Introduction
The Muscular Dystrophy Campaign supports the 60,000 people in England living with more than 60 different types of neuromuscular conditions. These are rare and very rare conditions that can be genetic or acquired, and can present in childhood or adult life.

Neuromuscular conditions can cause muscle weakness or wasting. They are multi-system disorders that require complex long-term care. There are currently no known cures and without multi-disciplinary care, most patients experience a reduction in quality of life and, for some conditions, shortened life expectancy.

Because of the rarity of these conditions and the current patchy provision of services for this patient group, a Strategic Clinical Network could be a vital way to improve services in the new commissioning structure. This briefing outlines the rationale behind designating neuromuscular services as a network and outlines how these services fulfil the criteria for being established as such.

1. A clear link to a national outcome
A Strategic Clinical Network (SCN) for neuromuscular conditions would clearly link to the second domain of the NHS Outcomes Framework: enhancing quality of life for people with long-term conditions. In particular, an SCN would contribute to:

- Ensuring people feel supported to manage their condition
- Improving functional ability in people with long-term conditions
- Reducing time spent in hospital by people with long-term conditions.

These areas will be explored in more detail below.

An SCN would also be vital to take forward the work of the Specialised Commissioning Groups (SCGs) who, over the past year, have been focusing on improving services for patients with a neuromuscular condition across England and developing a service specification for specialised neuromuscular services.

2. The need for co-ordination across complex pathways
Neuromuscular conditions are complex, progressive multi-system conditions which require multidisciplinary pathways of care. These pathways are recommended by the leading clinicians in this field as drastically improving the quality of life and need for emergency care: “Specialist multi-disciplinary care has been developed by leading clinicians as the best model for delivering effective care for such complex, multi-system diseases. The provision of expert physiotherapy, orthotics, early cardiac monitoring and intervention and corticosteroids has been shown to improve muscle function and maintain independent mobility. The judicious use of spinal surgery and expert respiratory services (including non invasive positive pressure ventilation) helps to improve quality of life, delay the onset of respiratory failure and prolong the life of these patients”.  


2 The need for co-ordination across complex pathways for Duchenne muscular dystrophy has been illustrated in the Duchenne standards of care, which were awarded NICE Accredited Evidence status in 2011.

Professor Katharine Bushby et al for the DMD Care Considerations Working Group, The Lancet Neurology - 1 January 2010 ( Vol. 9, Issue 1, Pages 77-93 ) Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management
People with neuromuscular conditions will also need to access vital non-specialised services such as physiotherapy, hydrotherapy and psychological support. It is therefore vital that care is co-ordinated across primary, secondary and tertiary care.

Dr Ros Quinlivan, a leading consultant in neuromuscular disorders, has outlined the effects of neuromuscular conditions and how they need to be managed:

**Neuromuscular conditions are rare and include: muscular dystrophies, metabolic myopathies, congenital myopathies, inflammatory myopathies, spinal muscular atrophies and peripheral neuropathies. Many of these conditions affect only skeletal muscle and thus cannot be considered to be neurological disorders, in fact skeletal muscle can be considered to be the largest organ in the body. Most neuromuscular disorders are genetic in origin and affect families, but the inflammatory myopathies are acquired and require specific treatment. Affected patients range from new-born infants to elderly people. The effect of many of these conditions is on the skeleton causing skeletal deformities due to muscle contractures and on the heart and lungs causing respiratory or cardiac failure which can significantly limit life-expectancy.**

The physical management of these disorders is quite distinct compared with conditions affecting either the nervous system or the musculo-skeletal system (bone and joints). Proximal and axial muscle weakness causes specific functional difficulties not seen in patients attending clinics in other specialist areas. The progressive nature of these conditions means that an anticipatory multi-disciplinary approach to care with experienced clinicians specialising in neuromuscular disorders is essential for best outcomes.

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**The South West Neuromuscular Network**

The work of the South West Neuromuscular Network is overseen by a multi-disciplinary Steering Group to aid co-ordination across complex pathways for patients with a neuromuscular condition.

The Steering Group comprises commissioners, neuromuscular clinicians, clinicians from co-specialities (respiratory and cardiac), allied health professionals, provider trust managers and patient representatives. The group meets regularly to monitor and record progress towards meeting the needs of people with a neuromuscular condition in the South West. When a particular topic or service is under review a working group is set up to engage the key people to address the issue and formulate a plan to move service provision forward. This approach has, for example, been successful in exploring physiotherapy provision in the region. The services within the provider trusts have been challenged and collaboratively reorganised to improve them. For example, the Network is gradually identifying a lead paediatrician in each district general hospital for children with a neuromuscular condition and helping them to pool these children together under their care to provide a neuromuscular clinic, which the neuromuscular care advisor and specialist physiotherapist can attend to deliver better care. The specialist paediatrician can then visit this clinic periodically to provide a specialist service.

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**3. Significant potential for quality improvement through a network model**

Existing neuromuscular networks in the South West and in Scotland demonstrate the value of networks to improve quality of care for patients with a neuromuscular condition.

As outlined in section 4 below, service provision for people with neuromuscular conditions has been patchy and disjointed. The network approach in the South West has fostered a more organised approach to providing for clients across the region, which has enabled the delivery of a more equitable service. The network approach has also raised awareness of the specific needs of these patients, enabling delivery of a better service.

Because the network is able to reach a large patient group across the region, they are able to facilitate a patient and public engagement programme to encourage self management and disease awareness. This empowers individuals to understand their own conditions and push for access to services when necessary.

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Conversely, a provider with a small number of patients will be unable to prioritise such programmes, meaning that individuals will be more vulnerable to deterioration and a health crisis. This vulnerability is demonstrated by data released as part of an audit of hospital admissions being carried out by Professor Michael Hanna (Consultant Neurologist and Director of the Queen Square Division of UCLH NHS Trust). These data, from eight Trusts across London, show that between 37 - 42% of emergency admissions could have been avoided if patients had been able to access the right specialist support.

Recent data from the Lane Fox Respiratory Unit (LFRU) at St Thomas’ hospital has demonstrated that co-ordinated respiratory care for Duchenne muscular dystrophy (DMD), in particular at the vulnerable time of transition from the paediatric to adult service, can reduce the incidence of unplanned admissions, length of stay and the need for invasive ventilation. The 3-year prospective study showed only 16% of the new ventilator set ups were emergency admissions with two patients requiring invasive ventilation. Furthermore, an additional study of prospective data over a 7 year period demonstrated that by providing home cough assist machines, the incidence and length of stay of unplanned admissions for chest sepsis was significantly reduced, which would be reflected as a cost saving to acute healthcare organisations.

Finally, in a recent prospective study, which investigated cause of death over a 6 year period in patients with DMD, it was found that that as a consequence of enhanced respiratory care, respiratory failure accounted only for 35% of deaths in DMD and importantly all of these patients died supported in their home environment. 60% of deaths in this cohort were cardiac failure which was on the whole unpredictable with 90% dying unexpectedly in the acute hospital environment. This model of providing coordinated care from children’s to adult services focuses very much on admission prevention and strengthening community support by maximising the role of the outreach service, and could be best supported and extended across England through a network model.

In addition, a network will be essential to ensuring that there is no fragmentation of the pathway between services commissioned by the National Commissioning Board and those services commissioned at clinical commissioning group level. A network approach would allow for joint planning with other agencies and encourage third sector involvement.

A network would also play a vital role in developing hub and spoke and outreach models whereby patients would receive the majority of their care closer to home whilst remaining, particularly for access to research, under the care of specialist tertiary services.

The role of the neuromuscular care advisor is also key to providing care co-ordination and signposting for all clients with neuromuscular conditions regardless of organisational boundaries. With the establishment of smaller commissioning groups and social enterprises there will more boundary issues, which could potentially cause further fragmentation of service delivery if a network were not established to oversee the provision of this vital service.

Additional evidence of the potential for quality improvement can be seen from the Scottish Muscle Network. In Scotland, the number of boys with Duchenne muscular dystrophy surviving into adulthood is increasing each year. Clinicians agreed that this is largely a result of the work of the Scottish Muscle Network and the improvements this has brought to the standard of paediatric care for this patient group across Scotland.

5 http://www.muscular-dystrophy.org/assets/0000/0454/buildingonFoundations.pdf
Other examples of inequitable care across England also exist. For example, patients in the South West now have access to one of five neuromuscular care advisor posts, whereas patients in the South East Coast only have one care advisor covering a similar population.

5. Clear rationale for why quality improvement cannot be driven by another means (e.g. by a clinical commissioning group)

Neuromuscular conditions are rare and, as such, many GPs and other health professionals may only come across one or two patients in their careers. Because the conditions are complex, multi-system disorders patients will require specialist input from a variety of health professionals and will need to access services commissioned by the National Commissioning Board. However, patients will also need to access non-specialised services, for example some types of physiotherapy, so a network approach, co-ordinated by a specialist lead clinician supported by a group of care advisors, is therefore vital to co-ordinate the patient pathway. Clinical commissioning groups will not have access to a specialist clinical group across a wide enough region to fulfill this co-ordination role.

The South West network provides an example of how a network can benefit patients across the region in a way that clinical commissioning groups could not. They have employed five specialist physiotherapists who work across the region and provide specialist support to all physiotherapists in touch with patients with a neuromuscular condition. This has proved a more effective use of resources than if they were delivering hands-on care and positive patient feedback has demonstrated the success of this approach.

A co-ordinated network approach is also likely to have a wider impact for this group of patients in achieving the key QIPP outcomes and in reducing unplanned emergency admissions.

6. An assessment of how the absence of an SCN would result in a lack of continuous quality improvement

The SCGs have been focusing on improving services for patients with a neuromuscular condition over the past twelve months. This work has required the SCGs to work collaboratively across local and regional boundaries to improve current services and to develop a national service specification to ensure the national gaps in care can be consistently addressed. In order to make sure that this work is taken forward and the service specification is implemented, it is vital that there is a focus on co-ordinating care and developing clear patient pathways. This can best be achieved through designating an SCN for neuromuscular conditions.

7. Commissioning by the National Commissioning Board

There are more than 60 different types of muscular dystrophy and related neuromuscular conditions, many of which are low incidence, orphan conditions and indeed some are very rare and are regarded as ultra orphan. Neuromuscular conditions can be genetic or acquired and, with the exception of a couple of acquired conditions, there are no known effective treatments or cures.

As these are rare and very rare conditions, they have been the responsibility of the 10 regional Specialised Commissioning Groups in England, and are listed in the Specialised Services National Definition Set (SSNDS). The ultra-rare conditions (affecting less than 400 patients each) are the responsibility of AGNSS (Advisory Group for National Specialised Services) and commissioning is expected to transfer to the National Commissioning Board from April 2013.

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8 National Specialised Commissioning Group Specialised Services National Definition Set http://www.specialisedservices.nhs.uk/info/specialised-services-national-definitions
9 AGNSS: http://www.specialisedservices.nhs.uk/info/agnss
This briefing has been endorsed by the British Myology Society and the following specialist clinicians:

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