European Reference Network | Neurological Diseases (ERN-RND)

ATAXIA Ataxia UK

euro ataxia

MUSCLE TIGHTNESS & STIFFNESS – "SPASTICITY" IN ATAXIA

Spasticity is one of the symptoms experienced by some people with ataxia and this leaflet:

a) gives a definition of spasticity,
b) explains the different treatment options
c) provides recommendations on how to get treatment.

The text has been adapted from the **"Management of the ataxias - towards best clinical practice" guidelines for healthcare professionals (Ataxia UK, third Edition, July 2016).**

It is recommended that this information be discussed with a healthcare professional.

Definition

Spasticity is the presence of increased muscle tone or hypertonia, which can cause muscle stiffness, spasms and pain. Persistently raised muscle tone can result in abnormal posturing of body parts which if prolonged can result in muscle and tendon shortening, fixed joints and ultimately contractures[1].

Contractures are characterized by permanent reductions in the range of motion of joints and muscles. Usually contractures occur in your arm or legs.

Spasms are sudden, involuntary and often painful muscle contractions which are often associated with spasticity and provoked by muscle stretch or other stimuli[1]. They may be transient or prolonged.

Ataxia and spasticity can coexist in a large number of congenital, genetic or acquired conditions including common conditions such as stroke, multiple sclerosis (MS), cerebral palsy and head injury. Spasticity is prominent at presentation in some people with ataxia who may subsequently be diagnosed with a spastic ataxic syndrome, or one of the hereditary spastic paraplegias (HSPs). Spasticity can be a less prominent or later feature of many of the ataxic disorders[2].

Spasticity can affect many parts of the body contributing to a range of symptoms seen in progressive ataxia including:

- difficulty with walking
- lack of manual dexterity (e.g. writing, clumsiness)
- dysarthria (slurred speech)
- dysphagia (swallowing problems)

Focal spasticity and **night cramps** will be discussed in a different section of the leaflet.

Treatment of spasticity

Why treat spasticity?
→ To optimize mobility, standing capacity, upper limb function, speech and swallowing
→ To reduce symptoms of pain and spasms, especially those at night which impair sleep and contribute to daytime fatigue
→ To improve transferring from a chair, sitting posture, washing and dressing, and so promote independence and reduce carer reliance
→ To prevent contractures and so reduce the development of chronic disability

Spasticity can be worsened by pain, infection, diarrhoea, constipation, being unable to pass urine, tight clothing or poor posture. Therefore, before treating the spasticity with medications or increasing an anti-spasticity medication, it is crucial to try to identify and treat any other cause of spasticity.

Treatments of spasticity available are:

- physiotherapy-based
- medications
- surgery

Ideally the treatment should follow the sequence above, moving from one to the next in that order if the previous has failed or provided incomplete benefit.

••• It is vital that medications and surgical techniques be discussed in detail with patients before proceeding with them. The doctor or therapist should outline their risks and benefits. •••

Physiotherapy

In addition to preventing spasticity complications, such as joint contractures or osseous deformities, physiotherapy has a vital role to play in providing exercises and educating patients and carers in correct posture, muscle use and the avoidance of spasticity triggers such as pain and infection.

Medications

Although there is little evidence of the efficacy of anti-spasticity interventions specifically in cases of spastic ataxia, a greater evidence base exists in commoner conditions causing spasticity like MS. Since the underlying mechanisms generating spasticity and spasms are similar, doctors normally use evidence from related conditions to make decisions on medications.

Anti-spasticity drugs can sometimes worsen mobility by uncovering underlying muscle weakness if the patient relies on stiffness for walking[7]. It is important to discuss this with your doctor before any medication for spasticity is started. If worsening of mobility or other functions occurs it is important to inform the doctor who may reduce the dose or stop treatment. Generally speaking, anti-spasticity medications should not be stopped abruptly, particularly from high doses.

Anti-spasticity medications should be increased slowly to minimise side effects.
Most doctors start with the following oral medications for treatment of spasticity (usually in this order):

1. baclofen,
2. tizanidine,
3. gabapentin,
4. clonazepam,
5. dantrolene sodium or
6. diazepam

Long-term use of diazepam is not recommended apart from in very severe cases. Many other oral medications have shown some evidence as anti-spasticity agents in other conditions including methocarbamol, levetiracetam, lamotrigine, pregabalin, progabide, clonidine, piracetam, vigabatrin, prazepam, cyproheptidine, L-threonine, thymoxamine, orphenadrine and 3,4-diaminopyridine. However, these are rarely used in practice.

If these are not successful or not tolerated, greater concentrations of baclofen in the central nervous system can be achieved with reduced side effects by the use of an intrathecal (into the fluid around the spinal cord) baclofen infusion. **This intervention is only offered in highly specialised centres and requires careful planning and long-term follow-up. It is only relevant in a small number of patients[3].**

Anecdotally, it is known that some patients with ataxia benefit from the use of cannabis products in reducing pain and spasticity. However, no trials focusing on spasticity in patients with ataxia and using cannabis have been done. Although some studies have shown benefits in treating spasticity in MS[4], the largest of these trials[5,6] failed to show significant reductions in objective markers of spasticity, so further research is required.

Focal Spasticity

Focal spasticity (i.e. spasticity localised in a part of the body), particularly in small muscles, is probably best treated with intramuscular botulinum toxin injections[8]. It is advisable to be referred to a specialised clinic for such treatment. There is evidence that this benefit is prolonged by additional therapies such as stretching, taping, casting, artificial external devices or electrical muscle stimulation.

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It is therefore very important that such injections are accompanied by a course of physical therapy or an exercise programme at the time or immediately after injection.

Night cramps associated with spasticity

Spasticity can be associated with painful nocturnal cramps. The medical treatments described above are commonly used for these, especially baclofen or gabapentin. In the elderly, quinine sulphate has been used extensively for these, but it is associated with serious adverse events (particularly cardiac problems, blood disorders and deafness) and so it is generally not recommended:

- for long-term use,
- for patients with cardiac conditions,
- or for patients with Friedreich's ataxia (because of associated cardiomyopathy)

Surgical treatments

Surgical treatments are generally only considered when physiotherapy and medications have not worked as they are often permanent and destructive. However, they may be considered first line in exceptional cases. Surgical treatments include a series of procedures depending on the severity and the cause of the spasticity. Surgical treatments include orthopaedic procedures such as tendon lengthening, tenotomy or tendon transfer; and neurosurgical procedures such as peripheral neurotomies, dorsal rhizotomies and microsurgical ablation of the dorsal root entry zone ('DREZotomy')[9].

Summary of recommendations

1. Careful assessment by a neurologist, with advice from a physiotherapist, to decide on the type of treatment.
2. Factors which worsen spasticity should first be treated, such as pain, infection, diarrhoea, constipation, urinary retention, tight clothing or poor posture.
3. Physiotherapy should then be used to treat spasticity.
4. If that does not provide complete benefit, medications should be considered. Medications for generalised spasticity include baclofen, tizanidine, gabapentin, clonazepam, dantrolene sodium or diazepam.
5. To treat localised spasticity, particularly in small muscles, refer to a specialised clinic for treatment with intramuscular botulinum toxin injections, followed by physiotherapy and exercise.
6. Surgery should be considered in cases where physiotherapy and medications have not worked.

References

This is the list of publications cited in this document and used to write the different sections:

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ERN-RND website: www.ern-rnd.eu
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About Ataxia UK

Ataxia UK is the leading national charity in the UK for people affected by any type of ataxia. We fund research into finding treatments and cures, and offer advice, information and support to people affected by the condition.

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Euro-ataxia is an international non-profit association whose member organisations work together to help people with progressive ataxia lead their best life. We do this by building a strong organisation that represents people with progressive ataxia throughout Europe.

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