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Foreword

"They were rehabilitation exercises that you'd use if you were injured playing sport. They were very aggressive and weren't designed for long-term conditions."



"I was born with merosin-deficient congenital muscular dystrophy. The doctors knew I had some sort of congenital muscular dystrophy from the time I was really young, but I wasn't properly diagnosed until I was nine or ten. There's really limited access to the therapies that could help maintain my muscle function.

"I had a physiotherapy appointment over a year ago, and it was horrible. The physio came to my house, walked right in and said, 'You need to do this, this and this,' without really knowing what he was talking about. He showed my carers stretching exercises that were completely inappropriate for my condition. He didn't understand that my legs don't bend that way. They were rehabilitation exercises that you'd use if you were injured, say, playing football. They were very aggressive and weren't designed for long-term conditions.

"I didn't feel comfortable doing the exercise because they weren't appropriate. With muscular dystrophy, it's not like you can go to the gym and just get stronger. If I get too much exercise, my condition can go downhill. It's a real balance, so it's important that you get the right kind of physio in the right way.

"I have noticed a worsening of my condition in the year since that physio session. My muscle contractions are getting tighter and tighter, especially in my hands and joints. It can be painful to even lift a glass sometimes. I also have scoliosis. Regular access to the right therapies would help maintain function, movement and independence for as long as possible, but it's really difficult to get the therapies that are designed for my condition."
Sulaiman Khan (31) from London

An expert view

"As physiotherapists, our primary aim should be to support people with muscle-wasting conditions to live well with their condition. In childhood, the developing body and the progression of the condition mean that excellent physical management optimises the child's function and minimises the onset of potentially preventable complications. Every child with a muscle-wasting condition deserves this, but the scant provision of specialist and community paediatric services means that many are missing out. Physiotherapists can bring together the joint effort of schools and communities to support families.



"In adulthood, access is even more restricted, with community service models being geared towards conditions that improve with rehabilitation, not progressing conditions. People with muscle-wasting conditions need to know they have longer-term support from knowledgeable therapists. They need to be re-assured that they can access services quickly, as they need them. Innovative models of self-referral and open access would work well with these conditions. The emphasis should be on enabling people to make their own choices, not to medicalise their lives, like support to enjoy leisure and physical activity. Access to respiratory equipment and specialist therapists is a more pressing issue, with the risk of hospital admission and loss of life. To live in fear of struggling to breathe is a torment too far.

"Finally, to ultimately improve the future lives of people with muscle-wasting conditions, medical and rehabilitation research in this area is paramount. Physiotherapists are central to this. We conduct important functional measurements for drug trials, ensuring that drugs being tested give real benefits to the person. We also conduct and lead our own research into the most effective physiotherapy treatments. Research time is not valued in our NHS work and many trusts do not support this. Our career structure does not reward research or develop future physiotherapy research leaders and innovators.

"It's time to recognise the barriers, see the benefits and support the many committed physiotherapists across the country united in wanting better lives for people with muscle-wasting conditions."

Gita Ramdharry PhD MSc MCSP

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Executive summary

Why is access to physiotherapy so important for people with muscle-wasting conditions?

It is agreed by leading neuromuscular experts that physiotherapy and exercise are good for people with muscle-wasting conditions.

With very few exceptions, there are no drug therapies available for these conditions to slow, halt or reverse muscle damage. Specialist physiotherapy remains a central management option. Physiotherapy aims to optimise and/or maintain independence and muscle function, to allow as good a quality of life as possible for people who, as a result of progressive loss of movement, are less able to participate fully in daily activities.

By developing interventions focused on an individual's needs, physiotherapy can help people with muscle-wasting conditions to:

- ▶ optimise and/or maintain muscle function for as long as possible
- ▶ anticipate, minimise and reduce the risk of secondary physical complications, working jointly with community nurses, respiratory teams and occupational therapists
- ▶ reduce the risk of contractures and loss of movement by developing individualised programmes of stretches, advice on postural management, suitable equipment and, where appropriate, exercises
- ▶ reduce the likelihood and impact of chest infections through timely intervention
- ▶ reduce pain and promote comfort.

Crucially, timely intervention from a multi-disciplinary team (including physiotherapists) can help keep people out of hospital. In 2012, the team at the National Hospital for Neurology and Neurosurgery, in conjunction with the NHS, conducted an audit of unplanned admissions for people with muscle-wasting conditions.¹

The audit found that nearly **40 percent of admissions could have been prevented had patients had access to better co-ordinated care, including timely physiotherapy reviews.**

This comes at a significant cost to the NHS: Muscular Dystrophy UK's report *Invest to Save*² found that the NHS wasted £31m per year on avoidable hospital admissions for people with muscle-wasting conditions. This is a result of a lack of provision of specialist care, of which timely physiotherapy intervention is absolutely central.

Physiotherapy also plays a crucial role in conducting and supporting research and clinical trials. This includes conducting assessments and tests that provide measurements and results, such as the six-minute walk test. Excellent standards of physiotherapy make it more likely a patient will be able to meet baseline entry criteria for a clinical trial. Some physiotherapists design and run research trials themselves, and for research into the physical management of muscle-wasting conditions, it is important that physiotherapists lead the way.

¹ *Unplanned admissions of neuromuscular patients: a collaborative audit*, Hana et al, June 2012

² www.muscular dystrophyuk.org/assets/0002/4945/Muscular_Dystrophy_Campaign_Invest_to_Save_Report.pdf

Key findings

This report follows an extensive survey of people with muscle-wasting conditions (conducted between March and June 2016), interviews with patients and physiotherapists, and two audits of clinical care and clinical trial capacity at UK neuromuscular centres.

Our findings indicate a serious shortage in physiotherapy access across many parts of the country. This lack of provision is having a damaging impact on health, quality of life and – in cases where a patient needs respiratory support – length of life. It is costing the NHS money through avoidable hospital admissions, and is slowing progress in the development of treatments and cures for muscle-wasting conditions.

In most cases, people living with muscle-wasting conditions report that they don't receive any physiotherapy for their condition. Those that do, often don't have access to physiotherapy for long enough, or often enough.

Inconsistencies in the funding and provision of vital aspects of respiratory physiotherapy and equipment – such as cough assist machines – are leading to avoidable admissions to hospital. In some tragic cases, this can even contribute to early loss of life.

Key findings include:

Lack of specialist physiotherapy or support in the community:

- ▶ 60 percent of people with muscle-wasting conditions are unable to access appropriate physiotherapy for their condition
- ▶ 20 percent are paying privately for physiotherapy, highlighting that this is because they feel there is a lack of appropriate provision on the NHS. Some individuals are paying over £6,000 per year for private support.
- ▶ short-term access to physiotherapy – often in six-week blocks – means that people with muscle-wasting conditions report they are receiving little or no benefit as a result
- ▶ 40 percent of UK neuromuscular centres say they urgently need increased physiotherapy capacity
- ▶ although there are examples of excellent community provision in the UK, some community physiotherapists may have limited experience working with people with muscle-wasting conditions, which individuals report is resulting in ineffective and even dangerous treatment. A shortage of specialist physiotherapists means it is harder to upskill these professionals.

Major gaps in respiratory physiotherapy care and provision of breathing equipment:

- ▶ refusal by over 95 percent of Clinical Commissioning Groups (CCGs) to adopt a funding policy for cough assist machines is increasing hospital admissions and can contribute to early loss of life
- ▶ a shortage of respiratory physiotherapists is restricting access to breathing equipment as there is no trained individual to ensure such equipment can be used safely in the community. This is increasing reliance on treatment in hospitals.

A shortage of physiotherapy roles to lead, conduct and support research and clinical trials:

- ▶ specialist neuromuscular centres in the UK are turning down promising new clinical trials owing to a shortage of key roles, including specialist physiotherapists who are funded to work on clinical trials
- ▶ there is a lack of funded research time for physiotherapists to lead and conduct trials in their specialist field of rehabilitation.

What needs to happen?

"My son sees a community physio, about every six months, who suggests stretches for him. We have no idea if we're doing them right. The lack of support we've received has just been catastrophic. There's nothing in our area."

Nick Brown from Lincolnshire, whose 13-year-old son has Duchenne muscular dystrophy

To ensure that people with muscle-wasting conditions are able to get the support they need, Muscular Dystrophy UK is calling for improved provision in the following areas:

Specialist and community physiotherapy

1. Hospital Trusts to ensure that their neuromuscular team includes a specialist physiotherapist, at a minimum level of Band 7, so that all patients with a muscle-wasting condition have a named specialist physiotherapist.

This would ensure that each service is compliant with NHS England's service specification³, which states that all neuromuscular services should include a specialist neuromuscular physiotherapist. It would also end the postcode lottery on specialist physio, and ensure that all patients with muscle-wasting conditions have access to a physiotherapist with knowledge of their condition, who can fully assess their needs, support and develop detailed treatment programmes and advise on equipment and exercise. This would comprise review in clinic, with either a 1 to 1 session for those who are able to travel to their specialist centre, or a referral to local services. Liaison with local teams – to share specific information about an individual's condition and necessary management – will help community teams to best understand the needs of individual patients. The specialist physiotherapist can also act as a point of contact for advice and support with local services and community teams: supporting effective communication between specialist and local teams and helping to coordinate and support joined up working. Specialist physiotherapists have a broad skill set and therefore can play an important role in coordinating patient care across different specialisms, such as neurology or respiratory care. In addition, Hospital Trusts and CCGs should review the caseload of their existing specialist physiotherapists, and support additional roles where caseload is unsustainably high.

2. NHS England, CCGs and Hospital Trusts to work to ensure that patients with muscle-wasting conditions can have access to physiotherapy when they need it, at a clinically appropriate time to allow them maximum benefit.

The current process by which individuals with chronic conditions such as muscle-wasting conditions receive short-term access to physiotherapy – usually in blocks of six weeks – is not appropriate. These individuals often require long-term support, rather than shorter-term provision based on a model more suited to people recovering from a relapse of MS or a stroke, for example. There are excellent examples of open access at specialist centres such as the John Radcliffe Centre in Oxford, and this should be rolled out across all provision. Such an arrangement does not need to entail frequent appointments: if patients are supported to

³ NHS England's Service Specification for Neurosciences: Specialised Neurology (Adult) D04/S/a – Annex B includes children 2013

develop independent management strategies and have access to expert advice, many can self-manage their condition with as few as six physiotherapy sessions per year. Self-referral services have worked especially well in key areas, such as management of musculoskeletal injuries. It has the potential to empower patients to seek help when they know they need it, and to bypass the need for a GP referral.

- 3. CCGs to increase provision of community physiotherapy. Community physiotherapists – linking with a specialist physiotherapist at a specialist centre – can play a crucial role in more regular management of patients at a community level. While requiring an upfront cost, an increase in this provision can actually reduce costs elsewhere.** For example, by implementing strategies that prevent issues such as falls or contractures – which have devastating consequences – patients stay well for longer and this can reduce the associated costs such as increased care hours. Some patients can also experience respiratory difficulties, which may first be identified by a community physiotherapist. In this way, concerns can be quickly escalated and more urgent help received. This also avoids long and costly periods of hospitalisation. Furthermore, CCGs must ensure that community teams are staffed appropriately for people with chronic conditions, and not solely to deal with individuals with set diagnoses.

Role of physiotherapy in clinical trials

- 4. Hospital Trusts to work in conjunction with organisations such as the National Institute of Health Research (NIHR), pharmaceutical companies and patient groups to create posts which maximise clinical funding and research funding.** Currently, already overstretched clinical teams are carrying out research activities outside of core hours and in their own time. Equally, centres report that it can be difficult to retain and recruit dedicated research physiotherapists whose work is restricted solely to clinical trials. Joint-funded initiatives would therefore meet both clinical and research needs at UK Neuromuscular Centres, while also offering an attractive and varied role for experienced physiotherapists. This would benefit patients both in the clinic and on clinical trials. This further enhances the achieving and exceeding of standards of care.
- 5. Development and integration of existing professional networks and organisations that deliver training.** This will ensure high quality delivery of endpoints as well as training in management of muscle-wasting conditions to benefit all patients, whether they are in a trial or not. It can also help ensure that there is a consistent approach to training on clinical trial work, and that all physiotherapists within that network are trained on important outcome measures.
- 6. NHS Trusts to work with patient groups and grant funders to raise the profile of senior physiotherapy researchers, supporting them to be Principle Investigators on grants, obtain PhDs, fellowships and follow professional career pathways.** Dual clinical academic appointments will recognise the achievements of research leaders in physiotherapy who are employed by NHS Trusts. This would reflect the current career structure of medical researchers.

- 7. The NIHR to support work with UK Muscle Centres, patient organisations and industry to ensure that these centres have the necessary research physiotherapy provision and expertise to deliver clinical trials for muscle-wasting conditions.**

Respiratory physiotherapy

- 8. CCGs across the UK to ensure that each respiratory service that sees patients with muscle-wasting conditions includes a specialist respiratory physiotherapist able to work across acute and community settings.**

Respiratory physiotherapists play a vital role in monitoring patients' respiratory status, showing them breathing techniques and providing a much-needed link between hospital and community settings. British Thoracic Society Guidelines⁴ recommend access to a specialist physiotherapist. However, a scarcity of these roles at some centres is resulting in increased hospital admissions because appropriate support is not available in the community. For example, equipment such as cough assist machines cannot be issued in the community because there isn't a member of staff who can provide training on the safe use and maintenance of the equipment. This is all happening at a significant cost to the NHS; better provision of respiratory physiotherapy and equipment in the community is therefore also a cost-effective measure. Recent studies at Alder Hey Children's Hospital and Nottingham Children's Hospital support this approach.⁵

- 9. All CCGs and Health Authorities in devolved nations to adopt a funding policy on cough assist machines.** CCGs have a responsibility for funding cough assist machines in England, as do Health Boards in Scotland, Wales and Northern Ireland. However, only four percent of CCGs in England have a funding policy in place for cough assist machines. This can contribute to funding requests from respiratory healthcare professionals being turned down or delayed unnecessarily, with devastating results for patients' quality of life. In some tragic cases, this can even contribute to loss of life. Recent research at Lane Fox Unit in London and Evelina Children's Hospital showed a reduction in hospital admissions and length of stay among patients issued with a cough assist machine at home (see Appendix III).

- 10. The National Institute for Health and Care Excellence (NICE) to commission research on evidence of clinical-and cost-effectiveness of cough assist machines.** Small-scale studies at centres such as the Lane Fox Unit - as well as patient-reported outcomes - provide evidence the use of cough assist machines in patients with respiratory weakness. However, there is a need to build on the body of evidence supporting the use of this intervention to ensure that it is routinely accepted and funded by CCGs. The Health Minister, Ben Gummer MP, has indicated that NICE is currently considering research proposals in this field⁶, and Muscular Dystrophy UK calls on NICE to commission studies without further delay.

⁴ *Guidelines for respiratory management of children with neuromuscular weakness – British Thoracic Society Guidelines 2012*

⁵ <http://onlinelibrary.wiley.com/doi/10.1111/j.1469-8749.2011.04193.x/pdf>

⁶ <http://www.theyworkforyou.com/wrans/?id=2016-04-27.35834.h&s=cough+assist+machines#g35834.q0>

Specialist and community physiotherapy

"I get muscle spasms which pinch nerves and need to be relieved. Lots of people who have muscle spasms and pinched nerves can get into physio very easily, and I get pushed back to the end of the queue because my condition is permanent and won't get any better."

Lynne Mead, who lives in Wales and has inclusion body myositis

Access to specialist physio

Having access to a specialist neuromuscular physiotherapist as part of a multi-disciplinary team is vital for people living with muscle-wasting conditions. Given that sometimes at a community level there is a lack of knowledge and experience in working with people who have muscle-wasting conditions, it is vital that there are sufficient numbers of specialist physiotherapists spread across the country with specialist knowledge to manage this group of patients.

Three key aspects of a neuromuscular physiotherapist's role are:

1. Direct patient support within specialist clinics

Specialist neuromuscular physiotherapists perform patient assessments and provide advice, support and signposting in consultation with local services. Working in dedicated neuromuscular clinics with consultants and clinical specialist nurses, neuromuscular physiotherapists will have a detailed understanding of treatment pathways and condition management. They disseminate this information and understanding with local teams, to provide the best patient support possible.

2. Supporting community physiotherapists

A vital function of a specialist neuromuscular physiotherapist is the support and education they offer to community health professionals who often will be offering most of the hands-on advice and support to patients in between specialist appointments. Neuromuscular specialist physiotherapists are able to provide both individual patient advice on specific questions and wider training and upskilling for community therapists. This gives them a better overall level of knowledge of muscle-wasting conditions. However, the rarity of many muscle-wasting conditions and frequent staff changes at a community level can add challenges to this.

3. Improving overall care for people with muscle-wasting conditions

Specialist neuromuscular physiotherapists play an important role in assisting the development of evidence-based guidelines, protocols and pathways which aid the care of people with muscle-wasting conditions.

NHS England has identified the importance of a neuromuscular physiotherapist as part of a specialist multi-disciplinary team in the adult neurosciences service specification⁷.

⁷ <https://www.england.nhs.uk/wp-content/uploads/2013/06/d04-neurosci-spec-neuro.pdf>

“The specialist MDT team will comprise of neuromuscular Consultants, neuromuscular physiotherapist, Neuromuscular Nurse, Occupational Therapist, Speech and Language Therapist (SALT), dietician, psychologist and neuromuscular care coordinator. The MDT role will be to assess, diagnose and review patients and their treatment plans.”

However, access to specialist physiotherapy varies regionally meaning that many patients lack the necessary specialist support and appropriate strategies to limit deterioration and help manage their condition. Other regions where provision does exist typically comprises one physiotherapist, whose caseload covers all patients under active follow up at that centre, which can often span well into the thousands.

Services who responded to Muscular Dystrophy UK’s Centre of Excellence audit and identified gaps in provision include:

Nottingham University Hospitals Trust – adult neuromuscular physiotherapist
Leicester University Hospitals Trust – adult respiratory physiotherapist
Cambridge University Hospitals Trust – paediatric respiratory physiotherapist
The Colman Centre – adult respiratory physiotherapist
Oxford University Hospitals Trust – adult and paediatric neuromuscular physiotherapist:
there is currently insufficient resource to support the number of patients under active follow up at the centre

Physiotherapy in the community

Some of the regular support to patients affected by muscle-wasting conditions comes from community physiotherapists. Specialist and community physiotherapy should not be viewed in isolation, as working together plays a crucial role in condition management and patient care. There are good examples across the country of community teams working with specialist neuromuscular teams to provide excellent patient care in the community. It is important to recognise that children will often access physiotherapy at school (for example in PE lessons) and the links between specialist centres and community teams tend to be stronger.

However, findings from our patient survey and reports from individuals living with muscle-wasting conditions reveal a number of difficulties encountered when individuals are treated in the community.

Key barriers to effective treatment in the community include:

- ▶ A lack of awareness of muscle-wasting conditions at a community level, and a shortage of specialist physiotherapists to advise and support individuals to implement tailored programmes of treatment
- ▶ Current commissioning of service models (for example, stroke Early Supported Discharge services) that are aimed at treating individuals with shorter term needs, rather than those with chronic, slowly progressive conditions such as muscular dystrophy

- ▶ Fixed treatment blocks usually of six weeks, which are inappropriate for individuals with long term conditions.

Given the rarity of the conditions, community physiotherapists may not have significant experience working with people with muscle-wasting conditions. Consequently, there can be a gap in knowledge regarding specific considerations necessary for effective treatment of these conditions. Individuals responding to our patient survey report that lack of knowledge of their condition at a community level has resulted in ineffective treatment. Muscular Dystrophy UK is working to upskill community physiotherapists, as is the London and South East Coast Network (for more information see Appendix II) and other teams across the country, such as in Bristol, Newcastle, Preston and Sheffield.

Furthermore, treatment at community level is often very goal orientated. This in itself is appropriate– and individually tailored goals are a central part of physiotherapy. However, due to limited resources at a community level, priority is often given to patients who are in rehabilitation and who would be expected to see marked improvement in time; individuals who are at risk of falling or those at risk of deterioration without rapid access to therapy. For slowly progressive, chronic conditions – such as neuromuscular conditions – goals should be aimed at maintaining function and independence. This is not always well recognised or reflected in eligibility/referral criteria at a community level. There can be a lack of appreciation that timely intervention is crucial for minimising deterioration, and therefore individuals are treated as having non-urgent needs and are not prioritised. In some cases individuals have been told there is no need for treatment as their condition will not improve.

There is therefore an urgent requirement for increased awareness within CCGs, so that the needs of people with muscle-wasting conditions are recognised and taken into account when it comes to the commissioning and design of local physiotherapy services.

A recurrent theme is that individuals are offered treatment blocks of six weeks. This can be dictated by service commissioners or implemented locally to try to best manage demand with limited capacity. Physiotherapy goals for people with muscle-wasting conditions may not always be achievable within such a time limited block.

These patients should therefore be able to have access to physiotherapy when they need it, at a clinically appropriate time to allow them maximum benefit.

Such an open arrangement can be cost effective, and requires a limited number of appointments per year if the patient is educated and supported to effectively self-manage and to live well with their condition.

Comments from our patient survey on specialist or community physiotherapy include:

- ▶ Community physiotherapy is infrequent and not specialised. Specialist physiotherapy at our specialist centre is good but we only see them twice a year

- ▶ My physio has little knowledge of muscular dystrophy, and there are no specialist facilities in my area for people with the condition
- ▶ The physio made it very clear that they were only there to help rehabilitate me, so I could get out of bed and into a chair. There was no greater goal of the therapy
- ▶ We need continuous access to physiotherapy – the length of treatment should reflect the chronic nature of our conditions
- ▶ When I go for physio I find that they are scared to treat me due to not understanding my condition. I have found they are doing more harm than good
- ▶ I feel there is a real lack of specialist physiotherapists
- ▶ In the past when I have sought support, I have found that knowledge and experience of my condition is extremely limited. Any treatment offered is of a general nature, and not tailored to my needs.

Muscular Dystrophy UK is calling for:

- ▶ **Hospital Trusts and CCGs to ensure that their local neuromuscular team includes a specialist physiotherapist.**
- ▶ **Hospital Trusts and CCGs should review caseload of existing specialist physiotherapists, and support additional roles where caseload is unsustainably high.**
- ▶ **Hospital Trusts to ensure that all patients with a muscle-wasting condition have a named specialist physiotherapist.**
- ▶ **NHS England, CCGs and Hospital Trusts to work to ensure that patients with muscle-wasting conditions can have access to physiotherapy when they need it, at a clinically appropriate time to allow them maximum benefit.**
- ▶ **CCGs to ensure that local services are commissioned to include appropriate input and support for individuals with muscle-wasting conditions.**

"Obviously maintaining what little things you can do for yourself is not important enough"

An individual whose daughter has mitochondrial myopathy told Muscular Dystrophy UK:

"My daughter is a full time wheelchair user and uses an electronic communication aid. She needs feeding, toileting, un/dressing, and all aspects of personal care, as well as support and assistance to have any kind of social life.

"She received regular physio while she was at a residential college but that fell apart when she returned home since we're under community services.

"We were referred for physio sessions at our local hospital, but after 3 sessions we were told it was a rehab centre and she needed a neuromuscular centre.

"After a long wait period, a physio finally came out and said they only worked on achievable goals and asked her what she wanted to accomplish. We stressed it was very important for us both that she maintains her ability to transfer from bed/wheelchair/toilet for as long as possible. The physio said while that was important, she needed a specific goal that would have measurable results.

"Obviously maintaining what little part of your life you can do for yourself is not important enough in today's target obsessed climate. We were then put into the physical disability team, where another physio came out and told us the same thing. It's ridiculous."

"There needs to be an ongoing system of support for us"

Alexandra Pawley-Kean has Bethlem myopathy congenital muscular dystrophy

"My experience with the community physio has been pretty frustrating. They're simply not educated on my condition. Education is really what it comes down to. They just don't understand. I mean if I had a pound for every time I had to explain my condition to a medical professional...they should be there to help us, I shouldn't be educating them.

"The system for getting a referral is very, very prescribed and restricted. You have to go to the GP, then they refer you, then you go to the hospital and then they give you a particular allocation of 6 weeks. But the GPs don't always know what you need. I was referred to a physio, but the physio said I needed a different referral. Except the physio couldn't give me that referral, so I had to go back to my GP.

"There needs to be a different system for those of us who aren't going to get better. The allocation system is designed for people with broken bones who will get better in a few weeks or months. That's not us. My muscles are deteriorating as I speak. They're not going to get better. There needs to be an ongoing, long-term system of support for us.

Respiratory physiotherapy

"Our community physiotherapist is not a respiratory physiotherapist. She's unable to help with exercises or help with secretion management"

Mother of a child with spinal muscular atrophy type 1

Improvements in respiratory care have been highlighted as the major reason behind an increased life-expectancy for some forms of muscle-wasting conditions (Eagle et al, Appendix IV).

For example, as recently as 15 years ago individuals with Duchenne muscular dystrophy would not have been expected to reach their 20s. Now, many with the condition are living well into their 30s and even 40s. Similar improvements in length of life have also been experienced by people living with conditions such as some types of spinal muscular atrophy (SMA).

Respiratory physiotherapists perform a central role in the ongoing respiratory care of individuals affected by these and other muscle-wasting conditions. They have an important link to community and family care, training those involved with the patient on how to manage their respiratory needs in a community setting. This includes ordering and training in the use of cough assist equipment, self-assisted cough techniques and ongoing monitoring of respiratory status.

In addition, respiratory physiotherapy plays an important role in weaning patients, enabling earlier discharge and lung function to be kept as strong as possible. 'Weaning' refers to the process in which patients are encouraged to breath without the help of mechanical ventilation.

A recent study at Alder Hey Children's Hospital (Turner et al) showed timely, proactive community based respiratory physiotherapy reduced both A&E admissions and stays in hospital.⁸

However, in many parts of the country respiratory physiotherapists are in significant shortage. Of those that are in post, their high caseload limits often prevents altogether their work in the community. In other cases, patients have no access to a respiratory physiotherapist. In Muscular Dystrophy UK's latest patient survey on respiratory care, 52% of patients in need of respiratory support reported that they had no access to respiratory physiotherapy.

With significant advances in the medical management of patients and an increase in life expectancy, community therapists are seeing patients in the community who previously would have been treated only in hospital. The high caseload of existing specialist physiotherapists – and lack of important roles in some areas of the country – has limited the support that can be provided to community teams.

⁸ <http://onlinelibrary.wiley.com/doi/10.1111/j.1469-8749.2011.04193.x/pdf>

This is a dangerous situation, meaning patients, family members and community physiotherapists have no support or training in airway clearance techniques, which can result in unsafe practice when using equipment, such as cough assist machines. It also means there is no regular assessment or monitoring of respiratory needs. All of this increases the likelihood of chest infections and avoidable emergency admission to hospital, at significant cost to the NHS and to the quality of life of those affected. A study at Nottingham Children's Hospital found that individuals were receiving sub optimal respiratory care due to a lack of physiotherapy support within the region.⁹

Muscular Dystrophy UK is calling for:

- ▶ **CCGs across the UK to ensure that each respiratory service which sees neuromuscular patients includes a specialist respiratory physiotherapist able to work across acute and community settings.**

Case study: Bristol Children's Hospital

The neuromuscular team at Bristol Children's Hospital supports 170 patients affected by muscular dystrophy or a related neuromuscular condition. It is the primary referral centre for children in the South West living with one of these conditions.

The centre has reported a growing number of children in the community who have long term respiratory needs. This requires support within the home setting and complex care plans to cater for their often severe needs.

However, whilst ventilation nurses are in post and a neuromuscular physiotherapist manages the children's physical needs, no support is available to provide respiratory physiotherapy input. This is despite the fact that these children are very vulnerable to respiratory infections which can necessitate lengthy and expensive admissions to hospital.

As a result, there is no support to escalate community physiotherapy treatment, which is resulting in lengthy hospital stays, which for many of these children is occurring on a regular basis.

When an inpatient, use of a cough assist machine is initiated in hospital for these patients. However, when they are discharged into the community they are unable to take this equipment home as they and their community physiotherapist are not qualified to use it and there is no individual within the neuromuscular team at Bristol who can provide the appropriate training and support. Adults in the Bristol area have access to a specialist community respiratory service,

To address this issue, the team at Bristol has put forward a business case for a Band 7 respiratory physiotherapist to improve respiratory support provided to children, and reduce avoidable and costly admissions to hospital.

⁹ <http://onlinelibrary.wiley.com/doi/10.1111/j.1469-8749.2011.04193.x/pdf>

However, local Clinical Commissioning Groups have not supported this, and the team has so far been unable to secure funding. This means that children in the region continue to go without the respiratory physiotherapy care that they need, and the Trust continues to spend scarce funds on preventable hospital admissions.

Access to cough assist machines

"As a parent you cannot help but think if he had access to a cough assist machine sooner, we might not be in this position."

Emma Kemp, whose late son, Freddie, had Duchenne muscular dystrophy

For individuals whose condition affects respiratory function, a weakened cough and difficulty in clearing secretions from the airways is common. Left untreated, this can lead to chest infections and potentially fatal respiratory failure.

Respiratory tract infection is in fact the most common cause of hospital admission for individuals whose muscle-wasting condition causes respiratory weakness.¹⁰

It's therefore imperative that these individuals are given the equipment and care that they need to help combat the effects of their condition, and avoid a hospital admission wherever possible. Evidence indicates that for this group of patients, non-invasive breathing apparatus that assists with coughing (known as 'cough assist machines') can reduce the number and frequency of chest infections and therefore help keep them out of hospital.

Cough assist machines are recommended in NHS England's *Service Specification for Neurosciences: Specialised Neurology (Adult) D04/S/A*. They are also recommended in the British Thoracic Society guidelines and NICE accredited guidelines for the '*Diagnosis and Management of Duchenne muscular dystrophy*'.

Cough assist machines require an upfront cost, but in the long run they save the NHS money. A cough assist machine costs about **£4,500**; a week-long stay in an Intensive Care Unit can cost over **£13,000**¹¹. Recent research at Lane Fox Unit and Evelina Children's Hospital showed a reduction in hospital stays and shorter hospital stays for those patients issued with a cough assist machine at home (see Appendix III)

However, in many parts of the country Clinical Commissioning Groups (CCGs) – whose responsibility it is to commission this equipment – are refusing to provide funding for cough assist machines, and are wrongly claiming it is not their responsibility. This is despite clear guidance, including from Health Ministers in response to questions from MPs.

In response to a Parliamentary Question from Mary Glendon MP on 4 May 2016, the Parliamentary Under-Secretary for Health, Ben Gummer MP, said:

"The provision of cough assist machines is the responsibility of clinical commissioning groups".

However, alarming findings from Muscular Dystrophy UK through Freedom of Information Requests to CCGs reveal that:

- ▶ Fewer than 4% of CCGs said that they had a specific cough assist policy
- ▶ One fifth either referred our FOI to a different agency or wrongly stated that cough assist machines were not their responsibility

¹⁰ Bach JR, Ishikawa Y, Kim H. Prevention of pulmonary morbidity for patients with Duchenne muscular dystrophy. *Chest* 1997;112(4): 1024-1028

¹¹ Figure based on data in '*Invest to Save*', May 2011, http://www.muscular-dystrophy.org/assets/0002/4945/Muscular_Dystrophy_Campaign_Invest_to_Save_Report.pdf

- ▶ Only 1 CCG had any idea of the number of people in their area that might benefit from a cough assist machine.

This lack of support from CCGs is having a devastating impact on people's lives, with many left struggling without the right equipment. Muscular Dystrophy UK is working with and supporting families whose relatives were refused funding for a cough assist machine from their local CCG, which may have contributed to tragic early loss of life.

To address the issue, families and Muscular Dystrophy UK joined with Walsall CCG and respiratory specialists to create a cough assist best practice commissioning policy for CCGs. We are now pushing for all 209 CCGs to end the postcode lottery by meeting their funding responsibilities and adopting a policy.

The House of Commons Public Accounts Committee has also raised the issue of provision and funding of cough assist machines in a session on NHS Specialised Services with Simon Stevens, Chief Executive of NHS England, and Dr Jonathan Fielden, Director of Specialised Commissioning at NHS England.

Muscular Dystrophy UK is calling for:

- ▶ **All CCGs and Health Authorities in devolved nations to adopt a funding policy on cough assist machines, and commit to the routine funding of this important equipment.**

Emma Kemp's son, Freddie, had Duchenne muscular dystrophy:

"While Freddie waited for a cough assist machine to be provided he started to develop a chest infection and generally felt unwell. Between us as a family we called his doctors and respiratory consultant and anyone else we could think of asking why he had not got a cough assist machine yet.

"We were finally told it would be ordered, but by this point though Freddie had really deteriorated and from the end of October he barely left his bed as he was struggling to breathe and was using his bipap machine almost constantly just to keep him comfortable as his breathing was so bad.

"As a parent you cannot help but think if he had access to a cough assist machine sooner, we might not be in this position. Freddie's last few weeks were awful as he was completely dependent on a ventilator we had at home. He had no energy or appetite. He had no quality of life at all. These are not luxury pieces of equipment that we are talking about.

"Commissioners need to realise that investing a small amount can make a huge difference. It's a postcode lottery. If Freddie had lived down the road in Devon, where the local NHS is funding cough assist machines, he would have been issued one two years ago and might still be alive today."



The role of physiotherapy in research

Access to excellent standards of physiotherapy for individuals living with muscle-wasting conditions not only improves health and overall quality of life, but also has valuable implications for research.

Senior physiotherapists have a central role to play in conducting and leading research, with some physiotherapists designing, running and leading research trials themselves. It is important that research into issues such as exercise, orthotics, postural management and seating is physiotherapist led.

However, it is rare for a physiotherapist to have both a clinical and academic role, given an exclusive focus by NHS Trusts on clinical activities in job plans.

It is vitally important that physiotherapists are supporting patients in clinic. However, there is also a benefit to quality of patient care and service developments based on evidence gathered through research. Muscular Dystrophy UK strongly believes that if physiotherapists have skills in this field, they should be allowed to utilise them in their practice. This would also provide physiotherapists with attractive clinical research career paths similar to colleagues in neuromuscular medical research.

Clinical trials

Research into treatments for muscle-wasting conditions such as Duchenne muscular dystrophy is a very promising area, and it's vital that the pipeline of trials and potential therapies is able to continue to grow.

Consistently high standards of physiotherapy has a central role in allowing patients to participate in research. For example, maintaining the best muscle strength and function possible makes it more likely that a child with Duchenne muscular dystrophy could meet the entry criteria for a recruiting clinical trial. When on the trial, it also makes it easier for them to comply with important outcome measures, such as the six minute walk test.

Similarly, if more patients are able to meet clinical trial entry criteria, more clinical trials are able to take place. For rare conditions, this is especially important because the number of patients able to be recruited for a trial is often limited. Physiotherapists themselves conduct assessments and play an important role in the running of the trial.

Clinical trial capacity audit

In a recent Muscular Dystrophy UK audit of clinical trial capacity in the UK, a shortage of physiotherapy and research time was highlighted as a major contributor to the lack of clinical trial capacity at UK centres.

The audit was conducted as part of the 'Newcastle Plan' – joint work with a consortium of Duchenne muscular dystrophy charities to address the underlying lack of clinical trial capacity at UK sites specialising in Duchenne and other types of muscle-wasting condition.

Key findings include:

- ▶ Leading UK muscle centres are turning down clinical trials due to a shortage of key roles, including physiotherapists
- ▶ Six leading muscle centres involved in clinical research highlighted a shortage of physiotherapists as a key concern, given that studies on muscle-wasting conditions often rely so heavily on physiotherapy input. An increase in roles was seen as crucial in maintaining and increasing capacity.
- ▶ There is a lack of funded research time available to physiotherapists at UK muscle centres, which is limiting clinical trial capacity.

Muscular Dystrophy UK is calling for:

- ▶ **Hospital Trusts to work in conjunction with organisations such as the NIHR, pharmaceutical companies and patient groups to create posts which maximise clinical funding and research funding.**
- ▶ **The development and integration of existing professional networks and organisations that deliver training.**
- ▶ **The National Institute for Health Research to work with UK Muscle Centres and patient organisations to ensure that these centres have the research physiotherapy provision and expertise to deliver clinical trials for muscle-wasting conditions.**
- ▶ **NHS Trusts to work with patient groups and grant funders to raise the profile of senior physiotherapy researchers, supporting them to be Principle Investigators or grants, obtain fellowships and follow professional career pathways. Dual clinical academic appointments will recognise the achievements of researcher leaders in physiotherapy who are employed by NHS Trusts. This would mirror the current career structure of medical researchers.**

Get involved

If you have been affected by any of the issues raised in this report, and would like to share your story and get involved with our campaign on physiotherapy, please get in touch with Peter Sutton on p.sutton@muscular dystrophyuk.org or call **020 7803 4838**.

If you are struggling to access support you need, our advocacy team is here to help you. Please get in touch with them at info@muscular dystrophyuk.org or call **0800 652 6352**

What is muscular dystrophy?

There are about 60 forms of muscular dystrophy and related neuromuscular conditions. These conditions cause muscles to weaken and waste over time, leading to increasing disability. The conditions may affect not only the muscles in the limbs, but also those of the heart and lungs, sometimes significantly shortening life-expectancy.

Many of the conditions are low incidence, rare conditions, with some regarded as very rare or ultra-orphan. Muscular dystrophy and related neuromuscular conditions can be genetic or acquired and, with few exceptions, there are currently no effective treatments or cures available.

Clinical trials in some forms of muscular dystrophy and related neuromuscular condition are now underway and it is hoped that these may lead to the introduction of new treatments that can slow or arrest the progression of these often devastating conditions.

About Muscular Dystrophy UK

Muscular Dystrophy UK is the charity bringing individuals, families and professionals together to beat muscle-wasting conditions.

- ▶ We are supporting high-quality research to find effective treatments and cures, and leading the drive to get faster access to emerging treatments for UK families.
- ▶ We are ensuring everyone has the specialist NHS care and support they need, with the right help at the right time, wherever they live.
- ▶ We are providing a range of services and opportunities to help individuals and their families live as independently as possible.

Appendix I: Specialist Neuromuscular Physiotherapist mapping

The following summaries are based on submissions to the Muscular Dystrophy UK Centres of Excellence Audit from 2015 as well as our best knowledge of service provision up until the time of printing.

We recognise that this information is likely to change over time and is only as accurate as what information individual centres are willing to provide us.

Where possible physiotherapy provision is written as the centres themselves have described it.

Centre	Adult physiotherapy provision	Paediatric physiotherapy provision
Addenbrooke's Hospital, Cambridge	1 x specialist adult Neurophysiotherapist with an interest in Neuromuscular conditions	2 x part time Paediatric Neuromuscular Physiotherapists
Alder Hey Children's Hospital, Liverpool		2 x part time Paediatric Neuromuscular Physiotherapists
Bart's Neuromuscular Centre, London	1 x part time senior neurophysiotherapist	
Birmingham Heartlands		1 x Senior Paediatric Physiotherapist for Neuromuscular clinic
Royal Bristol	1 x full time adult neuromuscular physiotherapist	1 x part time Paediatric Neuromuscular Physiotherapist
Great Ormond Street Hospital, London		2 x full time Paediatric Neuromuscular Physiotherapists, 1 x full time physio, 1x part time physio, 1 x full time research physio, 1 x part time research physio
Evelina Children's Hospital, London		2 x part time Paediatric Neuromuscular Physiotherapist
John Walton Muscular Dystrophy Research Centre, Newcastle	2 x part time adult and paediatric neuromuscular physiotherapists, 1 x full time physio TI	2 x part time adult and paediatric neuromuscular physiotherapists, 1 x full time physio TI

Centre	Adult physiotherapy provision	Paediatric physiotherapy provision
John Radcliffe , Oxford	1 x full time neuromuscular physiotherapist	1 x full time paediatric neuromuscular physiotherapist
King's College, London	2 x Specialised Muscle Neurophysiotherapist	
Leeds Teaching Hospitals Foundation Trust		2 x part time and paediatric neuromuscular physiotherapists
Leicester Teaching Hospitals	Rehabilitation physiotherapists only	Rehabilitation physiotherapists only
Morrison Hospital, Swansea		1 x Part time Paediatric Neuromuscular Physiotherapist
National Hospital for Neurology and Neurosurgery, London	2 x full time neuromuscular physiotherapists, 1 x part time academic physiotherapist	
Plymouth Hospitals NHS Trust	2 x part time physio	
Queens Medical Centre, Nottingham	none	1 x Part time Paediatric Neuromuscular Physiotherapist
Robert Jones and Agnes Hunt Orthopaedic NHS Trust, Shropshire	1 x full time adult and paediatric neuromuscular physiotherapists	1 x full time adult and paediatric neuromuscular physiotherapists
Sheffield Teaching Hospitals NHS Foundation Trust	2 x full time neuromuscular physiotherapists	
Royal Manchester Children's Hospital		1 x part time neuromuscular physiotherapist, 1 x part time neurosciences physiotherapist
Royal Preston		1 x part time neuromuscular physiotherapist

Centre	Adult physiotherapy provision	Paediatric physiotherapy provision
Sheffield Children's Hospital		1 x part time neuromuscular physiotherapist, 1 x full time neuromuscular physiotherapist
Suffolk Community Healthcare		2 x part time physiotherapists with specialist expertise
The Walton Centre for Neurology and Neurosurgery, Liverpool	1 x part time neuromuscular physiotherapist	
University Hospital of Wales, Cardiff	none	none
Queen Elizabeth Hospital, Birmingham	3 x part time specialist neuromuscular physiotherapists	
Derby Hospital	2 x part time community physiotherapists	2 x part time community physiotherapists
Wessex Neurological Centre, Southampton	2 x part time neuromuscular physiotherapist	
Bradford Teaching Hospitals NHS Foundation Trust	1 x full time neuromuscular physiotherapist	
St Georges Hospital	none	

Appendix II

Muscular Dystrophy UK's Physiotherapy e-learning module

The Muscular Dystrophy UK physiotherapist eLearning module was launched in November 2015 and has now reach almost 650 physiotherapists across the country. Since launching the course has gone from strength to strength and become a key part of our upskilling work.

After receiving feedback from people with muscle-wasting conditions who said that community physiotherapists sometimes struggle to understand the needs of their condition, we partnered with Kingston University & St George's University of London to develop this resource. The module has been designed to provide important information that will help physios build a foundation of knowledge and skills around the management of muscle-wasting conditions in adults.

The module focuses on 11 short topics that develop critical understanding of the management of muscle-wasting conditions. The topic areas, which include four core topics and seven optional topics, are:

- ▶ overview of neuromuscular conditions (core topic)
- ▶ pathology and presentation of neuromuscular conditions (core topic)
- ▶ assessment and outcome measures (core topic)
- ▶ exercise (core topic)
- ▶ orthotic management
- ▶ respiratory management
- ▶ transition
- ▶ contracture management
- ▶ postural management
- ▶ wheelchairs and mobility
- ▶ fatigue

The module can be access remotely via a website and physiotherapists who complete the core topics and two optional topics are able to request a certificate for their CPD (Continuing Professional Development) file.

Gita Ramdharry, Academic Neuromuscular Physiotherapist who was responsible for coordinating and developing the course content has since played an active part in the promotion of the course. Alongside Muscular Dystrophy UK she has worked with the Chartered Society of Physiotherapy and the Association of Chartered Physiotherapists in Neurology in order to promote the module to as many people as possible.

Jane Mellor (pictured), a community physiotherapist, has a daughter, Kate, with merosin deficient congenital muscular dystrophy

“The course is about using and adapting the skills we already have as physiotherapists to support the long term management of people with muscle-wasting conditions”.



Appendix III

Title

A Pre-Post Intervention Study of the Effect of Domiciliary Mechanical Insufflation-Exsufflation and the Incidence of Crisis Admission in Patients with Duchenne Muscular Dystrophy (DMD)

Authors

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#Guy's & St Thomas's NHS Foundation Trust & Kings College London NIHR Comprehensive Biomedical Research Centre, London, UK

Abstract

Although there are trials ongoing, there is limited evidence to support the use of mechanical insufflations-exsufflation as a method of secretion clearance in patients with DMD. Despite this, our local unit practice is to provide a cough assist machine to DMD patients that are established on non invasive ventilation with a peak expiratory cough flow < than 160 litres per min despite maximal physiotherapy adjuncts when they have either: (1) > 2 episodes of chest sepsis per year requiring antibiotics and hospital admission (2) 1 episode of severe chest sepsis requiring invasive ventilation. The aim of the use of the cough assist machine is to reduce the frequency of hospital admissions. We reviewed prospective data from purpose built database of our DMD patient cohort over a 7 year period. Over this period, 32 patients had been issued with a domiciliary cough assist machine. We reviewed the admission data for 12 months prior to issuing the device and the subsequent 12 months post issue in order to establish the effect on admission frequency and length of stay.

Number of patients 32	Pre home cough assist issue	Post home cough assist issue	Significance
Mean frequency of respiratory admissions in a 12 month period	3(1-6)	0.3 (0.1)	P<0.001*
Mean LOS of respiratory admissions in a 12 month period	12.9 (2-67)	11.4 (0-51)	P<0.001*

Data are expressed as absolute value (range).

* Significantly different using wilcoxin signed ranks test

The data shows a significant reduction in both hospital admission frequency and length of stay for those patients issued with a domiciliary cough assist machine. These data add support to the use of domiciliary cough assist machines in DMD patients with an ineffective cough and previous episodes of chest sepsis already established on NIV.

This project has been supported by the Guy's & St Thomas' Charity (Project Code: G080109)

Title

Timely Respiratory Review and Coordination of Care at the Age of Transition can Reduce the Incidence of Unplanned Respiratory Crises requiring ventilation in patients with Duchenne

Authors

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#Guy's & St Thomas's NHS Foundation Trust & Kings College London NIHR
Comprehensive Biomedical Research Centre, London, UK

Abstract

The introduction of timely home mechanical ventilation (HMV) in DMD has been associated with increased life expectancy. In 2009, we introduced a transitional care coordinator role to coordinate respiratory review and intervention during adolescence with the hypothesis that we would reduce the number of unplanned respiratory crises and length of hospital stay (LOS). We have prospectively gathered data in a purpose designed database for all children and young adult DMD patients currently under review. Out of a population of 75 patients under review in 2009, 34 were already receiving non invasive ventilation (NIV) with 60 under respiratory screening review. Data are shown in the table below.

	2009	2010	2011
Total number of DMD patients > 10 years old under review	75	86	101
Number of DMD patients receiving HMV	34	46	60
New elective HMV initiations			
Mean age of initiation NIV (years)			
Mean length of set up - elective (days)			
Mean length of set up - emergency (days)			
Number of new patients requiring invasive ventilatory support and requirement for ongoing HMV			
Number of new patients requiring invasive ventilatory support and tracheostomy formation and requirement for ongoing HMV			
Level 3 (ICU) bed days			
Level 2 (HDU) bed days			
LOS (days) from tracheostomy insertion to removal			

*this patient did not require long term nocturnal NIV; data are expressed as absolute values (range)

Analysis of these data was focused on the primary respiratory crisis admission requiring ventilatory support. Within the 3-year period, we identified 19 patients who required initiation of ventilation. All patients were known to the respiratory team prior to experiencing their first episode of respiratory failure and only 15.8% were as a result of an emergency admission. Out of the 19 patients initiated on NIV, 16 were electively admitted to our unit with a mean length of stay of 4.7 days (2-9) days. 1 patient was admitted directly from clinic as an emergency, requiring invasive ventilation but managed without tracheostomy formation. Of the two patients who required invasive ventilation with tracheostomy formation, one did not require NIV post discharge from hospital. These data lend support for the need for specialist respiratory review and careful coordination of care before the 1st respiratory crisis. The data supports that with timely intervention you can reduce the need for emergency initiation and the severity of the crisis which is also reflected in a reduced length of hospital stay.

[This project has been supported by the Guy's & St Thomas' Charity \(Project Code: G080109\)](#)

Appendix IV

Unplanned admissions of neuromuscular patients: a collaborative audit, Hana et al, June 2012

Number of admissions	All admissions (%)	Neuromuscular related admissions (%)
Yes	216 (37.5)	143 (63.0)
Possibly	28 (4.9)	13 (5.7)
No	327 (56.8)	67 (29.5)
Could not be determined	5 (0.8)	4 (1.8)
Total	576	227

Intervention / measure	All preventable admissions (%)	Prevented neuromuscular related admissions (%)
Surveillance of patients condition	139 (26.5)	114 (29.9)
Access to neuromuscular care	116 (22.1)	98 (25.7)
Having an emergency plan	60 (11.5)	59 (15.5)
Access to/liaison with other services*	41 (7.8)	13 (3.4)
Prevent delay in referral to a neurology service	32 (6.1)	11 (2.9)
Provision of equipment (including orthotics)	34 (6.5)	34 (8.9)
Prevent delay in initial diagnosis	29 (5.5)	8 (2.1)
Patient/parent education	23 (4.4)	18 (4.7)
Physiotherapy referral/review	22 (4.2)	16 (4.2)
Monitoring of repeat admissions for recurrent symptoms	15 (2.9)	4 (1.0)
Access to social services	6 (1.1)	1 (0.3)
Access to alcohol/substance abuse services	3 (0.6)	0 (0.0)
Having a discharge plan	3 (0.6)	4 (1.0)
Better transition to adult care	1 (0.2)	1 (0.3)
Total	524	381

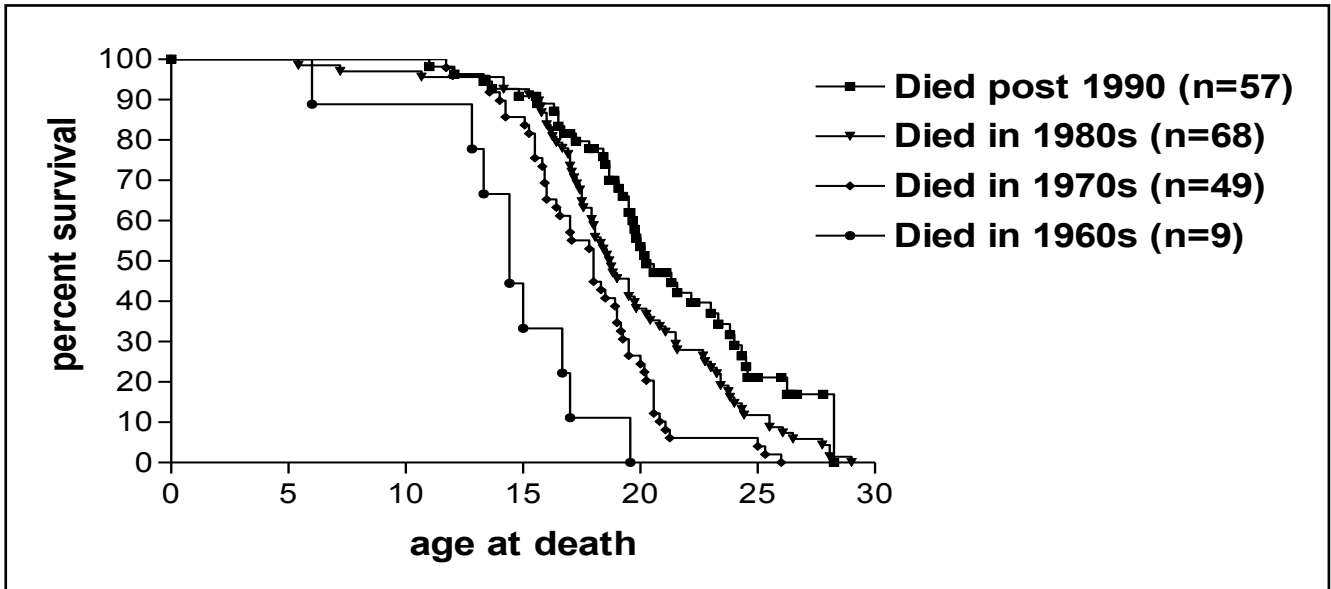
*other services include respiratory, cardiology, palliative care, oncology, psychiatry, diabetes, urology, ophthalmology and care of the elderly.

Appendix V

Duchenne muscular dystrophy survival

Duchenne Muscular Dystrophy Survival data 1960-1990

(Eagle et al Survival in Duchenne muscular dystrophy: improvements in life expectancy since 1967 and the impact of home nocturnal ventilation)



The authors reviewed the notes of 197 patients with Duchenne muscular dystrophy whose treatment was managed at the Newcastle Muscle Centre from 1967 to 2002, to determine whether survival has improved over the decades and whether the impact of nocturnal ventilation altered the pattern of survival.

Results:

1960s: mean life expectancy: 14.4 years – no survivors beyond 19.29 years

1990s: mean life expectancy: 19.5 years

Improvement as a result of multi-disciplinary care

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Muscular Dystrophy UK

Fighting muscle-wasting conditions



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