

Seeking and maintaining paid employment can be challenging for people with CANVAS, especially as their condition progresses. Despite these challenges many people in New Zealand with neuromuscular conditions carve out a career and work productively and successfully for a number of years. Research has shown that a paid occupation is achievable for others with the correct supports and environmental conditions (flexibility, adaptations, employer recognition, peer support).

When choosing a career, if possible choose something that you are passionate about and that meets your physical needs now and into the future as your condition progresses. Consider the workload; repetitive tasks, physicality of the job, or how much speaking is required if you struggle with slurred speech. Ask about opportunities for job shadowing to get a sense of daily tasks and expectations. Consider when are you more



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alert and more fatigued? Is there flexibility to work from home on certain days or to be flexible with work schedules so you can incorporate rests if needed? Will the job accommodate flexibility to meet these needs so you can be more productive in your role?

Volunteer work is an opportunity to build up skills and experience. It creates the same feelings of self-worth, sense of identity and purpose as a paid job.

The New Zealand government recognises the value people with a disability can bring to a workforce and the under representation of this community in the labour market. They have set up a number of employment related services and supports for people with a disability, including training and apprenticeships. The list of all government-funded or supported services are available on the website [Employment New Zealand](#).

Diversity Works New Zealand (formally the EEO Trust) is the national body for workplace diversity and inclusion. They can be contacted on 0800 348 377 or by visiting their website <https://www.diversityworks.nz>.

Remember, it is illegal for employers to discriminate against people because of ethnicity, sexual orientation, gender, marital status, religious belief, or disability. Equal rights are demanded by the Human Rights Act, 1993, and the Equal Pay Act, 1972. You can seek information about your rights on [Health and Disability Commissioner](#) website or [Human Right Commission](#) website.

### References:

- Online Mendelian Inheritance in Man, <http://www.omim.org/entry/614575>
- US National Library of Medicine, National Health Institute:  
<https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3128688/>
- <http://www.dizziness-and-balance.com/disorders/central/cerebellar/CANVAS.html>