CANVAS

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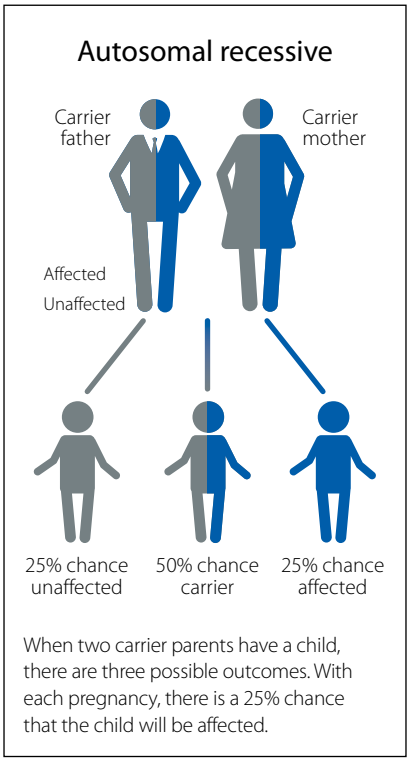
CANVAS is an easy-to-remember acronym that describes an adult-onset slowly progressive neurologic disorder that has an average age of onset of 50-60 years.

People with CANVAS have cerebellar ataxia (coordination problems), neuropathy (peripheral nerve damage), and vestibular areflexia (loss of vestibular function) and combined they form a recognisable syndrome affecting balance and gait, which is progressive, worsening over time.

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.

Common features of this condition include "dolls head reflex" upon examination, which is an 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 (dysarthria).

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.



When two carrier parents have a child, there are three possible outcomes. With each pregnancy, there is a 25% chance that the child will be affected.

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 CANVAS will show cerebellar atrophy.

CANVAS is a rare but likely under-diagnosed disorder. It is inherited in an autosomal recessive pattern. This means two altered genes are inherited, one from each parent. The parent's health is unaffected as the condition requires two altered genes to occur.

Diagnosis

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

Earlier this year it was established that CANVAS is caused by an alteration in a gene called RFC1 (replication factor C) on chromosome 4. Usually the gene contains a simple tandem pentanucleotide AAAAG repeat of 11 (AAAAG(11)), whereas in CANVAS the repeat expansion is different (AAGGG(n)) and the size ranges from about 400 to 2,000 repeats, with the majority of cases having about 1,000 repeats.

Genetic testing for this pentanucleotide repeat expansion will soon be available.

Treatment

There is no known cure for CANVAS and treatments are generally to manage symptoms such as chronic

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cough, pain and dizziness. CANVAS is progressive and a person with this 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 will be helpful.

Speech therapy and communication devices such as writing pads and computer-based devices may benefit those 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 regularly with visits to physiotherapists, occupational therapists and other specialists as needed.

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