

The State of the Nation:

# National Patient Survey

2013



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# Foreword

The latest national patient survey by the Muscular Dystrophy Campaign has revealed major gaps in the provision and quality of care and support for people with muscular dystrophy and related neuromuscular conditions. We found that too many patients are still struggling to access the specialist, multi-disciplinary care and support they need to manage their conditions and improve their quality of life. Particularly concerning is patients reporting a lack of regular heart and lung checks, which are absolutely vital for many people with neuromuscular conditions. Some people with these conditions are at high risk of death without regular monitoring of heart and lung functions by a specialist consultant. Other gaps include transition services between child and adult care across England, with few units to support this patient group. Our survey also revealed that too many vulnerable patients are lacking support from specialist physiotherapists, as well as from psychological services; these are gaps in care which must urgently be set right.

The forthcoming changes taking place within the NHS and in particular new guidelines on neuromuscular services<sup>i</sup>, set to be launched by the NHS Commissioning Board in April 2013, provide a real opportunity for the NHS to improve services for this vulnerable patient group. The Muscular Dystrophy Campaign will continue to push forward and highlight the continuing need for essential specialist support for people with neuromuscular conditions.

Since our last State of the Nation<sup>ii</sup> report in 2010, new research has revealed that millions of pounds are being wasted through people being admitted to hospitals as an emergency. An audit of hospital admissions,<sup>iii</sup> led by Professor Mike Hanna, showed that around 40 percent of emergency admissions could have been avoided if patients had been able to access the specialist support they require. Access to specialist teams, emergency plans, specialist physiotherapy and vital specialist equipment such as orthotics and cough assist machines were all highlighted as important factors that would prevent emergency admissions. While inadequate provision of vital specialist services persists, the NHS continues to waste money. The Muscular Dystrophy Campaign calls on the NHS and commissioners to invest more in vital specialist support so that more patients can avoid costly emergency admissions to hospital.

The Muscular Dystrophy Campaign also has major concerns about delays to investment in services and the potential of further cutbacks. We urgently call on NHS Trusts and Clinical Commissioning Groups to put in place plans and to set specific targets to ensure that neuromuscular services are brought up to the level set out in the new guidelines, so that people with neuromuscular conditions have access to the vital treatment, care and support they need.

This report focuses on alarming experiences at diagnosis, shocking gaps in access to specialist care and inadequate support from GPs in England, with further reports covering Scotland, Wales and Northern Ireland set to be published later this year.

A handwritten signature in orange ink, reading 'Robert Meadowcroft'.

Robert Meadowcroft  
Chief Executive  
The Muscular Dystrophy Campaign

# 1. Foreword

## Neuromuscular conditions and the Muscular Dystrophy Campaign

The Muscular Dystrophy Campaign supports the 70,000<sup>iv</sup> people across the UK affected by 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 or treatments.

Patients require vital multi-disciplinary care from specialist neuromuscular consultants, physiotherapists, nurses, care advisors and – where necessary – support from specialist respiratory and cardiac consultants. Without this level of care, patients experience a reduction in quality of life and, in some cases, shortened life-expectancy.

## NHS reforms: what is going to change?

The Muscular Dystrophy Campaign works closely with the ten regional NHS Specialised Commissioning Groups, responsible for ensuring that specialist care services for people with neuromuscular conditions are provided effectively. However, from April 2013, when the new NHS structures come into force, these Specialised Commissioning Groups will no longer exist.

As the NHS regional set-up changes, responsibility for the specialised services which people with neuromuscular conditions receive will be shifted to 10 local area teams, overseen by the NHS Commissioning Board, with community care being provided at an even more local level.

In terms of how this will affect people with neuromuscular conditions, it is hoped that the publication of the neuromuscular service specification will mean that the specialist multi-disciplinary care required will be improved by these changes.

## About the Muscular Dystrophy Campaign

**The Muscular Dystrophy Campaign is the leading UK charity fighting muscle-wasting conditions. We are dedicated to beating muscular dystrophy and related neuromuscular conditions by finding treatments and cures and to improving the lives of everyone affected by them.**

Our work has five main focuses:

- we fund world-class research to find effective treatments and cures we provide practical information, advice and emotional support for individuals with muscle-wasting conditions, their carers and families
- we campaign to bring about change and raise awareness of muscular dystrophy and related neuromuscular conditions
- we award grants towards the cost of specialist equipment, such as powered wheelchairs
- we provide specialist education and development for health professionals.



## 2. Executive summary

Our nationwide survey of people living with muscle-wasting conditions has revealed a continuation of the 'postcode lottery' of available care and support. Almost 600 patients and families responded to the survey.

### Key findings

- Some adults and children at high risk of death from heart and lung problems are not receiving regular checks – one patient had even been told, “go home, we can do nothing for you” by senior medical staff.
- One in five patients said their GP had offered inaccurate advice in the past year, and nearly half of patients surveyed said their GP did not understand their condition well enough to plan local care.
- More than one-third of patients surveyed had their condition misdiagnosed, with almost one in five patients waiting five years or longer for an accurate diagnosis.
- More than a quarter of respondents were forced to wait longer than a year to see a specialist consultant, and some were forced to wait several years.

### The Muscular Dystrophy Campaign is calling for:

- the NHS to ensure a faster diagnosis for both adults and children with symptoms of a muscle-wasting condition, with families supported by specialist, multi-disciplinary teams of health professionals within their local area
- **clinicians and the NHS to ensure regular heart and lung checks for patients with respiratory and cardiac problems**
- GP-led Clinical Commissioning Groups to work with specialist neuromuscular clinicians, regional NHS leaders and people affected by muscle-wasting conditions to ensure that (a) the warning signs are picked up sooner and (b) appropriate care and support arrangements are put in place locally
- the NHS to recognise the value of investment in specialist care, which will not only dramatically reduce unplanned admissions but it would also save money by keeping people out of hospital
- new guidelines for the treatment and care of people with neuromuscular conditions to be implemented as soon as possible.

### GP-led commissioning

People with muscle-wasting conditions experience a significant lack of understanding of their conditions by GPs, which re-affirms long-standing concerns about GP-led commissioning for community-based care services. Moving to more locally based commissioning, with GP-led Clinical Commissioning Groups (CCGs), is of concern to many people with the condition.

Neuromuscular expertise and knowledge will be needed within the new NHS structures to address these alarming gaps and commissioners must have access to the expert knowledge needed to ensure that the necessary specialist

services are available. The Muscular Dystrophy Campaign has recently set up a number of patient-led expert panels, which act as a forum for clinicians, patient representatives and Clinical Commissioning Groups to work together and to make sure there is a co-ordinated approach to patient care pathways. The panels, led by people with muscle-wasting conditions, empower neuromuscular patients to take the lead on working with commissioners to improve services in their area. The Muscular Dystrophy Campaign hopes to ensure the new NHS structure can meet the complex and specialised needs of the 60,000 people with muscle-wasting conditions across England.

### 3. Support from GPs

#### What the survey reveals:

- nearly half of all patients surveyed felt that their GP did not understand their condition well enough to put in place the right support for them
- two out of five patients felt that their GP did not understand their condition sufficiently to refer them to the appropriate specialists
- fewer than half of those surveyed felt that the advice their GP had offered was correct, while a third were uncertain whether the advice was correct or not
- close to half of the people surveyed said they felt their GP did not understand their condition well enough to plan the primary care services they needed locally, which raises concerns about the planned shift to GP-led commissioning in April 2013

#### What people have said:

- Justine McAlister from East Grinstead has a 6-year-old son, Benedict, who has congenital muscular dystrophy. The time leading up to his diagnosis was an extremely tortuous process for the family. After being told by two GPs that Benedict was a healthy young boy and the family had nothing to worry about, they decided to consult a private GP, who misdiagnosed Benedict with hyper-mobility. This delay in diagnosis has meant that Benedict has not received the vital care and support needed to manage his condition.

- Jeannette George, 68, from Colchester has limb girdle muscular dystrophy. She has had a long struggle to get the right care and support, and said:

*"I feel isolated and misunderstood by my local surgery. I have been left very much to fend for myself and push for the help I need."*

- Maxine, 40, from Lancashire, has facioscapulohumeral muscular dystrophy and has felt let down by her local GP, saying:

*"I have repeatedly attended my GP surgery for chronic pain, tiredness and sleep problems and have been made to feel like I am wasting their time."*

- Bruno Carrico, 16, from the South West has Charcot-Marie-Tooth disease (CMT) and felt let down by the care given by his GP. Bruno said:

*"My GP appears to have no knowledge of my condition and often asks me what is wrong with me. He does not appear to read any notes or letters sent to him and he even asked me what CMT stood for."*

- Sylvia, 78, from Worthing, has facioscapulohumeral muscular dystrophy and has been confronted by a lack of knowledge of her condition by GPs. She has therefore never seen a specialist about her condition, which means her condition has deteriorated in a way that may have been delayed with adequate specialist support.

*"Over the years, I have flagged up my condition to GPs, and their response is just that they have heard of the condition but know little about it. My walking has deteriorated and I have major concerns about my future and the support I will need."*

- Pauline Clark from Hereford has a 14-year-old son, Josh, who has Duchenne muscular dystrophy. When Josh was diagnosed with this severe and devastating condition, Pauline felt completely let down by the dismissive attitude of her GP. She said:

*"I was essentially told, 'Sorry, he has Duchenne, any questions? See you in three months.' There was not even a nurse to chat with, no tea or coffee, no time given to get myself back together, I just left with the news and leaflets."*

Stephen Meech is a GP and his son has Duchenne muscular dystrophy. Stephen has raised concerns about the move to GP-led commissioning and highlighted the urgent need for improved GP understanding of neuromuscular conditions, saying:

*"GPs work with children every day. It is important that GPs have openness and willingness to listen to parents' concerns and timely access to specialist support, if required, when a child presents with unusual symptoms."*

*"Although the new NHS Clinical Commissioning Groups are involving GPs more closely with local commissioning arrangements, they are focusing on commonly occurring conditions such as diabetes, heart and respiratory problems. Few GPs have in-depth knowledge of more than a smattering of rare conditions, which therefore risk being overlooked, and I am keen to see specialist and regional services for these commissioned more centrally."*

### 3. Support from GPs

#### Recommendations

The survey shows that a lack of knowledge about neuromuscular conditions by many GPs is a major problem for patients. This has led, in some instances, to patients receiving inaccurate advice and, in some cases, contributed to a delayed diagnosis.

- **From April 2013, people with neuromuscular conditions should have their own patient-held records which will be updated by those involved in the patient's care. These can be shared with other professionals who may not be familiar with the patient's condition or care needs, for example should a patient be admitted to A&E. Accurate patient-held records are absolutely vital to ensure people get the appropriate specialist support. While it is hoped these records will improve GPs' knowledge and understanding of conditions, we are keen that GPs work with the Muscular Dystrophy Campaign to develop their understanding of neuromuscular conditions.**
- While we recognise the difficulties faced by GPs when confronted with rare conditions, the Muscular Dystrophy Campaign is calling for improved training for GPs and for GPs actively to engage with patients, specialists and commissioners in order (a) to have a better understanding of the symptoms associated with rare conditions, and (b) to start to plan local services effectively.
- Specialist knowledge and expertise needs to be shared in an effective and co-ordinated way by the new NHS Local Area Teams (LATs) to ensure that GPs can provide the appropriate level of care and support, locally, for people with muscle-wasting conditions.

## 4. Information and support at diagnosis

### What the survey reveals:

- people have continued to experience long waits for a correct diagnosis with one out of six having to wait more than five years for their diagnosis, and fewer than half of people receiving a diagnosis in under a year
- of those who did not receive a correct and prompt diagnosis, 29 cases were reported where, whilst waiting for a correct diagnosis, parents had other children who have now being diagnosed with a neuromuscular condition
- more than one-third of patients had their conditions misdiagnosed
- around three out of five patients felt they were not well supported at the time of diagnosis.

### What people have said:

- Annette, from Petersfield, has a 10-year-old son who has Duchenne muscular dystrophy. After her son's creatine kinase (CK) levels were identified as being raised, he had a biopsy to identify whether or not he had muscular dystrophy. After a long wait for the results – they were lost in the post – the family felt completely let down by the support they received at diagnosis. Annette said:

*"When we were eventually told, the doctor said, 'Your son has Duchenne, here is a leaflet on steroids, enjoy what time he has left,' and sent us on our way."*

- Emma, from London, has a 16-year-old son who has facioscapulohumeral muscular dystrophy (FSH), and he was not diagnosed until he was 14 years old, despite his condition being at an advanced stage. Talking about her experience, Emma said:

*"Health professionals were heading down the wrong track with his condition, thinking that he had problems with his diet as he had had a dramatic weight loss. It wasn't the case of us not engaging with health professionals; my son was passed around from pillar to post. When we finally saw a neurologist at Great Ormond Street, he was shocked that paediatricians hadn't picked up on the vital signs – his was such an advanced case."*

### Recommendations

Linked to GPs' lack of adequate understanding of neuromuscular conditions is a concerning pattern of delayed and incorrect diagnoses of these conditions. Many patients are waiting too long; diagnoses should be confirmed in a matter of weeks rather than months.

Swift and accurate diagnosis is absolutely essential for people with neuromuscular conditions. The time between an initial indication that a patient might have a neuromuscular condition, such as raised levels of creatine kinase (CK) in the blood, and a formal diagnosis, based on a muscle biopsy, must be drastically reduced. This is especially important for young boys with Duchenne muscular dystrophy, where rapid progression of the condition means that the clinical management of the condition – often using steroids – has to start as soon as possible.

- **According to the new standards set out in the service specification, there must be appropriate referral practices in place to avoid delays in diagnosis. Clearly, neuromuscular patients are not experiencing fast and effective diagnosis and action needs to be taken to ensure this is addressed to avoid lengthy and distressing delays experienced by individuals and families.**
- There need to be shorter lead times between initial suspicions and confirmed diagnoses. The Muscular Dystrophy Campaign would want this lead time to be no longer than 18 weeks for both children and adults.
- Training and support must be given to GPs so that the early warning signs of muscle-wasting conditions can be recognised, a faster and more effective diagnosis can be made and appropriate support given to patients and their families.

#### Dr Amy-Jayne McKnight, a geneticist whose father has a neuromuscular condition, says:

*"Obtaining an accurate diagnosis as early as possible is crucial to enable individuals with muscular dystrophy to receive optimal treatment. Every day counts, when families are waiting for a diagnosis and we know that early interventions – such as corticosteroids to prolong ambulation for boys with Duchenne muscular dystrophy, and respiratory/cardiac/support therapies – substantially improve the quality and life-expectancy of affected individuals."*

*"As part of a family living with muscular dystrophy, and as a geneticist, I would like to see the NHS commit to a timeframe of a maximum of 18 weeks between the time concerns about symptoms are first raised to an accurate diagnosis being delivered. As many forms of muscular dystrophy are rare, it is particularly important that GPs and community health professionals are provided with appropriate information and a contact point to help manage each patient's condition and maximise their lifestyle."*



## 5. Specialist healthcare

### What the survey reveals:

- some adults and children at high risk of death from heart and lung problems are not receiving regular checks
- more than a quarter of respondents had to wait longer than 12 months to see a specialist consultant after they had been diagnosed with a neuromuscular condition
- a quarter of patients surveyed have never seen a neuromuscular consultant
- of those seeing a consultant, one quarter of patients see their consultant fewer times than once a year
- one-third of respondents reported that they did not have access to the support of a neuromuscular care advisor in their region
- around half the patients surveyed had not accessed any specialist services, such as specialist physiotherapists
- over the past two years, two-thirds of respondents had not received any physiotherapy, more than five out of six respondents had not received hydrotherapy and a similar proportion had not received any other specialised treatments.

### What people have said:

- one mum from Birmingham, whose son has congenital muscular dystrophy, told us how a lack of specialist knowledge of his condition meant that doctors decided they would not resuscitate on his behalf. Speaking about this traumatic experience, she said:

*"They didn't know anything about my son's condition, so they just put him into a category and made the decision not to resuscitate on his behalf. They just thought he would die, but it didn't work like that and it can't."*

- a 53-year-old woman from Buckinghamshire, who has facioscapulohumeral muscular dystrophy (FSH) – a muscle-wasting condition which causes the muscles in the face, arms and legs to waste, a condition which her mother and brother also have – said:

*"In both my mother and my brother's cases, their hearts have been affected. My mother has [recently] suffered a heart attack and stroke and my brother has developed heart problems. Even now, despite being at risk from further complications, neither of them is being monitored. My brother has had to retire, because he can't cope. After years of inequalities in care and support – it is about time that the NHS put words into action."*

- Emma, from London, has a 16-year-old son who has facioscapulohumeral muscular dystrophy (FSH) and she has struggled to get an appointment with a specialist, despite her son being at high risk of choking. Emma is concerned about the effectiveness of the planned changes to the NHS services.

*"Recently my son's school has flagged up issues with his swallowing and the high risk of choking. Despite his school highlighting this last summer, Louis' appointment keeps on being postponed. The current NHS system is a maze and I hope patients and families will see the planned improvements to the NHS on the ground sooner rather than later."*

- Mr Brown, from Bolton-le-Sands, has facioscapulohumeral muscular dystrophy (FSH) and has not seen a consultant for 50 years. Even with the intervention of the Muscular Dystrophy Campaign's advocacy\* service, it was a struggle for Mike to see a consultant.

*"When I was finally referred to a consultant, he refused to see me again as he said there were people worse off than me, despite the fact I had 40 percent lung capacity. I have given up any hope of getting help, my experience of local health care has been a shambles."*

- Rachel, 40, from Chichester has limb girdle muscular dystrophy. She has experienced patchy support from her consultant, saying:

*"After suffering from increasing pain in my legs, I was told that I would be booked in for five days of intensive therapy to work on my transfers and going up stairs, and I was terrified. I am a wheelchair user and have been for years, I physically cannot walk up stairs. There was a complete lack of understanding on the consultant's behalf. I am supposed to have annual appointments with my neurologist but he has cancelled three appointments this last year and the last time saw him was in January 2011."*

\* The Muscular Dystrophy Campaign's advocacy service helps people affected by muscle-wasting conditions to get the assistance they require to live a full and independent life. People approach the charity's service to help them fight for the services, benefits and specialist equipment they are entitled to.

## 5. Specialist healthcare

### Recommendations

- **New standards set out by the NHS recommend that patients will be reviewed at least every six months by a specialist multi-disciplinary team. However, our survey shows that one quarter of respondents do not see a specialist consultant more than once a year and two-thirds have not received support from a specialist physiotherapist in the past two years. This demonstrates that specialist support has been falling short of this benchmark.**
- Adults with severe muscle-wasting conditions, who have associated respiratory and cardiac issues, should have their heart and lungs checked at least once a year at a muscle centre or a clinic that has appropriate expertise from a specialist multi-disciplinary team.
- All adults and children with neuromuscular conditions should have access to support from a neuromuscular care advisor and the Muscular Dystrophy Campaign would like to see at least 70 such posts nationally, giving each one a caseload of about 1,000 people.
- While there are no cures for neuromuscular conditions, advice on managing health, physiotherapy and access to respiratory equipment to ward off dangerous chest infections can all protect length and quality of life. The NHS must invest in specialist, multi-disciplinary teams, increasing specialist physiotherapy provision and improving the transition from child to adult services to avoid costly unplanned emergency admissions.

### Progress since the last survey in August 2010

This patient report coincides with the NHS' publication of a detailed service specification for neuromuscular services. This document, set by the NHS Commissioning Board, sets out guidelines for healthcare service providers in England, outlining in detail the support someone with a rare neuromuscular condition should have access to. The service specification has just completed its consultation stage, with changes set to take place from April 2013. The guidelines pay particular attention to the multi-disciplinary and cross-organisational working, which is so essential for ensuring that patients, their families and carers, receive the most effective and efficient care. This offers a real opportunity to develop a more equitable, effective and ultimately a better service for people with neuromuscular conditions. With the excellent support of our campaigners across the country, the Muscular Dystrophy Campaign has fought hard to ensure that the planned improvements to the NHS make neuromuscular services a priority. The publication

of the service specification for consultation is also a pivotal moment in the charity's campaign to improve access to specialised neuromuscular care, extending life-expectancy and increasing quality of life.

#### Service specification: what does this mean for people with neuromuscular conditions?

The service specification is a document produced by the NHS Commissioning Board, the new England-wide body responsible for ensuring that care services for people with neuromuscular conditions are provided effectively.

The document outlines the objectives for Specialised Neuromuscular Services across England from April 2013, and provides detailed guidelines on the provision of standardised care, dietary and nutritional care, specialist equipment and transition as well as respiratory care and psychological support. The specification clearly states that: 'patients will be reviewed at least every six months by the specialist multi-disciplinary team.'

This is a vital development towards an integrated and consistent approach to the provision of specialist multi-disciplinary neuromuscular care across England, which has been a key campaigning priority for the Muscular Dystrophy Campaign.

Over the past two years, significant progress has been made and it is crucial that we use the NHS reforms as an opportunity to build on this progress to ensure that a specialised, national neuromuscular service is cemented.

The NHS now funds 32 specialist neuromuscular care advisors across the UK, up from 10 in 2004, who provide vital information and support to families affected by muscular dystrophy and related neuromuscular conditions. There are still significant gaps in support from neuromuscular care advisors and the Muscular Dystrophy Campaign would like to see at least 70 in post nationally, giving each one a caseload of about 1,000 people.

The 10 regional NHS Specialised Commissioning Groups across England have undertaken reviews of neuromuscular services in their areas, including a series of recommendations on how services will be developed and improved. However, the Muscular Dystrophy Campaign is alarmed that many of these vital recommendations have not been implemented and believes these recommendations must be put in place as a priority, alongside continued investment in specialised neuromuscular services.

## 6. Action needed

The Muscular Dystrophy Campaign is calling on the NHS to ensure that services meet the needs of patients with neuromuscular conditions at all stages of the care pathway from initial diagnosis to the ongoing management of the condition. The service specification goes some way to achieving this, but it is essential that the NHS is held to account if the standards in the specification are not met.

The Muscular Dystrophy Campaign's 2011 Invest to Save <sup>v</sup> report clearly shows that better use of existing resources can fund improvements in the quality of neuromuscular services. Figures show that investment in preventative services could result in savings of approximately £25 million in England and approximately £31 million across the whole of the UK, with a relatively small investment of £4.6 million.

### To improve patient services new NHS commissioners need:

- to ensure that the service delivered to people with muscular dystrophy and related neuromuscular conditions meets the standard outlined in the service specification and reduces the inequalities in provision of care which currently exist
- to ensure that people with muscular dystrophy or related neuromuscular conditions are supported by a specialist, multi-disciplinary team of health professionals who are accessible within their local region
- to use the South West Neuromuscular Network as a model for creating neuromuscular networks across other regions in England
- to ensure that people with muscular dystrophy or related neuromuscular conditions have access to the support of a specialist neuromuscular care advisor to support them and their carers, both practically and emotionally
- to recognise that investment in specialist care can dramatically reduce unplanned admissions and save the NHS money by keeping people out of hospital
- to encourage the NHS Commissioning Board to take into account the high cost of unplanned emergency admissions and develop a NICE Quality Standard on Duchenne<sup>6</sup> as well as other neuromuscular conditions.

### In addition, Commissioners must:

- give full commitment to developing and working with Neuromuscular Steering Groups – composed of clinicians, therapists, NHS commissioning staff and patients – which are a key part of monitoring and developing the services available to patients with a neuromuscular condition in a particular region
- use the South West Neuromuscular Network <sup>vii</sup> as a model for future service development in other areas. The South West has led the way for the country in developing its services and has secured four neuromuscular care advisors

and a network co-ordinator to manage the care of patients and work to bring together services

- ensure that specialist Muscle Centres, such as those in Newcastle and London, have the resources to provide essential outreach services in the community.

### The Government must:

- ensure that the NHS National Commissioning Board has the power to enforce the contractual requirements outlined in the service specification for neuromuscular services
- encourage the training and education of health professionals and professional bodies on neuromuscular conditions ensure the national neuromuscular lead, who has played a vital role in coordinating the neuromuscular plans in the new NHS set-up, continues in post after April 2013.

#### Developing neuromuscular networks

A neuromuscular network could be a vital way to improve services within the new commissioning structure, given the rarity of neuromuscular conditions and the current patchy provision of services for this patient group.

It has been recommended that neuromuscular services delivered through a network model would be 'good candidates' for new 'Operational Delivery Networks' which the NHS is developing across England to improve the way services are delivered.

The work of the South West Neuromuscular Network has demonstrated the improvements that can be made to specialist care if services are linked together in an effective and efficient way. The Network is overseen by a group of NHS and patient representatives and health professionals to help co-ordinate service provision for patients with neuromuscular conditions.

## 7. Reducing the cost of unplanned emergency admissions on the NHS

Unplanned emergency admissions have been identified as a significant problem nationally for patients with neuromuscular conditions. Unplanned admissions to hospital, for example as a result of a fall or a respiratory infection, negatively impact on the quality and experience of care received by people with neuromuscular conditions and are harmful to patients' health. In addition, emergency admissions often lead to a significant increase in length of hospital stay and therefore unnecessary costs to the NHS.

An audit published in 2012 of unplanned emergency admissions of 395 neuromuscular patients, led by Professor Michael Hanna, Consultant Neurologist and Director of the MRC Centre for Neuromuscular Diseases, showed that 37 to 42 percent of emergency admissions could have been avoided if patients had been able to access the right specialist support (Hanna et al 2012: 9). As well as access to specialist teams and emergency plans, the report highlighted three other important investments that could be made in order to prevent emergency admissions:

1. specialist physiotherapy
2. vital specialist equipment such as orthotics, and
3. cough assist machines.

Too many clinics are falling short of this, resulting in a high cost of preventable, unplanned admissions. The Muscular Dystrophy Campaign urges clinics to follow the recommendations made in Professor Hanna's report in order to bring down the number of preventable admissions.

### Unplanned admissions of neuromuscular patients: recommendations

1. Monitoring of known neuromuscular patients and access to neuromuscular services between clinic appointments should be strengthened. This could be co-ordinated in a more formal process by the service, for example by the clinical nurse specialist.
2. The specialist neuromuscular centre should co-ordinate care across different sub-specialisations (neuromuscular, cardiac and respiratory). Fragmentation of care across different hospitals should be avoided, where possible, to ensure good communication, avoid conflicting advice and provide an integrated care pathway.
3. All patients with a known neuromuscular diagnosis should have a documented emergency plan which specifies a clear point of access for emergency care. This may include telephone access for the patient to the specialist neuromuscular centre during times of worsening health.
4. Specialist neuromuscular centres should develop links, preferably with outreach, with local hospitals so that advice, diagnosis and referral can be managed in a timely fashion. Links should also be improved with local social services to ensure a patient's ongoing needs can be met and to prevent delays in being discharged owing to social care issues.
5. Specialist neuromuscular centres and commissioners should consider together whether other models of care or network arrangements would be an appropriate way to co-ordinate care for these patients.
6. Consideration should be given to undertaking further study of unplanned or emergency admissions (outside of London and outside of specialist neuromuscular centres) to try and gain an understanding of the broader neuromuscular population.
7. All patients with a known neuromuscular condition should have a:
  - a documented referral to the neurology team, even if the neuromuscular condition is not the prime reason for admission
  - b emergency plan on discharge.Health professionals should ensure that there is clear documentation of any review of a patient.

Hanna et al (2012), p.29

## 8. Improving provision of neuromuscular care advisors

Neuromuscular care advisors, who are based at NHS neuromuscular clinics in various regions throughout the UK, are key members of a neuromuscular specialist department and are responsible for specialist support and advice for patients diagnosed with a neuromuscular condition. There are currently 32 care advisors across the UK, but the Muscular Dystrophy Campaign would like to increase this number to at least 70 nationally, giving each one a caseload of about 1,000 people.

Neuromuscular care advisors provide practical and emotional support for people with muscle-wasting conditions, as well as their families. Their training backgrounds include nursing, social care, physiotherapy and occupational therapy and they work with statutory service providers, education authorities and other professionals to advise, support and best meet the needs of people with muscular dystrophy and related neuromuscular conditions.

They have a key role in assessing the patient's and family's needs and making necessary interventions. These could be telephone advice, single visits, short-term interventions or regular assessments and reviews of ongoing complex issues. They support families and patients with neuromuscular

conditions through signposting, liaising with local support teams, and developing care plans.

A neuromuscular care advisor also follows up with newly-diagnosed patients, for example through a home visit, to ensure families are given timely support. They organise and provide support and resources, helping patients and their families to adjust and adapt to a changed lifestyle in order to lead the fullest lives possible.

They also have a key role in establishing local links and networks. A neuromuscular care advisor is the primary co-ordinator between GPs, health providers and medical consultants to ensure systems are in place to provide the most appropriate support for the patient and their families.

### The role of a neuromuscular care advisor

Carol Wood was appointed as a neuromuscular care advisor in the East of England in November 2011 and is based in Stansted working with children and adults in Cambridgeshire and Essex. Carol says that each family has multiple issues that require help.

"The kind of information most of the individuals and families we work with need to know is generally very variable, sometimes dependent on the particular condition. Often what people need is information about their condition, or their entitlements (benefits, housing, health care, equipment etc) or how to navigate statutory bodies."

"The most satisfying part of my job is in making a difference and improving the quality of life for the patient and their family. The best feedback to get from patients is the knowledge that they feel supported and able to manage the problems that previously felt overwhelming to them," said Andrea Russell, the East of England neuromuscular care advisor covering Norfolk and Suffolk.

One person who has benefited enormously from the support of neuromuscular care advisors is Farrell Fox, 56, from Weston-Super-Mare in North Somerset. Farrell has muscular dystrophy and describes the work of his neuromuscular care advisor as a 'joyful necessity', and the help as a 'total revelation'.

"When the consultant first told me of my muscular dystrophy, she then said this is Carla, the neuromuscular advisor who will help you 'manage' your condition. Within two days, she had visited me at home and spent a lot of time assessing me. Within a week, I had visits from the Falls Prevention Service and two different occupational therapists. Shortly afterwards there were visits from physiotherapists and speech and language therapists. I had fire inspections, dieticians and the care agency 'Care and Repair'. Amazing numbers of adaptations, gadgets and equipment turned up and I was made a case study of how a person with neuromuscular disease could be helped to remain in their own home. Carla even came back and filled out forms for me and was always available by phone or email.

Before this, I was really depressed and had convinced myself that this was the last year in my upstairs flat, my home for over a quarter of a century. Now, a weight has been lifted and I look to the future with total optimism. Most of all, I am safe and secure in the knowledge that as my condition deteriorates, your (South West Neuromuscular) Network is able to help with a whole range of advice, adaptations, gadgets and support. This experience has been a total revelation and I would like to express my gratitude to the Network and Carla."

Farrell Fox, Weston-Super-Mare



## 9. Improving care through specialist centres

Research into Duchenne muscular dystrophy is accelerating and the prospect of potential treatments is on the horizon. It is therefore more important than ever before to ensure that the clinical infrastructure is put in place now to ensure that potential treatments can be delivered to patients.

**The Muscular Dystrophy Campaign is calling on NHS commissioners to work with clinicians and Hospital Trusts to ensure that existing Muscle Centres and clinics are able to provide:**

- two or more consultant physicians with a special interest in neuromuscular disease (for a combined adult and paediatric centre, this should include two adult and two paediatric consultants)
- access to medical specialists (ideally designated consultants) in orthopaedics, cardiology, clinical neurophysiology, gastroenterology, genetics, neuropathology, respiratory medicine and, for paediatric centres, endocrinology and spinal surgery. Joint clinics between the lead consultant and any of these specialists or appointments on the same day as the neuromuscular clinic are preferred but not essential
- at least fortnightly specialist clinics for adults or children with muscular dystrophy and related neuromuscular conditions
- a regular clinic attended by both an adult and paediatric neuromuscular consultant physician and one other designated professional to provide a transition service
- access to relevant para-clinical staff experienced in neuromuscular conditions, including a physiotherapist, occupational therapist, clinical psychologist, neuromuscular care advisor/support worker, dietician, speech and language therapist, genetic counsellor and orthotist
- signposting to relevant patient support groups
- support to fellow professionals in any future operational network, i.e. local hospital and community teams
- resource, training opportunities for health and social care professionals including continuing professional development and links (possibly through forums and networks), as well as regular dialogue with the surrounding local community services
- collaboration with high quality multi-centre clinical, scientific and social research
- contribution to the maintenance of national and local databases of patients attending the centre
- monitoring patient satisfaction including physical access, interactions with all staff, liaison between clinics and services, especially cardiology, spinal surgery, and respiratory support
- succession planning for all aspects of their service.

Muscular Dystrophy Campaign Services Development Committee (2012) <sup>viii</sup>

# 10. Conclusion

To conclude, the State of the Nation report shows:

- the NHS reforms are a unique opportunity to ensure the current postcode lottery of care is ended and people with neuromuscular conditions and their families can access a specialist multi-disciplinary team in their region
  - the NHS can save money and improve patient care by investing in specialised support
  - the recommendations given by Professor Hanna provide practical ways in which commissioners can reduce the number of preventable hospital admissions
  - the NHS Commissioning Board needs to work with Local Area Teams and Clinical Commissioning Groups so that all elements of patient care are looked after – from the highly specialised end to the care provided closer to home
  - One way to do this is through rolling out patient-led panels to ensure engagement between patients, commissioners and clinicians is taking place
  - GPs should work with neuromuscular clinicians, regional NHS leaders and people affected by neuromuscular conditions so that the early warning signs of conditions are picked up sooner and appropriate care and support arrangements are put in place locally.
- To join the fight against muscle-wasting conditions and to find out how you can support the recommendations made in the report, then please email [campaigns@muscular-dystrophy.org](mailto:campaigns@muscular-dystrophy.org) or call 0207 803 2865.**

## Glossary of conditions

### Charcot-Marie-Tooth disease

A condition that affects the nerves which carry signals to and from the muscles in the legs and arms. These nerves are known as the peripheral nerves. It is characterised by progressive weakness and wasting of the muscles in the calves, lower arms, hands and feet. There may also be loss of sensation in the hands and feet.

### Congenital myotonic dystrophy

Congenital myotonic dystrophy is the early childhood form of myotonic dystrophy. It occurs only when the mother already has myotonic dystrophy. The condition is usually identified at birth or soon after; myotonic means “involving muscle stiffness”; and dystrophy, “muscle wasting and weakness”. This condition is rare.

### Duchenne muscular dystrophy

A genetic disorder in which muscle cells break down, and are eventually lost, causing progressive muscle weakness. The condition usually affects only boys and is caused by a lack of dystrophin protein.

### Facioscapulohumeral muscular dystrophy (FSH)

A muscle wasting condition, caused by a genetic defect. The name describes the distribution of weakened muscles: ‘facio’ - facial; ‘scapulo’ - shoulder blade; ‘humeral’ - upper arm.

### Limb girdle muscular dystrophy (LGMD)

A group of conditions affecting primarily the limb girdle muscles (i.e. the muscles around the shoulders and hips). There are at least 19 different forms of LGMD and the genes involved in 15 of these have been identified. The symptoms of the condition appear differently in different people, with regard to age of onset, areas of muscle weakness, heart and respiratory involvement, rate of progression and severity.

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