CRAMPS AND JERKS: ABNORMAL MOVEMENT

In the 1999 survey, 81% of respondents experienced problems with muscle twitches/cramps/jerks.

These neuromuscular disorders are very common and troublesome problems for many arachnoiditis patients. Stiffness affected 79% of respondents in the survey.

This non-specific term may however, include joint stiffness as well as muscle stiffness.

There is a considerable range of muscular problems, from small, painless, transient twitches right through to extremely painful spasms, and sustained muscle stiffness.

There are various manifestations of abnormal muscle fibre activity. It is important to clarify the terminology in order to best understand the different problems.

Twitches:

Fasciculations: Spontaneous discharge of an axon causing contraction of muscle fibers in rippling unit, thus producing visible rippling of the muscle; it tends to be in small, isolated areas; it can occur in healthy people, usually in the calf or hand, but in those who have spinal problems, it may signify a dorsal horn involvement.

It may also be a consequence of motor nerve fibre irritability. Fasciculations may be chronic, lasting for weeks or months without evidence of weakness or muscle wasting or indeed any evidence of disease.

Twitching associated with low serum calcium is another fascicular muscle activity.

Fibrillations: spontaneous contraction of a single muscle fibre, not usually visible.

May increase with muscle warming. Fibrillation potentials seen in EMG testing do not necessarily indicate neurogenic denervation, but can also arise from muscle diseases like polymyositis.

Myokymia: irregular firing of multiple muscle fibres spontaneously; it tends to be in bursts; if persistent, it is termed Neuromyotonia, which is associated with sustained muscle contraction. Myokymia may affect the eyelids (benign); it may signal an underlying problem such as thyrotoxicosis (hyperthyroidism). Myokymia in the facial muscles may signal MS.

Spasms: arachnoiditis patients tend to refer often to ?muscle spasms', which in fact may well actually be cramps or myoclonic jerks. The following section attempts to cover the commonest spasmodic problems.

Aldrete's survey([1]) found that

64% of respondents experienced ?muscle spasms'.

Cramps: Sudden involuntary painful muscle contractions. Most often, cramps occur following voluntary contractions (e.g. after exercise), and occasionally during rest or sleep. They usually involve single muscles rather than groups.

Cramps may be seen in healthy people, usually in the calf muscle. In patients with neurological disorders, cramps may occur in other muscles and may be associated with partial denervation or other neuromuscular conditions, as well as in hypothyroidism (underactive thyroid gland) electrolyte disturbances (metabolic abnormalities affecting salts in the blood). Nocturnal leg cramps are a common problem in arachnoiditis.

Causes:

- Familial
- Hypothyroid
- Electrolyte & metabolic disorders
- Dehydration: Diuretics; Haemodialysis
- Metabolic: Low Sodium, Magnesium, Calcium, Glucose
- Neurogenic
- Spinal Stenosis
- arachnoiditis

- <u>Drugs & amp; Toxins</u> e.g. Caffeine, Diuretics, Labetalol, Lithium, Nifedipine, Terbutaline, Tetanus

, Theophylline,

Vitamin A

(not an exhaustive list)

- Note: Myotonic cramps are not usually painful Non-muscle disorders: Usually with

Normal CK* & EMG (* creatine kinase, an enzyme which is raised in inflammatory disorders)

- Connective tissue disorders: Fasciitis ; Systemic
- Bone: Osteomalacia; Fracture (Hip); Myeloproliferative; Fluorosis
- Endocrine : Thyroid ; Parathyroid
- Vascular: Ischaemia; Thrombophlebitis
- Polymyalgia syndromes
- Joint disease: Immune; Hypermobility syndromes; Gout; Degenerative
- Polyneuropathy including Guillain-Barr? syndrome
- CNS: Restless legs; Dystonias (Focal)

Treatment:

Acute cramp may be relieved by stretching the relevant muscle.

Preventive measures include: avoidance of excessive sugar intake and caffeine.

A diet with plenty of potassium rich foods such as bananas is helpful.

Regular stretching of calf muscles during the day, using a footboard at night, or dangling the feet over the edge of the bed if lying prone.

Quinine 300mg (especially for nocturnal cramps); phenytoin or carbamazepine (anti-convulsants). Note quinine interacts with other medication such as: cimetidine, digoxin, anticoagulants, antacids. You can also obtain quinine in Tonic Water.

Vitamin E 400-800 IU per day has also been reported as being helpful. Calcium supplements (0.5-1g four times a day) may be useful, as may riboflavin 100mg 4 times a day (vitamin B2): note that some common medication used in arachnoiditis patients: e.g. tricyclic antidepressants (amitryptiline being the most frequently used) can contribute to riboflavin deficiency. Magnesium 400mg daily may also be used.

The anticonvulsants carbamazepine 200mg twice or 3 times a day, Gabapentin 400mg three times a day or phenytoin 300mg 4 times a day; methocarbamol (Robaxin), verapamil 120mg 4 times a day, tocainide 200-400mg twice a day, and diphenhydramine (Benadryl) 50mg 4 times a day have also been reported as helpful but there are no specific scientific studies of their use. It is essential normalise any metabolic abnormalities.

Myoclonus: a brief, sudden, shock-like muscle contraction, mediated by an electrical nerve discharge originating in the central nervous system. Secondary*

Myoclonus is seen in conditions in which there is central nervous system damage, which, in arachnoiditis, is likely to be related to the spinal cord, so would be termed spinal myoclonus (other types include peripheral myoclonus from an electrical impulse in a peripheral nerve).

Myoclonic jerks can be extremely debilitating as they interrupt normal posture and movement. The muscle spasms may be uncontrollable and may be both forceful and painful.

They may be triggered by movement (?action' myoclonus), so may not be present when at rest or asleep.

There may also be sudden reduction in muscle contraction which prevents normal movement: this is termed negative myoclonus (asterixis).

A very mild, harmless form of myoclonus is seen in healthy people when they hiccup or jerk involuntarily as they fall asleep.

*primary is either inherited or of unknown origin (idiopathic)

Drug-induced myoclonus: about 80 causal agents (toxins and drugs) including:

- Tricyclic antidepressants e.g. amitriptyline
- SSRIs e.g. Prozac
- Penicillin
- Morphine
- Hydromorphone (an opiate related to morphine)
- Phenytoin
- Midazolam
- Pseudoephedrine (available in some over-the-counter common cold preparations)

Treatment:

Clonazepam (benzodiazepine), valproate (anticonvulsant); some reports of baclofen, fluoxetine (an SSRI antidepressant), propanolol (antihypertensive) and 5hydroxytryptophan (5-HT) being of help.

There is no treatment for negative myoclonus (asterixis, postural lapses) <u>Complex repetitive discharges</u>: Bursts of recurrently firing complex muscle action potentials which cause discharges of groups of muscle fibres, firing in a regular pattern.

They can begin spontaneously or after muscle irritation and are often associated with radiculopathy (nerve root pathology) such as that seen in arachnoiditis. Occasionally, frequent discharges will produce muscle hypertrophy (increase in size) which is usually in one limb only (unilateral).

Less common miscellaneous problems:

Myoedema: Localized, persistent, electrically silent swelling of muscle after percussion, lasting a few seconds; it may occur in myxoedema, a feature of underactive thyroid, which may be seen in arachnoiditis patients who have had a myelogram in the past.

Myotonia: Delayed relaxation of skeletal muscle following voluntary contraction, present with initial activity, which usually abates after repeated muscle activity.

Notably, it can be caused by colchicine, which has been occasionally used to treat arachnoiditis patients. Most cases, however, are congenital and not relevant to arachnoiditis.

Dystonia: movement disorder in which sustained muscle contractions cause twisting and repetitive movements or abnormal postures. The movements are involuntary and may be painful; they may affect a single muscle or a group of muscles, or even the entire body.

Writer's cramp is a form of dystonia. It is a spasm induced by a specific task. It affects both men and women and age of onset is 20-70; it may only affect one hand, but in some patients (around 25%) who learn to write with the other hand, it may become bilateral.

It affects the ability to write legibly, the hand being held stiffly, so that writing becomes jerky. The act of writing may become painful. In patients in whom the condition progresses to affect the ability to perform other tasks, it becomes known as dystonic cramps. Roughly half of dystonia cases are primary or idiopathic, i.e. there is no known cause.

Some causes of secondary dystonia include:

- Drugs: e.g. tricyclic antidepressants, SSRIs, buspirone, antihistamines, calcium channel blockers, haloperidol, levodopa, lithium.

- Syringomyelia (complication of arachnoiditis); meningitis
- Vascular disease including arteriovenous malformation

The commonest from of dystonia is torticollis, in which the neck muscles are affected, causing the head to turn to one side, or be pulled forward or backward.

Blepharospasm: the second most common dystonia, is defined as

" a chronic, unremitting, bilateral, variably progressive dysfunction of the nerve that controls the muscles around the eye."

It causes an uncontrollable closure of the eyelids, sometimes for longer than the blink reflex, which therefore may interrupt the ability to see; it usually only affect one eye, although both may be affected.

In some cases, other muscles in the face also twitch.

Hemifacial Spasm is a variant of blepharospasm which causes chronic twitching or spasms on one side of the face, affecting the muscles supplied by the facial nerve. It can result from Bell's palsy, which is an inflammation of the facial nerve causing temporary facial weakness, if there is abnormal regeneration of the nerve.

In severe cases, it may be difficult to open the mouth, thus impairing the ability to eat, speak and swallow.

Meige's Syndrome is a similar condition affecting the long facial and neck muscles as well as the muscles around the eye.

Spasticity

Stiffness of the muscles, often with generalised increased muscle tone, is a common problem in arachnoiditis patients.

The term, Spasticity (from the Greek spastikos, meaning ?to draw in') encompasses:

- involuntary muscle contractions
- stiffness of the muscles; may alternate with floppiness
- hyperactive reflexes (may include clonus: see below)

Commonly there is reduced strength, impaired dexterity. This problem can be quite debilitating for the patient. It may also accompany other forms of hyperactive muscles such as cramps and spasms.

These symptoms signify damage to the central nervous system rather than to the muscles or the peripheral nerves which supply them, (peripheral nerve damage tends to cause the opposite problem: flaccidity, or loss of tone).

The problem arises because there is some degree of loss of the regulating influences of the brain, so that the automatic, reflex activity which occurs at a spinal level, is unopposed.

Any sensory stimulus below the level of spinal pathology, such as a change in body position or having a bowel movement, can trigger this reflex activity of muscle movement.

Spasticity may be seen in the following conditions:

- cerebral lesions including stroke, MS, cerebral palsy etc.

- spinal lesions including spinal cord injury, inflammatory disorders (MS, arachnoiditis),

compressive lesions (e.g. osteophytes: bone spurs, in the neck) etc.

Terminology:

Muscle tone: the resistance of muscles to passive stretch: the amount of tension a muscle has at rest. Normal tone is designed to resist the effects of gravity on posture and movement whilst being low enough to allow freedom of movement.

Hypertonia (increased tone) resistance to passive movement; not dependent on speed of movement; can be with or without spasticity.

Spasticity: an increase in resistance to sudden, passive movement; velocity dependent (the faster the movement, the stronger the resistance).

Definition:

"a velocity-dependent increase in tonic stretch reflexes, where faster passive movements meet with increasing resistance".([2])

This resembles the response of a bicycle tyre pump.

Clonus: a reflex which is a spasmodic alternation of muscle contraction and relaxation, usually in the calf muscle, the foot being sharply bent upwards towards the thigh and being held in mid position. Persistent clonus can interfere with putting shoes on.

Associated problems:

Spasticity tends to affect lower limbs more than upper limbs and can be exacerbated by tight clothes, infection, or pressure sores.

A significant increase in spasticity can arise due to:

- skin problems: sores/ingrowing toenail
- bladder problems: high residual volumes, infection or bladder stones
- bowel problems: constipation, impaction or haemorrhoids
- general medical problems: e.g. viral infection: ?flu etc.

As well as being uncomfortable, spasticity can significantly impair activities of daily living (ADL), including the ability to dress and take care of personal hygiene (in particular perineal self-care: bladder and bowel function).

It can also affect gait (walking), as normal gait requires normal muscle tone and good range of motion in several joints. Severe spasticity may severely affect mobility.

Treatment:

- 1. regular Range of motion (ROM) exercises: these can reduce the reflex activity
- 2. application of heat/ice in some patients

3. physiotherapy to improve function in some muscle groups: e.g. trunk control, elbow extension.

4. orthotic devices: e.g wrist splint: placing a joint in a position that allows maximum function

5. Drug therapy: Baclofen, diazepam and dantrolene sodium are the most commonly used medications.

Tizanidine (Zanaflex) is a relatively novel drug being used.

Baclofen: 5mg 3 times a day (can be increased every few days to maximum of 80mg total daily dose). Side effects: lethargy, weakness, nausea, pins and needles;

note: must not be discontinued suddenly as this could precipitate seizures.

Diazepam and clonazepam: benzodiazepine drugs; primarily used in patients with spasticity of spinal origin. Side effects include sedation, muddled thinking and dependence (hence withdrawal on stopping: not the same thing as ?addiction' in the generally accepted sense of the term).

Dantrolene: more often used in spasticity of cerebral (brain) origin.

Tizanidine: thought to cause less weakness; has been used in patients with MS.

Dose starts as 4mg at 6-8 hour intervals (max. daily dose should not exceed 36mg).

The UK Tizanidine Study Group found that whilst tizanidine improved spasticity without compromising muscle strength, there was no apparent improvement in functional measures or activities necessary in ADLs. Night-time insomnia may be more significant in Tizanidine than Baclofen.

Other drugs: clonidine: a drug developed to treat high blood pressure; rarely used specifically to counter spasticity; available as a skin patch.

Gabapentin: may be prescribed to relieve neuropathic pain: has also been found to be effective in treating spasticity in MS.

Botulinum toxin: injections of the toxin responsible for botulism (Clostridium

botulinum) into the relevant muscles has been found to relieve localised muscle spasm (e.g. blepharospasm); it achieves this by damaging the nerve fibres which transmit the signals to the muscles to contract.

Localised paralysis of the muscles begins within 24-72 hours, being maximal at 5-14 days.

However, the effects are temporary, lasting 12-16 weeks.

It has been helpful in patients with severe spasticity after stroke or with severe MS.

Intrathecal baclofen: administered via the ?pump': is being used in children with cerebral palsy.

Therapeutic nerve blocks: use of phenol (3-6% solution) and alcohol (50%) solution may effect a block for weeks to months; however, there is a risk of abnormal nerve regrowth.

Note: Invasive treatments are not recommended by the Arachnoiditis Support Groups but are included here for completeness.

Restless legs syndrome

RLS is an unpleasant sensation in the legs (and occasionally the arms) that occurs at rest and is relieved by movement. It is thought to affect more than 10 million Americans (other figures suggest around 5% of the population).

The incidence increases with age, although it has been described in adolescents. It is 30% more frequent in women than in men.

It is described as feeling like "pins and needles", "internal itch", "creeping and crawling sensation", "jittery feeling", "feeling fidgety".

Sufferers try to relieve these sensations by vigorously moving their legs around or by walking about; as this occurs chiefly at night, their sleep is considerably disturbed. They are therefore tired and sleepy during the day.

There have been 4 types of RLS described:

- 1. Restlessness due to paraesthesia (pins and needles)
- 2. paraesthesias and restlessness occurring independently
- 3. voluntary restlessness to achieve relief
- 4. repetitive movements without paraesthesias

Diagnostic criteria as defined by the Restless Legs Study Group are:

- desire to move the extremities, often associated with paraesthesias/dysaesthesias*
- motor restlessness
- worsening of symptoms at rest with at least some relief by activity
- worsening of symptoms in the evening or at night.

*dysaesthesias= pain, often burning in nature

RLS may be primary or secondary to diseases or drugs.

Systemic diseases associated with RLS include:

- diabetes

- rheumatoid arthritis
- spinal cord and cauda equina damage
- radiculopathies (nerve root damage: as in arachnoiditis)
- thoracic spinal lesions
- complete spinal cord injury
- neuropathy
- vascular disease including congestive heart failure
- Parkinson's disease
- End-stage renal disease
- Iron deficiency
- Folate deficiency

Drugs include:

- Paroxetine
- Mianserin
- Phenytoin
- Caffeine
- Alcohol
- Nicotine

A paper last year ([3]) has suggested that there are 2 subtypes of RLS:

1. triggered by painful dysaesthesias associated with sensory loss (later onset, no family history)

2. no pain or sensory loss, earlier onset, family history.

Obviously by these criteria, RLS associated with arachnoiditis corresponds to the first subtype. The authors of the paper suggest that this form is amenable to treatment with neuropathic pain medications.

At the 14th. Annual Meeting of the Associated Professional Sleep Societies, Dr. Meir Kryger presented the findings of a study which established that patients with RLS are very likely to have been previously diagnosed with other conditions: including musculoskeletal disorders. 21% of men with RLS in the study (compared with 13% of the control population) had been diagnosed with back problems and 38% of women versus just 15% of the controls. 36% of men and 50% of women had been diagnosed with joint disorders (cf. Around 23% of controls for both men and women).

Furthermore: psychological disorders had been diagnosed in 44% of the men and 46% of the women (cf. 10% and 23% of controls respectively).

This amply demonstrated the point that RLS is under-diagnosed.

Treatment:

1. treat underlying causes: e.g. iron deficiency, folate deficiency

2. general measures: limit smoking, alcohol, caffeine; discontinue aggravating medication if possible.

3. drug treatment: Clonazepam (benzodiazepine); Carbamazepine (Tegretol) or Gabapentin (anti-convulsants); Sinemet, Pergolide, Pramipexole (anti-Parkinsonian); Clonidine

Muscle pain

Inflammatory myopathies (disorders of muscles) and other ?collagen-vascular diseases' can feature muscle pain and tenderness. Bearing in mind the possible link between arachnoiditis and autoimmune conditions, disorders such as polymyalgia rheumatica need to be considered.

However, in the majority of cases, muscle pain will arise secondary to the abnormal spinal dynamics seen in most arachnoiditis patients who have had (or continue to have ongoing) spinal problems.

Often a diagnosis of fibromyalgia or myofascial pain syndrome is put forward by way of

explanation; if so, it must be remembered that these features are likely to be secondary to the underlying arachnoiditis, rather than a separate disease entity. Corticosteroid withdrawal can cause muscle(and joint) pain.

Neuromuscular disorders and endocrine disease:

Note that there appears to be some association between thyroid disease of various types and previous myelogram: this is a feasible situation because myelogram dyes contain iodine.

Hypothyroidism (underactive thyroid) can cause:

- muscle pain, cramps, stiffness; pins and needles, weakness. Muscle contraction is slow, with delayed relaxation; there may be myoedema (33%), with local contracture after muscle tap or pinch and myokymia.

- ?hung up' reflexes

- Neuropathy (see below): including carpal tunnel syndrome and cranial nerve involvement (deafness, hoarse voice) and a polyneuropathy

- Ataxia (clumsiness)

Hyperthyroidism (overactive thyroid)

- Fasciculations and cramps
- Brisk tendon reflexes
- May be associated with inflammatory myopathy

Hyperparathyroidism:

- Fasciculations

Adrenal insufficiency:

- Muscle fatigue, mild weakness, cramps and muscle pain

Myopathy: "disease of muscle" that causes problems with the tone and contraction of skeletalmuscles (muscles that control voluntary movements.) ranging from stiffness (myotonia) to weakness, with different degrees of severity.

Toxic myopathies: causative agents include alcohol, Organophosphates, Colchicine, D-penicillamine etc.

Focal muscle damage can be caused by Pethidine, Pentazocine, Heroin, antibiotics in infants, Penicillin and Diphenhydramine.

Vitamin E overdose can also cause muscle problems.

Local anaesthetics can cause necrotising myopathy.

Any substance which lowers potassium levels can induce muscle weakness: e.g. diuretics (water tablets), laxatives, licorice, carbenoxolone, amphotericin B.

Prolonged corticosteroid treatment is associated with a chronic proximal myopathy.(limb girdles especially).

[1] Aldrete JA Arachnoiditis :The Silent Epidemic JGH Editores 2000

[2] Treatment of Spasticity, Mariko Kita, MD Mount Zion Multiple Sclerosis Centre 1999:

[3] Polydefkis M, Allen RP, Hauer P, Earley CJ, Griffin JW, McArthur JC Neurology 2000 Oct.

24; 55(8): 1115-21 Subclinical sensory neuropathy in late-onset restless legs syndrome.