FOR HEALTH PROFESSIONALS

MS AUSTRALIA

PRACTICE

Ataxia and tremor in people with multiple sclerosis (MS)

Ataxia and tremor in people with multiple sclerosis (MS)

Ataxia and tremor are common yet difficult symptoms to manage in people with MS — often requiring the involvement of a multidisciplinary team. Early intervention is important in order to address both the functional and psychological issues associated with these symptoms.

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1.0 Definitions

Ataxia

Ataxia is a term used to describe a number of abnormal movements that may occur during the execution of voluntary movements. They include, but are not limited to, incoordination, delay in movement, dysmetria (inaccuracy in achieving a target), dysdiadochokinesia (inability to perform movements of constant force and rhythm), and tremor.[1] Ataxia can be the result of damage to the cerebellum (cerebellar ataxia) or the posterior columns of the spinal cord (sensory ataxia) or dysfunction of the vestibular system (vestibular ataxia).

Tremor

Tremor is defined as a rhythmic, involuntary, oscillating movement of a body part. There are two main classifications of tremor — resting tremor and action tremor. Resting tremor is present in a body part that is completely supported against gravity and is not voluntarily activated. Action tremor is produced during body part movement.[5] Action tremor may be further sub-classified into intention, postural, kinetic, and isometric tremor, of which intention and postural are the two most prevalent forms (refer to section 3.1 'Pathophysiology and clinical characteristics of tremor').

2.0 Incidence and impact

Studies report that up to 85% of people with MS may experience ataxia and/or tremor at some point in time. Ataxia and tremor are often accompanied by other symptoms such as weakness, spasticity, and reduced sensory or visual input.[6] It is not surprising that ataxia is reported to affect function in up to 32% of people with MS.[7] Greater levels of ataxia in people with MS appear to correlate with higher levels of respiratory symptoms,[8,9] and severe tremor correlates with the presence of dysarthria.[10]

3.0 Pathophysiology and clinical characteristics

3.1 Ataxia

There are three forms of ataxia that may occur in isolation, or in combination, in people with MS. These are sensory, cerebellar, and vestibular ataxia.[6]

Sensory ataxia is a result of damage to the sensory pathways responsible for transmitting proprioceptive information.[11] Sensory examination usually reveals the loss of position and vibration sense, with a significant worsening of symptoms when the eyes are closed.[3] When comparing movement tests at constant speed, performance is significantly worse when the eyes are closed. This is due to loss of proprioception (sensation of joint and body part position).

Vestibular ataxia may result from damage to the vestibulocochlear nerve (VIII cranial nerve) or its central connections, which are located in the brainstem and vestibular nuclei. More commonly, vestibular ataxia results from cerebellar damage. The cerebellum is the major port of integration for all the aforementioned neural connections. Symptoms include, but are not limited to, vertigo (dizziness), nausea, loss of balance and nystagmus (rapid rhythmic and repetitious involuntary eye movements).[11]

Cerebellar ataxia is a result of damage to the cerebellum, or parts of the brain that connect to the cerebellum. This includes cerebellar peduncles and the pons or red nucleus. The symptoms of cerebellar ataxia are dependent on the location of the damage and may include upper limb incoordination, disturbance of posture and walking (ataxic gait), dysmetria, dysdiadochokinesia, nystagmus or speech (dysarthria).[1] Cerebellar ataxia is often significantly worse when comparing movement tests at a faster speed. Closing the eyes may only minimally worsen test movement execution.

3.2 Tremor

Tremor is often experienced as rhythmic shaking movements that can be small or large in amplitude. Tremor in people with MS can affect the head, neck, vocal cords, trunk or limbs, and is predominantly action tremor.[10] A classification system for tremor was published by Deuschl *et al*. in 1998 and may be a useful reference for the practicing clinician.[5]

The upper limbs are the most commonly affected body parts for tremor in people with MS.[10,13]

The pathophysiology of tremor in MS is complex and not well understood due to the multi-focal damage typical of MS. However, the predominance of action tremors suggests that the cerebellum and its connections are the most likely source of tremor production.[12]

The two most prevalent forms of action tremor in MS are: [12]

Intention tremor – tremor present during target-directed movement. Tremor amplitude increases during visually guided movement towards a target.

Postural tremor – tremor present while voluntarily maintaining a position against gravity.

4.0 Assessment

The assessment of ataxia and tremor should be integrated into a functional analysis assessment using general measures of function, (such as the 10-metre walk test, timed up-and-go test, 9-hole peg test and the spiral test) as well as specific measures of ataxia and tremor.

Three of the specific tools devised to measure ataxia or tremor in people with MS include:

International Cooperative Ataxia Rating Scale (ICARS): [14] This scale utilises traditionally known tests such as the finger-to-nose, heel-to-shin, walking, drawing, speech and oculomotor movements tests with qualitative analysis. ICARS has been shown to be a reliable and repeatable measure of ataxia.[15]

Fahn Tremor Rating Scale (FTRS): [16] The FTRS assesses intention tremor in the terminal period of the finger-to-nose test. A modified version, without the functional disability rating scale, has been assessed for use with people with MS. It was found to be a reliable and potentially useful tool for assessing movement disorders in people with MS.[17]

0–10 tremor severity scale: The scale was devised by Bain *et al*. in 1993. It has been shown to be a valid and reliable measurement tool for tremor severity in people with MS.[18]

The cerebellum has a highly integrative role for many of the body systems including vision, sensation, temperature, limb position and movement. The results of any assessment must be interpreted in light of these systems due to their potential to influence the measures.

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5.0 Management

Although ataxia and tremor are very common, few studies have investigated the efficacy of management of these symptoms, which may still be perceived as nonresponsive to medical and rehabilitative therapies.[19] The National Institute for Health and Clinical Excellence in the United Kingdom (www.nice.org.uk) has developed a set of guidelines known as the 'NICE' guidelines for the management of MS symptoms. These guidelines, which are based on a critical review of available evidence, recommend that:

- Any person with MS who experiences a limitation of activities due to tremor should be assessed by a specialist rehabilitation team for appropriate pharmacological management, treatment techniques, and equipment.

- If problems remain severe and intractable, the person should be assessed by a neurosurgical team from a specialist centre. Their suitability for surgery to reduce ataxia can be addressed.

Randomised controlled studies that demonstrate changes in locomotion function through exercise interventions for people with cerebellar dysfunction (with or without MS) are limited. A Cochrane review of ataxia in MS found limited support for pharmacotherapies, neurosurgery or neuro rehabilitation for the improvement of ataxia, tremor or function.[19] Two studies investigating physiotherapy failed to exclude individuals with other deficits, consequently compromising the integrity of the research. However, strong evidence from animal studies show recovery from cerebellar dysfunction through adaptation and compensation of neighbouring areas of the cerebellum.

Success of rehabilitation for people with MS is dependent on a number of factors. Cerebellar dysfunction has been highlighted as one such inhibiting factor.[20] People with MS with cerebellar damage seem to experience greater difficulty improving movement execution than healthy individuals. More severely affected individuals demonstrate no learning of automated movements.[4] The combination of cerebellar and other symptoms, such as weakness, spasticity, and/or visual loss, may further worsen rehabilitation outcomes, as is experienced by people with stroke.[21]

Conversely, the management of these other symptoms may assist in reducing the overall impact of ataxia and tremor on physical and social abilities. The decomposition and slowness of movement may be difficult to improve as these strategies are likely to be employed by people with ataxia in order to optimise function.

5.1 Physiotherapy

Few studies have looked at the effectiveness of different physiotherapy interventions on reducing ataxia and tremor, as well as improving functional activities. To date, the scientific evidence used to support clinical practice in physiotherapy is of low levels (levels 2a–4). In addition, translating research success into practice is hampered by a lack of appropriate detail regarding these therapeutic interventions.

Physiotherapy interventions used to manage ataxia include:

- Exercises that promote dynamic and static stability of the trunk and limbs. This is achieved by practising tasks that rely on adapting or developing strategies to cope with increasingly demanding conditions. For example, tandem walking and walking on different surfaces, walking with the eyes open and closed, and starting movements slowly with gradual increase in movement complexity, postural stability, or speed (evidence level 2a–4).[23–27]

- The use of patterning and proprioceptive neuromuscular facilitation.[27] Use mat activities with proprioceptive neuromuscular facilitation techniques, combined with balance training, to help enhance postural stability and balance reactions. Introduce crawling, kneeling, half-kneeling positions, as well as co-contraction of agonist and antagonist muscles in static and dynamic balance training. These techniques employ the underlying clinical assumption that repetition and practise of patterned and facilitated movements can improve coordination.

- Current evidence suggests that the use of weights to manage ataxia, or reduce ataxia and improve function, is of no benefit. In fact the use of weights is more beneficial in reducing tremor and improving functions (see below). One of the reasons for misinterpretation around the clinical use of weights is that the majority of studies on ataxia and tremor simultaneously investigated the effect of weights on intention tremor and functional tasks, or with tests traditionally used to quantify ataxia.[28,29]

Each of these aforementioned strategies requires extensive practise, especially when the aim is to enhance cortical re-organisation for skill acquisition and re-learning.

Physiotherapy interventions used to manage tremor include:

- Weights – The use of weights to manage tremor is currently demonstrated to have positive outcomes with improvement of intention tremor frequencies;[29–31] however, this benefit has not been found among other types of tremor.[31] The mechanism by which the improvement of tremor occurs appears to be mechanical, with increased muscle activity dampening tremor amplitude.[29] Caution is required when using weights to dampen tremor since the application of too much weight may worsen tremor frequencies (evidence level 4).[30,31]

- Full limb cooling – This technique is advocated to produce a temporary reduction in tremor, which can last for up to 30 minutes (evidence level 4).[32] Undertaking full limb cooling without appropriate training is not recommended.

Overall, there is no strong evidence to support the clinical use of the aforementioned therapies for ataxia and tremor. However, physiotherapists should consider the value of these therapies for the temporary relief of ataxia and tremor symptoms. They may allow a client to perform functions that are otherwise difficult or impossible.

Compensatory strategies may be employed when physiotherapy techniques fail to improve ataxia and tremor symptoms or general function. Compensatory strategies aim to reduce joint movement complexity, reduce movement speed and/or encourage single joint movements. Occupational therapy expertise plays a key role in this form of ataxia and tremor management and may involve splinting, equipment and/or adaptive supports.[3]

In Australia, geography can limit access to health services for people with MS. However if local physiotherapists are educated about MS, people with MS are better serviced and can access support from specialists as required.

5.2 Pharmacotherapy

To date, medications used to treat ataxia have shown limited efficacy but are associated with extensive side effects. There are currently no drugs available that can completely subdue tremor.[6] Several drugs are being trialled for the reduction of tremor such as propranolol, clonazepam, primidone, isoniazid, buspirone, ondansetron. The hepatotoxic effects of some of these drugs often prevents their use at optimal doses and limits their long-term use.[27]

In recent years anecdotal evidence has suggested that cannabis and its major component, cannabinoid, may have beneficial effects on MS-related symptoms such as pain, tremor, and spasticity. However, double-blind, randomised, controlled trials have found no beneficial application of cannabinoid in the management of tremor or ataxia. In addition the use of this drug can have serious side effects.[33–36]

5.3 Surgical intervention

Surgical intervention for the management of tremor is only considered when pharmacotherapy has been unsuccessful and the client's functional status is severely impaired by the tremor. Surgical interventions used to alleviate MS-related tremor include thalamotomy and thalamic stimulation (deep brain stimulation).[6,37] The long-term results of deep brain stimulation are theoretically more promising than thalamotomy. Several case series have demonstrated a significant reduction in tremor following thalamic deep brain stimulation in people with MS. However at long-term follow-up tremor had returned in all subjects, albeit to a lesser extent than prior to surgery.[38] Deep brain stimulation carries a significant risk of transient and permanent complications.[39] Investigations of thalamotomy have been limited to case study reports only. To date, few investigations have been undertaken in the area of surgical management of ataxia.

6.0 Summary

- The best therapy outcomes are achieved in individuals with minimal ataxic symptoms who commence intervention soon after symptom onset.
- Other symptoms associated with ataxia and tremor (e.g., weakness, spasticity, dizziness or fatigue) must be addressed to minimise their influence on function.
- Physiotherapy strategies that involve repetitive and progressively more challenging functional training, or repetitive neuromuscular facilitation techniques, may be useful in the management of ataxia in people with MS.
- Weights may be useful in the management of tremor to achieve improved function.
- Full limb cooling may be useful to achieve a temporary reduction of tremor and a corresponding temporary improvement in function or practise of tasks.
- Training compensation strategies may be the most effective intervention in people with severe ataxia or tremor.
- Individuals with severe ataxia should be monitored and treated as required for respiratory impairments.
- People with MS should be considered for referral to their doctor or specialist to discuss pharmacological or surgical interventions that support their overall therapy goals.

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MS Practice//For Health Professionals

MS Practice is an initiative of MS Australia (MSA). MS Practice is an online resource designed to support allied health professionals in the symptom management of people with multiple sclerosis (MS). The series addresses the various symptoms associated with MS, providing health professionals with evidence-based information and clinical practice recommendations to enhance the quality of care and outcomes for people with MS. The MS Practice topics were identified by the MSA Physiotherapy Network.

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Credits

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MS Australia

MS Australia is a not-for-profit organisation that has been supporting people with MS since 1956. Through state-based MS Societies, MS Australia strives for a world without MS through quality research and service excellence for people with multiple sclerosis, their family and friends, and healthcare professionals.

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This handout is intended to provide information to support current best practice for the management and treatment of physical impairments in people with MS. While the information is available to all health professionals, there are details that are most relevant to physiotherapists, exercise physiologists, and people who are qualified to provide exercise opportunities for people with MS. MS Australia has made every effort to ensure that the information in this publication is correct. MSA does not accept legal responsibility or liability for any errors or omissions, or for any physical or financial loss incurred whilst participating in the exercises or activities outlined in the MS Practice handouts. Be sure to seek advice from the sources listed.

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