



**Neuromuscular patients with complex respiratory needs must have access to the right care and equipment when they need it.**



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## Respiratory weakness in patients with a muscle-wasting condition

Respiratory difficulties are a common feature of muscle-wasting conditions. This is because of weakness in muscles in the throat, chest wall and diaphragm, which are required to move air in and out of the lungs.

This weakness can also lead to a weakened cough, which makes chest infections and blocked airways more likely.

Many people with muscle-wasting conditions are vulnerable to under-breathing (hypoventilation), caused by a build-up of carbon dioxide, which leads to drowsiness, frequent headaches and can even result in a person's becoming unconscious. Often this problem first becomes evident at night because one's breathing can be shallower when asleep. The high carbon dioxide levels and under-breathing also decrease oxygen levels, thereby increasing a person's difficulties. Added to this, some patients are very sensitive to oxygen treatment, which unless used carefully, can make the hypoventilation more severe.

Given these difficulties, it is essential that patients at risk of respiratory difficulties have their respiratory function monitored regularly. It is also vital they should have access to specialist support and equipment to keep respiratory function as strong as possible and prevent potentially fatal respiratory problems.

## Foreword

"My sister, Christine Forrest, was diagnosed with oculopharyngeal muscular dystrophy (OPMD) ten years ago, at 55 years of age.

"In the last two years of her life, she had four bouts of pneumonia and also suffered from bronchiectasis (a condition where the airways of the lungs become abnormally widened, leading to a build-up of excess mucus). Aspiration issues (whereby liquids or food are breathed into the lungs or airways) were so severe that she was admitted to hospital three times during this period.

"Christine was never referred to a respiratory specialist for assessment, or given any respiratory support even though she clearly had respiratory complications.

"At all times, she was given treatments at her local hospital for symptoms as they presented. There was no co-ordinated approach between her GP, neurologist and the professionals in acute and secondary care.



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## **"Experiences like my sister's must not be allowed to happen again."**

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"Christine spent the last seven months of her life in hospital, the majority of which was spent on an acute ward. At this time, she was given extensive respiratory physiotherapy to stabilise her severe chest infection.

"Experiences like my sister's must not be allowed to happen again. Neuromuscular patients with complex respiratory needs must have access to the right care and equipment when they need it."

"In December 2013, she left hospital briefly and was admitted to a residential care home. At no stage during transfer was provision made for her respiratory requirements, despite her being at clear risk and having received respiratory support while in hospital.

"Christine had to be admitted urgently back to hospital on 26 December and died at the end of January 2014.

"With Muscular Dystrophy UK, I am heavily involved in campaigning for better care and support. I have found out much more about respiratory care in other parts of the country. I have no doubt that had Christine had access to care at one of these centres, she would have got the respiratory support her condition required.

"Experiences like my sister's must not be allowed to happen again. Neuromuscular patients with complex respiratory needs must have access to the right care and equipment when they need it."

**Bryan Gould**

February 2015

Given the extremely distressing time Christine and her family went through, Muscular Dystrophy UK believes there must be a clear care pathway established for patients with muscle-wasting conditions who have severe respiratory complications. There should be close co-ordination at all times between professionals at different levels of care, with awareness of the local support that can be provided by specialist respiratory teams.

Bryan is taking forward the fight for specialist respiratory care for people affected by muscle-wasting conditions. We are pleased to give Bryan the opportunity to tell Christine's story and to be able to dedicate this report to her memory.

## **Calls to action**

- 1. We call on the Secretary of State for Health and the Chief Executive of NHS England to issue a joint, written directive placing a clear obligation on all Clinical Commissioning Groups (CCGs) to routinely fund the use of cough assist machines, where a clinical need has clearly been identified.**

Cough assist machines are recommended in the NICE (National Institute for Health and Care Excellence) Accredited Guidelines for Duchenne muscular dystrophy and in NHS England's neuromuscular service specifications. They are also recommended and used by respiratory experts across the country. Published research and patient-reported outcomes point to their efficacy in clearing secretions and preventing hospital admissions. Far from saving the money, not investing in equipment such as cough assist machines is actually increasing NHS costs. A cough assist machine costs about £4,500: a week-long stay in an Intensive Care Unit can cost more than £13,000<sup>1</sup>. CCGs and Health Authorities in some parts of the country, in particular in the West Midlands, are not routinely commissioning these items of equipment. We believe that the need for CCGs to fund cough assist machines must be reinforced at the highest levels, given the clear risk at which these patients are placed without them.

- 2. We call on all CCGs and relevant Health Authorities not already doing so, to firmly commit to the routine commissioning of cough assist machines in order to improve patient quality of life, reduce preventable emergency admissions to hospital and save significant amounts of money for the local NHS.**

- 3. We call on Health Authorities across the UK to introduce targets with clear timescales for discharging complex respiratory patients. Throughout the process, there must be close liaison with the patient's specialist centre, who we believe are well placed to provide care agencies and community health professionals with training and advice.**

Every day counts for people living with severe and life-limiting conditions, including many types of muscle-wasting conditions. People living with these conditions have every right to independence and as good a quality of life as possible. However, this is not achieved when people with complex respiratory needs spend weeks and months stuck in hospital owing to avoidable delays in their being discharged from hospital. Unless these delays are addressed, bed capacity will continue to be lost and the strain and stress placed on patients and families, already living with life-limiting conditions, will remain.

**4. We call on NHS England and relevant Health Authorities in Scotland, Wales and Northern Ireland to ensure that equipment reliant on electricity, such as ventilator machines, have battery back-up. Individuals should also be given grants to purchase generators that switch over automatically, so they can continue using vital equipment in the event of power failures. Carers must also be fully trained in manual breathing assistance techniques for times when there is no access to a power supply.**

**5. We call on Health Authorities across the UK to ensure that each respiratory service includes a specialist respiratory physiotherapist able to work across acute and community settings.**

Specialist 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. It is alarming that 52 percent<sup>2</sup> of neuromuscular patients requiring respiratory support say they have no access to respiratory physiotherapy.

**6. We call on the Health Authorities in all four UK countries to ensure that there is a specified trainer within each Regional Centre who can liaise regularly with an identified respiratory care lead within the patient's care agency. In this way, care workers will be equipped with the necessary skills and competencies.**

Patients and specialists report that home care workers are often unaware of how to maintain breathing support measures and there can be difficulties in managing equipment. Centres such as the Lane Fox Respiratory Centre in London have a Home Mechanical Ventilation team, with responsibility for liaising with care workers and overseeing support for complex ventilated patients in the community. We believe this is an excellent model with potential to be delivered elsewhere.

**7. We call on NHS England to ensure that the tariff respiratory services receive is sufficient to enable comprehensive management and monitoring of respiratory status for all patients who need it.**

Many experts in respiratory care reported concerns that their service did not receive sufficient funding to enable comprehensive inpatient and outpatient assessment. For example, in some services there is a need for increased consultant time so patients can have regular reviews. This shortfall was also reflected in our *Patient Survey*<sup>3</sup>, with nearly 20 percent of patients reporting that they had no access to a sleep study. Sleep studies monitor an individual's breathing when they are asleep, and help detect respiratory difficulties.

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<sup>2</sup> State of the Nation: National Patient Survey, Muscular Dystrophy Campaign, 2013

<sup>3</sup> State of the Nation: National Patient Survey, Muscular Dystrophy Campaign, 2013

## Executive summary

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).

Whereas boys and young men living with Duchenne muscular dystrophy would not have been expected to reach their 20s as recently as 15 years ago, some with the condition are now 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).

There are many excellent examples across the UK of respiratory care. However, following an extensive *Patient Survey*<sup>4</sup>, analysis of NHS emergency admissions and consultation with specialists across the country, this report reveals how many patients with complex respiratory needs are going without the right specialist support and equipment. This leaves them vulnerable to life-threatening respiratory failure.

People's quality and – in some cases – length of life are being severely reduced owing to a lack of access to respiratory care.

### Key findings include:

- ▶ as many as one third of people in need of a cough assist machine are unable to access one. Failure to routinely provide equipment such as cough assist machines is putting patients' lives at risk, placing enormous strain on them and their families and leading to avoidable and costly admissions to Intensive Care Units
- ▶ families are being forced to consider funding cough assist machines themselves, with one family in the West Midlands resorting to looking for a cough assist machine on eBay, as their local NHS would not provide funding
- ▶ delays in setting up care packages is leading to delays in discharging complex ventilated patients from hospital. This is resulting in lost bed days, further costs to the NHS and lengthy and unnecessary stays in hospital
- ▶ alarming findings from Muscular Dystrophy UK's recent *Patient Survey*, including over half of patients reporting they were unable to access vital respiratory physiotherapy.

These findings all reflect an unacceptable situation, which needs to be addressed as a matter of urgency.

Improvements in respiratory care and the resulting increase in life-expectancy have allowed people a quality of life and opportunities that previously would not have been possible.

We believe all people affected by muscle-wasting conditions should be able to enjoy these opportunities, regardless of where in the country they happen to live.

Muscular Dystrophy UK is calling on the organisations listed in this report to act urgently to bring about change.

## Access to specialist equipment

“Treating patients with neuromuscular conditions using a mechanical insufflator-exsufflator can be extremely cost-effective. We have found that by using these machines, we can prevent admissions to intensive care or high dependency units. In fact, a mechanical insufflator-exsufflator pays for itself if it prevents just one two- or three-day admission to a paediatric intensive care unit.”

### Dr Stefan Spinty

Consultant Paediatric Neurologist and Neuromuscular Lead Clinician, Alder Hey Hospital

Some people with a muscle-wasting condition have a weak cough which can lead to lung infections and respiratory failure. Evidence indicates that non-invasive breathing apparatus that assists with coughing (‘cough assists’) can effectively reduce the number of chest infections people get. In addition, there is evidence to suggest cough assist machines can reduce treatment time, when added to standard courses of treatment for airway clearance (Chatwin 2009).

For neuromuscular patients with respiratory weakness, respiratory tract infections are the most common cause of hospital admissions<sup>5</sup>. Having access to equipment that can provide an effective cough is therefore essential for keeping patients out of hospital and avoiding emergency admissions.

The introduction of cough assist machines in a timely manner significantly improves patients’ quality of life. This is reflected in a substantial body of expert guidelines, which recommend the use of these machines. These include NHS England’s *Service Specification for Neurosciences: Specialised Neurology (Adult) D04/S/A*. They are also recommended in the British Thoracic Society and NICE accredited guidelines for the *Diagnosis and Management of Duchenne muscular dystrophy*.

Cough assist machines also save the NHS money, with clinical evidence and patients’ experiences pointing to fewer hospital admissions.

As an example, the table below documents a study of 39 patients who received respiratory support (including a cough assist machine) at home. Through this provision, 34 hospital admissions were prevented for this group of patients. An audit of unplanned admissions estimates the median length of hospital stay is six days (Hana et al. 2011). Admissions to a specialist ward can cost the NHS up to £1,925 per day<sup>6</sup>. Therefore, in the case of the study below, an estimated £346,500 of admissions was avoided by the use of respiratory support, including cough assist machines. This contrasts with £4,500, which is what one cough assist machine costs the NHS.

	Total Patients Having Home Visits (n = 27)	Patients With Home Visits Only (n = 15)	Patients With Home Visits Plus MI-E <sup>a</sup>	TIV Users Having Home Visits and Provision of MI-E (n=9)	NIV Users Having Home Visits and MIE (n=3)
Total respiratory exacerbations, n	67	20	47	40	7
Home visits without MI-E delivery, n	13	4	9	9	0
Home visits plus provision of MI-E, n	21	0	21	18	3
Patients hospitalized, n	18	8	10	7	3
Total hospitalizations, n	33	16	17	13	4
Avoided hospitalizations, n	34/67 (51%)	4/20 (22%)	30/47 (64%)	27/40 (67%)	3/7 (42%)

Avoided hospitalizations are defined by the domiciliary need for continuous ventilatory support and correction of a persistent decrease in baseline oxyhemoglobin saturation by assisted coughing. / <sup>a</sup>only the TIV and NIV users required home visits plus MI-E, all of which resulted in correction of SpO<sub>2</sub> baseline by some combination of adjusting ventilator use and assisted coughing. / TIV, tracheostomy intermittent positive pressure ventilation; NIV noninvasive intermittent positive pressure ventilation; I-E, mechanical insufflation-exsufflation.

Source: Muscular Dystrophy Campaign, Invest to Save

<sup>5</sup> Bach JR, Ishikawa Y, Kim H. Prevention of pulmonary morbidity for patients with Duchenne muscular dystrophy. *Chest* 1997;112(4): 1024-1028

<sup>6</sup> 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](http://www.muscular-dystrophy.org/assets/0002/4945/Muscular_Dystrophy_Campaign_Invest_to_Save_Report.pdf)

## Patient survey findings

We are concerned that in some cases these vital pieces of equipment are not being funded at all. This leaves patients with very complex respiratory needs vulnerable to respiratory tract infection and life-threatening respiratory crises.

In our 2013 *Patient Survey*, nearly 30 percent of respondents whose condition affected respiratory function reported they had no access to a cough assist machine. We are concerned that thousands of people may be going without this vital equipment.

While in some areas of the country, patients have access to cough assist machines, in other areas the local NHS will not fund them. For example, in the West Midlands, cough assist machines are not routinely provided at hospitals. This means specialists are being forced to submit complex and lengthy individual funding requests. In other cases, requests are being refused altogether and we have heard reports of boys and young men affected by Duchenne muscular dystrophy having been admitted to an Intensive Care Unit with respiratory failure. This could be avoided if patients were given the correct equipment and support, including cough assist machines.

In answers to Parliamentary Questions in late 2014, the Parliamentary Under-Secretary of State for Health, Jane Ellison MP, made clear that “the equipment and respiratory support of cough assist is the commissioning responsibility of clinical commissioning groups”<sup>7</sup>

**We therefore call on all CCGs and relevant Health Authorities not already doing so, to firmly commit to the routine commissioning of cough assist machines. This will improve patient quality of life, reduce preventable emergency admissions and save significant amounts of money to the local NHS.**

We believe this must be reinforced at the highest levels, given the clear risk at which patients are placed without a cough assist machine.

**We call on the Secretary of State for Health and the Chief Executive of NHS England to issue a joint, written directive placing a clear obligation on all CCGs to routinely fund the use of cough assist machines, where a clinical need has clearly been identified.**

## **Gill Finlayson's son, Christopher (24), from the West Midlands, had Duchenne muscular dystrophy and suffered from severe respiratory difficulties**

In the last two years of his life, Christopher had been admitted to hospital four times with serious respiratory difficulties. Each admission lasted at least a week.

However, despite requests from the professionals involved in Christopher's care, he was never provided with a cough assist machine. This was owing to the funding policies of his local NHS.

At one point, Gill even took to eBay to look for a cough assist machine. However, because of the high costs of maintaining the machine (costs which the family's local NHS also refused to fund), the family was unable to purchase the machine themselves.

In October 2014, Christopher was admitted to hospital and tragically died from respiratory failure.

Gill's younger son, Robin, also has Duchenne muscular dystrophy. She is passionate about continuing the fight to ensure CCGs routinely fund vital cough assist machines.

## **Trish Kerrison's sons, Steve (21) and Joe (19), from the East Midlands, have Duchenne muscular dystrophy**

"At Christmas in 2012, our boys both developed colds and we visited their specialist centre to use a cough assist machine. However, despite repeated efforts from healthcare professionals over the next 12 months, all requests to obtain a cough assist machine to use at home were turned down.

"During this period, Steve developed a debilitating cough which could last for up to five hours. He spent increasing lengths of time on a ventilator, interfering with his studies, making him reluctant to go out and reducing his quality of life.

"In November 2013, Joe experienced extreme breathing difficulties and his oxygen saturation deteriorated to 85 percent. He was taken by ambulance to the resuscitation department at Nottingham and was then transferred to intensive care. He spent the next seven days on the High Dependency unit. The CCG still refused a cough assist machine for home use after he was discharged.

"After a long battle and campaign, in which we involved our local MP, we were finally able to secure funding for cough assist machines for Steve and Joe.

"Both Steve and Joe play powerchair football and regularly meet young men from across the country with muscular dystrophy. They were well aware that had they lived in Newcastle, a machine would have been funded without difficulty.

"Now finally having obtained cough assist machines, the boys have peak flows of 120-130L/min. Steve has no need for a ventilator during the day and we now know that if they get colds during the winter, we will have some weapons to deal with it.

"However, in the long period without this equipment, my sons' health was severely compromised. Other families should not have to go through the same strain and distress we have been through. It is unacceptable that vital equipment is not being routinely provided."

## Respiratory physiotherapy and support for patients in the community

The support of professionals with expertise in respiratory care is essential to the effective management of some muscle-wasting conditions. While in some parts of the country there are examples of excellent respiratory services with a full range of professionals, we are concerned that this is not the case in all areas.

Respiratory physiotherapy can be done by a specialist physiotherapist or there are techniques, including a self-assisted cough, that carers and family members can perform themselves.

In some teams, respiratory physiotherapists are attached to the specialist respiratory service and operate in both acute and community settings. This is as part of a Home Mechanical Ventilation Team (HMT) and there are excellent examples of this practice across the country.

Specialist physiotherapists play an important role in training those involved with the patient on how best to manage their respiratory needs and equipment in the community. They are also able to monitor respiratory status and introduce self-assisted cough techniques wherever necessary.

In addition, respiratory physiotherapy plays an important role in weaning patients, enabling earlier discharge and keeping lung function as strong as possible. Weaning refers to the process in which staff try to get a patient to breathe without the help of mechanical ventilation.

In our recent patient survey, 52 percent of patients in need of respiratory support reported that they had no access to respiratory physiotherapy. This is a dangerous situation, which could mean patients are going without access to vital techniques to manage their condition. There may also be reduced monitoring of their respiratory status. All of this increases the likelihood of an avoidable emergency admission to hospital.

**We therefore call on Health Authorities across the UK to ensure that each respiratory service which sees neuromuscular patients includes a specialist physiotherapist able to work across acute and community settings.**

The model of care in the UK for patients with muscle-wasting conditions is very much within the home or, if at adult stage, with support to live independently. There is consequently a lot of pressure on carers and family members (who often act as primary carers) to understand and maintain breathing support measures, as well as maintain complicated equipment.

Centres such as the Lane Fox Respiratory Centre in London have an HMT team, with responsibility for liaising with care workers and overseeing support for complex ventilated patients in the community. We believe this is an excellent model with potential to be delivered elsewhere.

**We also believe there is a role for specialist centres to play in providing training in generic tracheostomy or ventilator care. We call on the Health Authorities in all four UK countries to ensure there is a specified trainer within the patient's regional centre, such as a respiratory physiotherapist. By liaising regularly with an identified respiratory care lead within the patient's care agency or local community healthcare team, necessary skills and competencies can best be ensured.**

## Delayed discharge from hospital for neuromuscular patients with complex respiratory needs

For some individuals affected by muscle-wasting conditions, respiratory difficulties may be so severe that mask ventilation, so-called non-invasive ventilation, proves ineffective. In such cases, a tracheostomy may be considered.

A tracheostomy is a small tube inserted in the front of the neck to access the trachea (windpipe). The tracheostomy tube is then attached to a ventilator to assist breathing. A patient will take an active decision, following an extensive discussion with the medical team, to have a tracheostomy, because non-invasive ventilation is not proving effective. In some cases, a conversion to tracheostomy ventilation is out as an emergency procedure during a respiratory crisis when non-invasive ventilation is no longer effective, or swallow function is severely impaired. The conversion to tracheostomy ventilation, albeit a simple procedure, requires a lengthy stay in hospital before the patient is discharged into the community as a nursing home or home care package needs to be put in place. This requires careful assessment of the patient, agreement of funding by the Clinical Commissioning Group (CCG) and the community care package to be developed.

Muscular Dystrophy UK is concerned at the lack of co-ordination between the acute trusts and the CCG assessment. There are also delays in the funding decision from the CCG and then further delays in setting up the care package. This may be the consequence of difficulty sourcing an appropriate provider and ensuring that the correct equipment is available. As a result, patients are often waiting months before they are able to leave hospital following initiation of tracheostomy ventilation.

In 2010, Neil Patel, a 33-year-old with Duchenne muscular dystrophy, spent five months in hospital following a tracheostomy, while his local Primary Care Trust (PCT) recruited and trained his carers.

At the time, Neil said:

"It was a really hard decision to have a tracheostomy but I had become reliant on the ventilator to keep me alive. I was assured by the PCT that my carers would be trained and ready for me to go home within weeks of the operation. They made a complete mess of it. It took them five months to sort out, leaving me no choice but to stay in hospital unnecessarily.

"I feel so frustrated that I have wasted a massive chunk of my life - if I'd known that it would take so long to employ my carers, I would probably have not had a tracheostomy."

Four years on, this situation may have been expected to have improved. However, the move from PCTs to CCGs has done nothing to address the issue of delayed hospital discharges, and patients continue to report unacceptable delays before a discharge into the community.

### Case study: Lane Fox Unit, London

The Lane Fox Respiratory Unit at St Thomas' Hospital, London, is a national referral centre, specialising in both invasive and non-invasive ventilation.

- ▶ 1,400 patients on a home mechanical ventilation contract
- ▶ 250 adult patients started on home mechanical ventilation per year
- ▶ 120 complex neuromuscular disease patients under active follow up
- ▶ 10 'difficult to wean' patients referred per month
- ▶ 88 tracheostomy ventilated patients in the community

The team at the centre is experienced in the discharging of complex ventilated patients with tracheostomies. The centre is a specialist home mechanical ventilation unit and provides the home ventilation support for these patients.

However, staff continually find it difficult to discharge these patients in a timely manner. These delays are leading to a loss in bed capacity and increased costs to the National Health Service.

The team at the unit believe that to improve the knowledge and care coordination of these complex respiratory tracheostomy ventilated patients, the clinicians and the CCG commissioners need to work closely, and that this holds the key to resolving the problem.

We understand that clinicians from the Lane Fox Unit have been working with NHS England to reduce delays in discharge from specialist centres.

Unless these delays are addressed, bed capacity will continue to be lost and the strain and stress placed on patients and families, already living with life limiting conditions, will remain.

The process must be streamlined.

Muscular Dystrophy UK believes that Health Authorities across the UK must introduce targets with clear timescales for the discharge of complex respiratory patients. Throughout the discharge process, there must be close liaison with the patient's specialist centre, who we believe are well placed to provide care agencies and community health professionals with training and advice.

## Ventilator power failure

Power cuts present an emergency situation for people reliant on ventilation in order to breathe. Without any back-up power supply for their vital equipment, individuals are placed at serious risk. There have even been tragic instances where people have lost their lives when power to ventilation equipment was cut.

**We call on NHS England and relevant Health Authorities in Scotland, Wales and Northern Ireland to ensure that equipment reliant on electricity, such as ventilator machines, have battery back-up. Individuals should also be given grants to purchase generators that switch over automatically, so they can continue using vital equipment in the event of power failures. Carers must also be fully trained in manual breathing assistance techniques for times when there is no access to a power supply.**

Muscular Dystrophy UK Trailblazer, Michaela Hollywood, has written a powerful blog post<sup>8</sup> on this issue:

I'm blogging from the middle of the ocean on board the Independence of the Seas. The irony is that even to get me here has been a military operation. I have one huge suitcase full of medical equipment in my room – and that's just the stuff not currently in use. I have two ventilators, a drip to feed me, a cough assist and a mattress with me as well as my own hoist and shower and toileting chair.

Anyone who has been on holiday knows you only bring essentials. That's exactly what we have done.

At the end of last summer we became aware of a grant that had funded a generator which would kick in automatically during a power cut. During bad weather we can be out for almost 3 days. I can't use the toilet, I can't be washed or dressed and all my carers aren't allowed to even so much as knock the door. If one is in the house when the power cuts they must leave immediately.

But that's if power cuts during the day.

At night the story is much worse.

My life support machine cuts out, taking my breath away and potentially causes a respiratory arrest or a life threatening infection. The battery that everyone keeps saying exists doesn't – because I'm under doctors orders to not use my machine without humidification, which doesn't run on battery. My mattress deflates in a matter of 20 seconds, leaving me on a metal frame. I have no access to my cough assist, allowing my lungs to drown in themselves. Carers leave immediately, and due to my bad sleeping pattern the vast majority of my caring hours which give mum and dad a break, happen at night.

This leaves me on a metal frame, unable to breathe or move at all, completely deaf and often unable to see because of dry eyes – and alone in the dark with no way of calling for help other than shouting, which is something I don't have the lung capacity to do.

Tonight when you go to bed, close your eyes and don't move. Just breathe for thirty minutes. Imagine what it feels like to have to be alone and not be able to move or get help in a situation where the power cuts. And then add in not being able to breathe.

This is a situation which has caused an infection which could kill me in the past. It has the capacity to kill me faster.

Tonight I heard that two young men in much the same position as me passed away after power cuts in Australia. You can read about them here: <http://www.news.com.au/national/western-australia/beaconsfield-housemates-and-muscular-dystrophy-sufferers-conor-murphy-and-kyle-scolari-die-after-storm-cuts-power-to-vital-medical-equipment/story-fnii5thn-1226989470874#3YearsDH2Film>

I have been working with the fantastic Seán Rogers MLA and Cllrs Maria McCarthy and Laura Devlin from the SDLP to work with Health Minister Edwin Poots to have me provided with an automatic switchover generator which costs £5,500 – including weatherproofing and soundproofing.

The South Eastern Health and Social Care Trust won't provide me with a grant – the same grant that was provided to another person just a few miles from my home who doesn't rely on a ventilator. I don't qualify because I'm on contributory Employment and Support Allowance (ESA) instead of income-based ESA.

The South Eastern Trust have a different plan. They want to give me an ambulance transfer – who we now know can't carry all my essential equipment. They then want to admit me to hospital – which they keep reminding me is perfect for me because it's a neurological specialist hospital. I'm a NEUROMUSCULAR patient, which is a stark contrast to someone with epilepsy which IS neurological. In that hospital they will expose me to life-threatening infections like pseudomonas which has already wreaked havoc on my lungs, and influenza which killed my sister. Not only that, but they will expose me to antibiotics that all have potential to give me a life threatening allergic reaction. And all this against what my doctors want to do – keep me at home. I stay at home even at my sickest on hospital at home, so why should I go to hospital and be institutionalised when I'm well?

Technology exists that has saved my life for decades now. More so, technology exists that can sustain that technology for several days with power in a cut. That technology – an automatic switchover generator – is being denied to me. Not only does that leave me with no safety while I wait on an ambulance which could take several hours or more, but it also means that when mum and dad are gone I will have no safety net and I will be completely alone.

## Summary

Vital respiratory support, such as cough assist machines or access to a respiratory physiotherapist, should not be dependent on the area of the country in which an individual happens to live.

This support enhances an individual's quality and length of life and the NHS in all regions of the UK has an obligation to provide it.

Muscular Dystrophy UK is committed to working with patients, families, healthcare professionals and the NHS to address the serious gaps in care that this report highlights.

## 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 respiratory care, please get in touch with Peter Sutton on [p.sutton@muscular dystrophyuk.org](mailto:p.sutton@muscular dystrophyuk.org) or call 020 7803 4838.

If you are struggling to access the respiratory care and equipment you need, such as a cough assist machine, our advocacy team is here to help you. Please get in touch with them at [info@muscular dystrophyuk.org](mailto:info@muscular dystrophyuk.org) or call 020 7803 4808.

## What are muscle-wasting conditions?

There are about 60 types of muscle-wasting 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-wasting conditions can be genetic or acquired and, with few exceptions, there are currently no effective treatments or cures available.

Clinical trials for some muscle-wasting conditions 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 (formerly Muscular Dystrophy Campaign) 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.

We know we can beat muscle-wasting conditions more quickly by working together. Please join us.

## Appendix 1

### Evidence for use of mechanical insufflation / exsufflation (cough assist machine)

East Midlands Regional Neuromuscular Respiratory Network Group Oct 2013 (reproduced with permission of Nicola McNarry, Clinical Specialist Physiotherapist)

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

#### Recommendations

- ▶ Children with ineffective cough (including children over 12 years of age with cough peak flow <270 l/min), particularly if they have had episodes of deterioration with respiratory infection, should be taught augmented cough techniques
- ▶ Mechanical insufflation / exsufflation (cough assist machine) should be considered in very weak children, those with loss of bulbar function, and those who cannot cooperate with manual cough assist or air stacking or in whom these methods are not effective.
- ▶ Mechanical insufflation / exsufflation should be available in the acute setting in all hospitals that treat neuromuscular patients as an alternative method of airway clearance with the purpose of preventing deterioration and the need for intubation and mechanical ventilation.

#### Evidence statements

- ▶ Use of airway clearance techniques which increase cough peak flow, as part of a homecare treatment package which includes NIV, is associated with decreased hospital admission for respiratory infection and improved survival.

*Concise BTS / ACPRC guidelines; Physiotherapy management of the adult, medical, spontaneously breathing patient May 2009*

The use of strategies, such as manually assisted coughing or mechanical insufflation – exsufflation can be used to increase Peak Cough Flow to effective levels and enhance cough effectiveness and must be considered and introduced where indicated.

#### Recommendations

- ▶ Consider mechanical in-exsufflation as a treatment option in patients with bulbar muscle involvement who are unable to breath stack.
- ▶ Consider mechanical in-exsufflation for any patient who remains unable to increase peak cough flow to effective levels with other strategies.

*Management of Respiratory Insufficiency in MND / ALS patients; an evidence based review 2006 MND Association*

#### Recommendations

- ▶ Use non-invasive cough assist devices, if available, in patients with a weak cough who develop a lower respiratory tract infection.

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

### Annex B

P11; Shared outcomes

Specialised Neuromuscular Services will:

- ▶ improve the quality of neuromuscular services and patient experience
- ▶ reduce the number of unscheduled admissions and re-admissions to hospital
- ▶ reduce length of hospital stay, and thus occupied bed days
- ▶ reduce inequalities in health between those with neuromuscular conditions, both across the region and in other parts of the United Kingdom.

P17 under Respiratory Care:

- ▶ If a patient's coughing continues to be ineffective and PCF is around 160L/min / FVC<40%, cough assist machines should be introduced to increase PCF and thereby assist secretion clearance (Lancet 2009). This should also be taught and used in all neuromuscular patients with PCF < 270L/min who are undergoing anaesthetic / sedation, to prevent post-operative respiratory complications (Birnkranz 2007).

*Consensus Statement for Standard of Care in Spinal Muscular Atrophy 2007*

Consensus recommendations from pulmonary working group:

- ▶ chronic respiratory management includes providing methods for airway clearance, including mechanical insufflation-exsufflation or manual cough assist and non-invasive ventilator support
- ▶ acute respiratory illness management requires increased airway clearance and secretion management techniques using mechanical insufflation-exsufflation or manual cough assist, increased respiratory support (including non-invasive ventilation), nutrition and hydration management, and a low threshold to start antibiotics.

*Respiratory Care of the Patient with Duchenne Muscular Dystrophy. American Thoracic (ATS) Statement 2004*

- ▶ The committee strongly supports use of mechanical insufflation-exsufflation in patients with DMD.

Diagnosis and Management of Duchenne Muscular Dystrophy, part 2: implementation of multidisciplinary care 2009 – accredited by NICE as produced by the highest standards (2011)

Respiratory interventions indicated in patients with DMD:

**Step 1:** Volume recruitment / deep lung inflation technique

- ▶ Volume recruitment / deep lung inflation technique (by self-inflating manual ventilation bag or mechanical insufflation-exsufflation) when FVC <40% predicted.

**Step 2:** Manual and mechanically assisted cough techniques

Necessary when:

- ▶ respiratory infection present and baseline peak cough flow <270L/min
- ▶ baseline peak cough flow <160L/min or maximum expiratory pressure <40cm water
- ▶ baseline FVC<40% predicted or <1.25L in older teenager / adult

## Appendix 2

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 (0.9)	4 (1.8)
<b>Total</b>	<b>576</b>	<b>227</b>

Intervention / measure	All preventable admissions (%)	Preventable neuromuscular related admissions (%)
Surveillance of patient's condition	139 (26.5)	114 (29.9)
Access to neuromuscular services	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)
<b>Total</b>	<b>524</b>	<b>381</b>

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

## Appendix 3

Emergency admissions in the West Midlands (figures based on estimated costs of unplanned emergency admissions from *Invest to Save*, Muscular Dystrophy Campaign report, 2011)

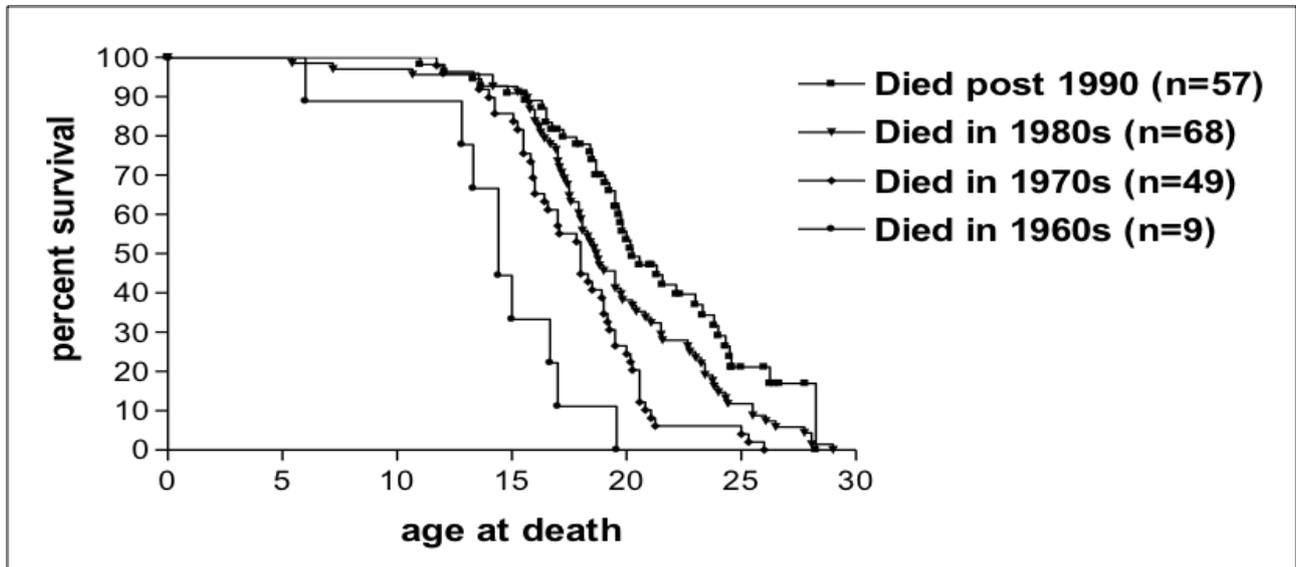
CCG Name	Sum of population	Estimated population with nm conditions	Estimated costs of unplanned emergency admissions	Potential savings on emergency admissions
NHS Birmingham <u>CrossCity</u> CCG	589300	638	£728,000	£291,000
NHS Birmingham South and Central CCG	242400	263	£300,000	£120,000
NHS <u>Cannock Chase</u> CCG	131200	142	£162,000	£65,000
NHS Coventry and Rugby CCG	461700	500	£571,000	£228,000
NHS Dudley CCG	313300	339	£387,000	£155,000
NHS East Staffordshire CCG	132700	144	£164,000	£66,000
NHS Herefordshire CCG	181500	197	£120,000	£48,000
NHS North East Birmingham CCG	131400	142	£285,000	£114,000
NHS North Staffordshire CCG	208700	226	£224,000	£90,000
NHS <u>Redditch</u> and Bromsgrove CCG	170800	185	£162,000	£65,000
NHS <u>Sandwell</u> and West Birmingham CCG	527800	572	£258,000	£103,000
NHS <u>Shropshire</u> CCG	296300	321	£211,000	£84,000
NHS <u>Solihull</u> CCG	234200	254	£150,000	£60,000
NHS South East Staffs and <u>Seisdon</u> Peninsular CCG	209600	227	£652,000	£261,000
NHS South Warwickshire CCG	270200	293	£366,000	£147,000
NHS South Worcestershire CCG	290800	315	£290,000	£116,000
NHS Stafford and Surrounds CCG	144000	156	£334,000	£134,000
NHS Stoke on Trent CCG	279600	303	£359,000	£144,000
NHS Telford & <u>Wrekin</u> CCG	170400	185	£649,000	£260,000
NHS <u>Walsall</u> CCG	269200	292	£346,000	£138,000
NHS Warwickshire North CCG	182600	198	£211,000	£84,000
NHS <u>Wolverhampton</u> CCG	258000	279	£199,000	£80,000
NHS <u>Wyre Forest</u> CCG	112100	121	£333,000	£133,000

## Appendix 4

### 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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