Relief of symptoms in end-stage neurological conditions

Effective multidisciplinary team working is essential to manage neurological symptoms as end of life approaches. Coordination of care ensures timely input of specialist therapies, pharmacological measures and occasionally invasive techniques including surgery. This can impact not only on function but also aid passive care and comfort.

Many individuals with either single insult neurological conditions such as stroke or traumatic brain injury, or chronic progressive conditions including multiple sclerosis, Parkinson's disease or motor neurone disease, will have troublesome symptoms related to their underlying impairments. Specific symptoms that are often difficult to manage include spasticity and spasms, neuropathic pain, bladder and bowel dysfunction, speech, respiratory and swallowing difficulties, mood and cognitive dysfunction. Towards the end of life these can have a significant impact on not only function but also on passive care and comfort. Appropriate management of these symptoms is therefore essential; this necessitates effective multidisciplinary team working with the individual and her/her family and/or carers across health- and social-care sectors.

As an individual develops severe disability, access to hospital-based services is increasingly difficult and management becomes focussed within community teams with increasing importance of both formal and informal care networks. During this stage there are often multiple interacting problems which necessitate a holistic approach focussing on maintaining autonomy, supporting the individual and the carers in decision making, providing information and ensuring coordination of care. Effective liaison between neurology and palliative care services is essential to ensure both symptom control and end of life issues are appropriately addressed (Higginson et al, 2006).

Spasticity and spasms

The impact of spasticity on an individual is extremely variable ranging from minor discomfort to severe pain, devastating loss of function and the development of long-term problems such as pressure ulcers and contractures.

During end-stage neurological conditions, for example multiple sclerosis, the individual is usually dependant on assistance for all mobility including a hoist for transfers. The presence of severe spasticity or spasms can compromise safety in the hoist and make washing and dressing very uncomfortable for the individual.

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Poorly managed spasticity can also have serious consequences; muscle shortening and tendon or soft tissue contracture can lead to restriction of passive movement, physical deformity, difficulty with positioning resulting in the inability to seat an individual and the development of pressure ulcers.

With these issues in mind it is imperative that management is always individualized and involves families and/or carers. Knowledge of the trigger and aggravating factors detailed in *Table 1* are particularly important as they can exacerbate spasticity and its associated features. Far too often pharmacological treatment is escalated before appropriate strategies to manage bladder and bowel function, skin integrity, soft tissue length and positioning are instigated. Attention to these simple but essential areas is paramount at all stages of management and will ensure drugs and more invasive therapies are used at appropriate times and dosages (Stevenson, 2006).

Pharmacological measures to treat spasticity

The identification of appropriate treatment goals will help optimize drug therapy in terms of choice of agent but also in timing and dose. For example painful nocturnal spasms may best be managed with a long-acting agent taken at night-time which has sedative side effects (e.g. clonazepam). Alternatively stiffness and spasms, which interfere with a person's morning transfers and personal care, may benefit from medication taken on waking before the person transfers out of bed.

The oral agents most commonly used to treat spasticity are baclofen, tizanidine, benzodiazepines, dantrolene and gabapentin; all can be used alone as monotherapy or in combination with each other (Stevenson, 2006). It is of course also possible to combine oral therapy with local or regional treatments such as botulinum toxin, focal chemical neurolysis or intrathecal therapies including baclofen and phenol.

Focal pharmacological therapies

Botulinum toxin is the most widely used treatment for focal spasticity; however, for botulinum toxin injections to be effective it is essential that treatment is performed in conjunction with physiotherapy. Several randomized-controlled trials have demonstrated that botulinum toxin is effective in reducing tone in stroke, multiple sclerosis,

cerebral palsy and head injury, often through improving passive function such as ease of hygiene or dressing (Sheean, 2006).

Local injection of ethyl alcohol or, more commonly, phenol is an alternative option to botulinum toxin for focal spasticity management. Chemical neurolysis by phenol or alcohol is irreversible and results in destruction of neural tissue by protein coagulation. Injections may be targeted at peripheral nerves or motor points (intramuscular injections aimed at the parts of muscle most sensitive to electrical stimulation). Most commonly applied in end-stage neurological conditions are obturator nerve blocks with the aim of improving ease of perineal hygiene and aiding in seating posture (Barnes, 1993) or musculocutaneous nerve blocks in the upper limb to relieve elbow flexor spasticity aiding in dressing.

Intrathecal therapies (baclofen and phenol)

If oral medication in combination with appropriate physical measures is inadequate at controlling lower limb spasticity or is not tolerated then intrathecal therapies should be considered. Delivery of baclofen intrathecally via an implantable abdominal programmable pump allows very small dosages of baclofen to be effective without causing any systemic side effects (Becker et al, 1995). However, this requires general anaesthetic and frequent visits to hospital for pump dose adjustments and refills.

If this is not appropriate and spasticity is severe causing contracture and inability to seat or hoist an individual safely, these patients may be considered for intrathecal phenol treatment. As phenol is a destructive agent which indiscriminately damages motor and sensory nerves, it is reserved for those individuals who do not have any functional movement in their legs, who have lost bladder and bowel function and who have impaired sensation to their legs. Intrathecal phenol can be an effective treatment which, although it requires expert administration, does not have the long-term maintenance or cost issues that go with intrathecal baclofen treatment. The effect of a single injection often lasts many months and can be repeated if necessary (Jarrett et al, 2002). There are two distinct phases in the treatment stage; the trial and the phenol injection.

Phase one: the trial

This consists of injecting bupivacaine into the intrathecal space to produce a targeted, short-acting anaesthetic to ascertain whether or not the person's spasticity, spasms and/or pain could be managed more effectively with the longer acting phenol injection. This trial allows the patient and family or carers to temporarily experience the likely effect of having the phenol injection and can assist them in deciding whether to proceed.

One of the values of doing a local anaesthetic trial is the opportunity to re-measure and assess if there are any contractures which may limit the achievement of the goal, for example the ability to be seated. It is not uncommon for individuals, their families or carers to hope that the treatment will result in legs that can be straightened; the trial therefore allows them to appreciate the extent of any contractures and avoids any false hopes from the phenol injections.

Table 1. Common cutaneous and visceral stimuli aggravating spasticity

Cutaneous stimuli	Altered skin integrity	Red or inflamed skin
		Broken skin
		Infected skin
		Pressure ulcers
		Ingrown toenails
	Tight-fitting clothes or urinary leg bag straps	
	Uncomfortable orthotics or seating	
Visceral stimuli	Any systemic or localized infection	
	Bowel dysfunction, e.g. constipation, overflow or diarrhoea	
	Bladder dysfunction, e.g. infections or incomplete emptying	
	Deep vein thrombosis	

Phase two: the phenol injections

Typically patients require both lower limbs targeted and will therefore need two phenol injections.

Immediately following treatment the impact on a person's spasticity and positioning can be dramatic; if their posture is changed significantly this often will have consequences for their seating and lying position necessitating a seating review to accommodate the sudden change in posture and to prevent complications such as pressure ulcers developing (Jarrett, 2006).

Neuropathic pain

Pain in neurological conditions is unfortunately very common, for example in multiple sclerosis it has been reported to be present in between 29 and 86% of patients (Beard et al, 2003).

Central post-stroke pain is reported to develop in about 8% of stroke patients, and the pain is moderate to severe in 5% of patients (Andersen et al, 1995). Usually individuals present with central post-stroke pain within 1 or 2 months after a stroke, but occasionally it may be as long as 6 years after injury (Nicholson, 2004). The clinical features are extremely varied and include muscular sensations, dysaesthesias, hyperpathia, allodynia, shooting pains or visceral pain such as bloating or fullness of the bladder. Treatment is similarly varied and may include opioids, tricyclic antidepressants, anticonvulsants and even intravenous lidocaine (Nicholson, 2004). Central pain may also originate from the spinal cord where it is usually a consequence of traumatic spinal cord injury. The incidence in such patients is thought to be in the order of 60–70%. It is treated in the same way as central post-stroke pain.

Commonly chronic pain, in any neurological condition, may be dysaesthetic in nature and often involves the extremeties (Irving, 2005). This can be very debilitating for the individual interfering with mobility, sleep and contributing to depression. The most useful drugs are carbamazepine, gabapentin, pregabalin and the tricyclic antidepressants, particularly amitryptiline. Some of the newer anticonvulsants including lamotrigine, levetiracetam and oxcarbazepine have also shown some benefit in small trials (Irving, 2005).

Other forms of chronic pain may be secondary to spasticity or to immobility; back pain is particularly common in wheelchair users. Pain relief should include local measures such as heat pads and transcutaneous electrical nerve stimulation (TENS); medication may be necessary and includes short-term non-steroidal anti-inflammatory drug use and simple analgesia (Stevenson and Thompson, 1998; Beard et al, 2003).

Bladder and bowel dysfunction

Unfortunately bladder and bowel dysfunction are extremely common in individuals with neurological conditions, particularly those with spinal cord damage. Common bladder symptoms are those of frequency, urgency and nocturia. However, as bladder dysfunction increases problems of incontinence, retention and urinary tract infections can occur. Most of these are a result of a combination of detrusor hyperreflexia (causing urgency and incontinence) and sphincter dyssynergia (causing failure to empty and consequent residual volumes) (DasGupta and Fowler, 2003). Owing to the often mixed aetiology of urinary symptoms it is essential to assess bladder emptying by measuring the post-micturition residual volume before initiating any therapy; this can be done by either catheterization or transabdominal ultrasound (Fowler, 1996). If there is no residual then detrusor hyperreflexia can be treated with anticholinergic agents such as oxybutynin or tolterodine. If nocturia fails to be controlled with anticholinergics the use of desmopressin delivered by a nasal spray can be useful. Other potential agents to reduce detrusor hyperreflexia include intravesical capsaicin or botulinum toxin. Detrusor sphincter dyssynergia requires effective bladder emptying; initially this is often managed using clean, intermittent catheterization. As disability progresses an indwelling catheter needs to be considered, if this is long term a suprapubic catheter is usually preferred.

Bowel dysfunction is less frequent than urinary dysfunction but can be extremely distressing. Most commonly individuals complain of constipation and urgency, incontinence is less frequent. Management is more difficult than bladder dysfunction, but establishing a routine is important. Often treatment with oral agents used regularly is enough (lactulose, senna, Movicol) but glycerine suppositories and micro-enemas can be extremely useful. Incontinence often linked to urgency can be helped with loperamide (DasGupta and Fowler, 2003).

Speech, respiratory and swallowing dysfunction

The speech and language therapist is essential in assessing, advising and treating speech and swallow disorders. Dysarthria may be a feature of conditions causing facial or bulbar muscular weakness; often patients can continue to communicate effectively with friends or relatives who have become accustomed to their speech, however, when very severe the use of communication aids can be explored by the therapist.

Dysphasia often associated with stroke, mass lesions or head injury is usually more problematic to manage. In randomized controlled trials of speech therapy post-stroke it has been shown that language recovery is improved with speech therapy; however, this is related to the intensity of therapy with a mean of 8.8 hours per week of therapy provided in the positive studies (Bhogal et al, 2003).

Swallowing difficulties may be independent of speech problems and can occur in the acute setting following stroke or brain injury or insidiously in chronic conditions such as in multiple sclerosis or mass lesions where the incidence is probably underestimated (Hughes et al, 1994). Many individuals will describe fluctuations in their swallow according to factors such as the time of day, temperature and general fatigue. The speech therapist is invaluable in providing education to the individual and if applicable carers in compensatory mechanisms such as posture (tucking the chin in or turning the head) or change in diet. Education should include the dangers of aspiration with advice on how to identify this. If swallowing becomes severely compromised a percutaneous gastrostomy should be considered; although often dreaded by patients and carers alike, quality of life can often significantly improve.

Respiratory dysfunction, although often a pre-terminal feature, is important to consider, particularly in degenerative conditions such as motor neurone disease or multisystem atrophy. Awareness that this may occur can enable individuals and their families to make decisions and plans before situations such as the need for a tracheostomy or ventilation arise. Non-invasive positive pressure ventilation may be acceptable to individuals not wishing to have invasive ventilation and can have a very positive effect on quality of life, improving daytime somnolence, increasing appetite, relieving morning headache and dyspnoea (Lyall et al, 2001).

Mood and cognitive dysfunction

Assessment of cognitive function by a neuropsychologist following a single insult brain injury or in chronic neurological conditions can be useful in defining the extent of any dysfunction in domains including memory, attention, speed of processing, language and executive function. Cognitive dysfunction can have a devastating impact on psychosocial functioning (Rao et al, 1991), however, identification and assessment of the nature and extent of cognitive deficits can help both the patient and

his/her family or carers instigate effective strategies for orientation and decision making.

Patients with chronic neurological conditions such as multiple sclerosis or Parkinson's disease may also develop mood disorders or other psychiatric symptoms. These are usually mild and commonly include low mood, irritability, poor concentration and anxiety (Ron and Logsdail, 1989). Psychological support or cognitive behavioural therapy is often all that is required, although if medication is indicated it should be used as in the normal population but with greater attention to possible adverse effects, particularly exacerbation of bladder dysfunction.

Conclusions

Although great advances have been made in the management of neurological conditions such as multiple sclerosis and Parkinson's disease, there is still a specific need for effective symptom control at all stages of their condition, particularly as end of life approaches. Much can be done to help the individual and his/her family or carers manage the symptoms from effective and timely education to the input of specialist therapists, pharmacological measures and occasionally more invasive techniques including surgery.

The input of a multidisciplinary team is essential to ensure appropriate input is provided in a coordinated and effective manner while ensuring the individual remains central to the management process and is actively involved in monitoring the impact of symptoms and in the effectiveness of therapeutic interventions. **BJHM**

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KEY POINTS

- Neurological symptoms such as spasticity, neuropathic pain, bladder and bowel dysfunction, speech, respiratory and swallowing difficulties, mood and cognitive dysfunction can impact on not only function but also on passive care and comfort.
- Much can be done to help from effective and timely education to the input of specialist therapists, pharmacological measures and occasionally invasive techniques including surgery.
- The input of a multidisciplinary team is essential to ensure appropriate input is provided in a coordinated and effective manner while ensuring the individual remains central to the management process.
- Effective liaison between neurology and palliative care services is essential to ensure both symptom control and end-of-life issues are appropriately addressed.