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**Men with Duchenne muscular dystrophy and end of life planning.**

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## **Abstract**

There is very limited evidence about the views of men with Duchenne muscular dystrophy (DMD) and end of life issues including death and dying. Studies have shown the physiological and psychological benefits of talking about and planning for end of life. Despite policy documents and guidance in the UK about end of life planning, there is consensus on the need for improvement. The study reported here is a qualitative one with 15 men with DMD (aged 20-45 years). Participants could not recall any significant conversations with clinicians about end of life and assumed that clinicians were reluctant to discuss the issue. The men in the study wanted to be given proactive cues that they could bring up topics such as death and dying and wanted to have these conversations with clinicians who combined expert knowledge about the condition as well as good listening skills. Topics of interest to participants included likely nature and place of death; practical planning for funerals and wills; and sources of information and support. Emotional or psychological support to think about end of life was not routinely offered and participants found it very difficult to discuss these issues with family members. The study suggests that more could be done to encourage clinicians, men with Duchenne, family members and the wider NMD community to pay attention to end of life planning issues and the associated need for emotional support and high quality interactions between patients and clinicians.

## **Keywords**

Duchenne muscular dystrophy; end of life; palliative care; death; neuromuscular disease.

## 1. Introduction

Duchenne muscular dystrophy (DMD) is an inherited neuromuscular disease which affects males (in about 1 in every 3500 UK births.) By the age of 13, boys with DMD have generally lost the ability to walk independently and by the mid-teens there are other complications: curvature of the spine, respiratory difficulties and cardiac failure. The mean age of death without specialised treatment was 19 but during the last decade there have been important improvements in the ways DMD is managed. In particular, teenagers who have been using nocturnal home ventilation to support their breathing can expect to live to around 27 years [1]. The mean age of death is continuing to rise as more effective medical interventions impact on the current generation of men who are reaching stages of adulthood that were, at the time of their childhood, largely unexpected and unplanned.

Recent social science research in the UK [2, 3] suggested that there were challenges associated with planning for and living the life of an 'unanticipated adult'. These included not routinely having the opportunity to talk about sensitive subjects such as sex, death, and living with the physical and psychological aspects of DMD. This was either because the topics were not brought up, or because men did not want to alarm or upset close family members. Concern about death and dying came into focus for example at a point of major medical intervention or significant illness or when friends with DMD died. Men with DMD may not have a predictable 'step down' in functional domains to warn them that the end of life is encroaching which reinforces the importance that these discussions happen in planned fashion early on in the disease course.

It is well documented that care of patients approaching the end of life in the UK is often less than ideal. In 2008 the Department of Health published its End of Life Care Strategy, a strategy aiming to promote 'high quality care for all adults at the end of life' [4]. How these documents should be implemented to improve end of life care for all was laid out in a series of 'Route to Success' documents [5]. The first stated aim of the End of Life Care Strategy was that it should improve culture around end of life care and death and dying by encouraging us all to talk about the issues around death and dying. The update of the 2008 strategy recognised that person centred care and decision making at end of life was still not universally good [6] and that an end of life care 'offer' should be, '...based on honest conversations with health and care staff which support the person to make informed choices.' A National Institute for Health Research review of the evidence on the organisation and quality of care in end of life services [7] concluded that improvements were needed in delivering the right care, at the right time and in the right place.

Temel and colleagues study [8] demonstrated the benefits of good end of life care. In a randomised controlled trial, patients with metastatic non-small cell lung cancer (and thus a poor prognosis) were randomised to receive standard oncological treatment with or without early referral, i.e. referral at diagnosis of malignancy, to palliative care services. Patients received monthly palliative care outpatient review. Quality of life was statistically significantly better in patients receiving palliative care. Also these patients had fewer depressive symptoms and received less aggressive end-of-life care. Patients referred to palliative care at diagnosis survived for longer than those who were referred to palliative care 'as usual', with a median survival of 11.6 vs 8.9 months. This was the first ever study to show that palliative care might improve survival as well as improving patients' quality of life.

The literature around end of life care in neuromuscular disorders (NMD) is not extensive. In one retrospective case note review of 24 men and one symptomatic female carrier referred to an adult neuromuscular clinic over a 7-year period, nine patients died during the observation period [9]. The paper noted that 'patients and their families were aware of disease progression and usually understood the natural history of the condition.

Nonetheless, and understandably, patients rarely wished to discuss the later stages of their illness. Death was often more precipitate than expected.' There was no justification in the paper of why this was 'understandable' or recognition that such an approach might lead to 'precipitate' deaths.

A questionnaire survey sent to families of males with DMD in the USA enquired about their knowledge of and involvement with palliative care services [10]. 34 families responded. Of these, 85% had never heard the term palliative care. 50% had received specialist skilled nursing services and 18% respite care. 12% had received pain management and 6% reported receiving hospice care. The authors concluded that knowledge about palliative care was low, most families were not using services commonly associated with palliative care and there was little evidence of the use of documents to direct care approaching the end of life. In an Australian study involving 11 family carers of men with Duchenne and 5 with other forms of muscular dystrophy [11], family carers reported gaps in services which would offer support in the later stages of life. Health professionals who were invited to reflect on the research messages acknowledged that they had difficulty in discussing death and dying.

The increasing technological opportunities for care for patients with NMD, particularly with regard to ventilation, can delay their referral for palliative care [12]. Because active management options are possible, this tends to delay conversations about the ultimately terminal nature of these disorders. Sadly the active/palliative care interface is still viewed by many as very black and white, tending to relegate palliative care and the professionals who offer it as being labelled as the 'last resort' when there is nothing further 'active' to be done. This is unfortunate, since palliative care alongside active management might well enhance quality of life as the Temel paper suggests. The American Thoracic Society consensus statement [13] on respiratory care of patients with DMD suggests that patients and families need education so they can understand and make informed decisions about end of life care.

The study described in this paper was informed by a scoping and feasibility study carried out by the authors [14] which highlighted the paucity of evidence in the field of end of life planning and NMD. This study aimed to focus on the views and preferences of men with DMD and how they could best be supported to approach end of life planning. Our hypothesis was that despite it being a sensitive topic to address, men with DMD may want to engage with their concerns and knowledge gaps about end of life. As mentioned, this hypothesis was borne out to an extent by data in social science studies with men with Duchenne carried out by one of the authors and colleagues [2, 3]. Men in these studies said that they tended to not think about living with DMD very much in order to stay positive. However, they also felt that they did not want to ignore it and said they would value the chance to talk to people whom they trusted about it.

What did we mean by 'end of life' in this study? We were aware of the differences between and interchangeable use of the terms palliative care, advanced care planning and end of life care but were not sure that these differences would be meaningful to research participants (or indeed much of the general public). We therefore used the term 'end of life' with little additional description and told research participants that we were interested in their views on 'end of life care issues, conversations and decision making.' This elicited interesting conversations which spanned the whole possible range of topics including death, dying, options around advanced medical interventions, assisted suicide, wishes about funerals and planning and so on.

## **2. Methods**

This was a qualitative study conducted with 15 men with DMD in the UK. A semi-structured topic guide was developed having reviewed other data collection tools in research with young people with life limiting conditions about death and dying [15], having conducted a review of relevant literature and in conversation with project advisors (made up of local clinicians and men with DMD from DMD Pathfinders). It was piloted and amended in the light of feedback and discussion. The study was supported by DMD Pathfinders (a user led organisation of men with DMD) who advertised for participants on their closed Facebook page. Those who expressed an interest were given the researchers e-mail address and we subsequently arranged to meet on a mutually convenient date. Interviews were carried out in the participant's homes. Interviews lasted, on average for 75 minutes and participants were given a £20 gift voucher for taking part. Research data was digitally recorded (with permission), transcribed and then analysed using established qualitative thematic analysis



techniques. The data was examined using the constant comparative method to test emerging hypotheses [16]. The two principal researchers met to discuss transcripts and the data coding which they both carried out. Both interviewers were experienced at carrying out 'sensitive' qualitative research and were mindful that a methods review on end of life care research [17] had concluded that despite the challenges of undertaking research in this area, people often valued the opportunity to participate in and find benefit from their involvement in research. The study was given ethical approval by a University of Bristol ethics committee.

### **3. Findings**

#### **3.1. What end of life conversations have men had?**

None of the men in our study could recall discussion about end of life with any clinician in children's services. In adulthood none of the men said that they had ever had a conversation about end of life with a medical professional. Most had experienced conversations initiated largely by respiratory consultants which were solely about views on resuscitation i.e. in what circumstances would the person want to be resuscitated, or not. By and large, men were pleased that the topic had been covered:

It was good to get it out of the way. You don't worry that someone isn't going to go along with your wishes. [R3]

But on its own, all of the men said that the conversation had been limited and overly medicalised and they did not view these exchanges as tantamount to a conversation about end of life. Conversations about resuscitation were regarded as important for the reasons stated above but only a small part of the picture. R12 felt this way, as well as feeling that the conversation about resuscitation itself had not been handled very well:

From the medical point of view they just say how it is. They don't think about the social stuff. My respiratory consultant is quite brutal. Talks about medical stuff but doesn't talk about end of life. [R12]

R1 said that the conversation about resuscitation raised by his respiratory consultant *had* been handled well describing her approach as, 'respectful', but in common with others did not equate this to a conversation about, '...what it will be like to be in the last stages of dying.'

They need to give you a route to talk about it from an emotional perspective as well, more than just the medical standpoint. Maybe tell people where they can go if they want to have those emotional conversations. [R1]

Most men highlighted the positive reasons to address the topic of end of life:

Cos to me death is upsetting but it's a natural thing. It comes down to having to accept it. You may even prolong your life a bit more if you can accept it and talk about it. You know, if you can get rid of your hurt and anger and depression about it. You can get on

and have a more fulfilling life. I used to be scared and I used to get a lot of physical symptoms like stomach issues and headaches and I couldn't breathe. Once I realised it was out of fear and anger and stress I kind of had to release that to improve my life. [R9]

In commenting on the absence of clinicians proactively talking about end of life issues, most men assumed that clinicians were simply too anxious to bring it up. R4 highlighted the fact that whilst other people might be anxious about the topic, he had talked with other friends with DMD at school about death:

They're frightened to talk about it and yet at 12, 13, we would talk about it at school, so I don't know why there's this hang up because we talk about it anyway. [R4]

With the health clinicians I've not really talked to them about dying. Doctors say a lot when you're diagnosed – you won't be able to walk, you'll have to go into a wheelchair – then they suddenly stop when you come to death. It's odd really because they're leading you to the door of the conversation but they're keeping it closed which makes it more scary really. If a doctor can't talk about it and you don't have a close relationship with your parents when it comes to these type of issues then who you gonna call? [R9]

In two instances, workers from a children's hospice had come to the family home to talk about, amongst other things, end of life planning and death and dying. R8 was 18 when this happened. R9 also worked with someone from a local children's hospice to create a document of wishes:

She's coming to talk about some of my plans for end of life. All this helps me because I didn't want my family to worry when I die. Doing it eases the pressure on me because I've taken responsibility for it.

Some men mentioned that they felt open conversations about end of life seemed more common and routine amongst other patient groups:

You watch programmes and they talk a lot about end of life, like cancer and Alzheimer's and that sort of thing. I've never heard any of that for MD. [R10]

### **3.2. What do men want to know about the progression of DMD, end of life, death and dying?**

Most men had been told about the progression of Duchenne by a combination of doctors and parents as they were growing up. Some recounted finding out that they were likely to die younger than the norm from friends or the internet. What men wanted to know about in adulthood varied to an extent, but a common factor were questions about what death would be like to which no-one had been given an answer. To be clear this was not men asking about what happened after death but about what the very last stages of life would be like:

How does life tend to be when you get near to the end? I've asked that and they say, 'Don't know. Can't tell you.' Which doesn't really help. [R2]

All you really want to know is, like, is it painless, or you're asleep or something? Or would you suffer? [R3]

It's specific things you want to know about what is the last stage going to look like? Are you going to be gasping for breath and in pain and all sorts of horrible things? Or will it be potentially peaceful? [R1]

If someone dies suddenly, no-one ever speaks about it... but you want to know how they died. You want to know was it the MD, the heart, the lungs? You can't ask but you'd quite like to know. [R10]

Three men spoke about assisted suicide. R4 was keen to talk about this in the interview saying that he tried to talk about it with carers, had thought about the most suitable methods and thought that it should not be off limits as a subject of conversation. To be clear, R4 was a very positive man but he wanted to discuss possible scenarios in which he may have found being alive to be much less tolerable. R5 was slightly less categorical in his view reflecting that the debates about assisted suicide needed contextualising in the overall lack of good support for disabled people in everyday life.

### **3.3. Practicalities such as funerals and wills.**

None of the men said that they had yet made wills or written anything formal about their wishes about place of death, funeral arrangements , but all of them had thought about it.

One of the main impediments to recording their wishes was that they felt they would want

some support to do it, but did not know who could help. One of the things that worried some of the men was that their wishes for settling their personal affairs after death would not be known. :

One thing that worries me is that people might not understand what I really want. I sometimes think about doing a will but I haven't got round to it. [R3]

DA2 wanted a non-religious funeral with his favourite music played and for his life to be celebrated by family and friends. He did not feel he could bring this up with his family, he thought he would need some support, and he thought his views would have to be captured in a formal document. The 'right' music choices for their funeral mattered to most of the men.

I quite like the idea of, 'Who Wants to Live Forever'. Sort of sarcastic. But I change my mind all the time. I'm more concerned about the after show party. Put my I-pod on random! [R4]

Those who discussed a preference for place of death said they would rather die at home or in a hospice than in a hospital if possible and cited reasons such as dignity, comfort and having space for family and friends to come and visit. A minority of the men had used hospice provision and were positive about it, 'It's like a second sort of home and the staff are wonderful there.' [R2]

R3 voiced a concern shared with others which was how would other people cope with his death. This was actually his primary concern:

The biggest thing I worry about is what happens to everyone else. It sounds weird but it's like if you die you're letting them down or something. I worry that they will be upset.

[R3]

R5 also talked about his uncertainty about some of the practical issues that would follow on from his death:

How's all your stuff taken care of afterwards? What happens to your staff, your house? Do your family do the right things? Because that's my biggest fear about death. It's not the fact of me not being here, it's the issue of how do people carry on when I'm not.

[R5]

### **3.4. How do these issues get talked about amongst family and friends?**

None of the men bar one said that they were talking openly with their families about end of life issues including death, and for everyone this was for fear of causing upset.

When you're talking about what will happen when you're at death's door, that's kind of hard to talk about with my mum, dad, sister and that. [R7]

Some had tried to broach the topic with parents but none recounted particularly successful experiences: 'It hasn't always been smooth talking to my parents', said R13. When R12 was told by a friend that he had a life limiting condition at age 14 he went to talk to his mum:

I came home and I spoke to my mum. I asked her if I had a short life. She said yes but they didn't want me to think about dying, they just wanted me to be positive about life.

[R12]

In one interview it was suggested that parents might also find it cathartic to talk and discuss the issues in the open:

They probably don't realise we would want to have those conversations. Really everybody wants the same thing but it's that initial barrier, getting over that really. [R1]

None of the men mentioned talking to friends who did not have DMD about end of life. The general view was that only other people with DMD would understand. R12 said that when he tried to talk to friends they would just tell him to 'be positive' which actually seemed like an unhelpful way of closing down the conversation. All said that they valued social media and the internet as a way of finding out information about Duchenne and sharing thoughts about end of life. On-line, some men said they took part in or read conversations about death and dying:



I always think it's easier to talk to lads with MD because they're in the same boat and they understand. There's always dark humour and stuff. Yeah we do talk about ourselves and stuff like end of life. [R3]

### **3.5. Is emotional support available?**

In common with previous studies, effective social work input was rare and no one reported ongoing contact with social services. All of the men talked about being depressed and/or anxious at various points in their adult lives. Most had accessed counselling or psychology services. These had not routinely proved useful in exploring issues about DMD and end of life. Several men commented on the unavailability (and physical inaccessibility) of counselling and that it was rarely offered. R2 had a psychologist who came to the house, '...every so often', but despite the fact that he wanted to talk about end of life issues, he had never brought it up:

I wouldn't like to bring it up with him. They would have to bring it up really for me to talk about it. Sometimes you just need a little prompt that'll help you bring it up. And then it's like an opening, which is good. [R2]

In fact R2 went on to say that it was in part the absence of understanding about the end stages of life which caused his anxiety:

I was getting a bit anxious and stuff at the time. And I thought that knowing would help, help calm me down a bit. But they couldn't really tell me anything to be honest. [R2]

R1 said that he had helpful discussions with a counsellor about death and dying:

I found it really useful to work through some stuff about how I felt about death and dying and to have someone to talk to that wasn't my parents. [R1]

But in common with others he also felt that the conversations left quite a lot unresolved because the counsellor lacked clinical knowledge and couldn't answer questions about end of life:

You don't just want someone who's going to be a passive listener, you want them to have some kind of insight into what you're talking about... they're not giving anything back because they have no idea about Duchenne. [R1]

### **3.6. What would good end of life conversations look like?**

There was a lot of consensus about this. What characterised a good overall approach was giving people cues that they could discuss any topic relating to end of life, life expectancy, death and dying. Somebody else 'bringing it up' was the most commonly expressed preference. That said, R5 noted that perhaps there was a learned history of passivity amongst men with DMD in terms of interactions with clinicians which could usefully be broken:

Guys should feel more confident about talking about their own issues without their parents being there. It's almost like sometimes they're waiting on permission to talk about things rather than realising they can bring it up. [R5]

Men wanted the person offering conversations to be personable. Personality counted for a great deal and this, coupled with expert knowledge i.e. being able to answer questions, outweighed any preference for the person being any particular *type* of clinician or professional.

It's more about the personal qualities, approachable kind of thing, in a gentle way, so it's not harsh. Approach it more like you would with your own family rather than your medical head. It needs to be more human. [R2]

It was perhaps this mix of people skills and clinical knowledge in one person that was thought to largely missing with R1 describing what he thought was the right mix of traits in an as yet undiscovered professional:

You want someone with expert knowledge about the condition and what might be involved, and they can answer your questions with authority, but they can also be supportive as well. [R1]

R9 cited these kind of traits in his respiratory consultant who he said he would feel comfortable discussing end of life with, in contrast to other clinicians he had met:

You feel that he is actually listening. Whereas other doctors I see, you feel that they're not emotionally invested in you. He asks about my life and that's the difference and it means you don't mind talking about anything really. If you can talk about how you live then you can talk about dying. It's difficult to talk about your death to someone who may not be that interested in your life.

In addition, they wanted whoever was offering the conversation to recognise that they may or may not actually want to talk at that point in time but they said they would not be phased by someone bringing it up – 'if you don't want to have the conversation you can just stop' [R11].

Views varied a little more on the best time to initiate conversations and most men settled on suggesting that everyone was different and it would need to be worked out on a person by person basis:

Everyone's different. You sort of have to judge it. Or simply just ask. [R2]

R14 felt that having conversations about future possible scenarios were best had in advance when you were still well, '...better to have conversations when you're stable so that nothing is a surprise if you get ill.' R11 was one of the few men who thought conversations should happen earlier in children's services and as a way of informing transition processes. Most men, as discussed above, said that there would be times in life that they would welcome a conversation but times they would not.

My perspective has varied over time. So I don't think there is just one approach. It's about being open to those conversations and giving people the route if they do want to talk about them. [R1]

Whether or not these conversations happened with and in front of parents was a matter of differing opinions. Some who were more open with their parents said that they should be involved, others were more reticent:

End of life conversations should happen but I don't want to ask about stuff in front of my mum, I don't want to upset her. [R14]

R10 suggested that clinicians should try and offer some time alone without parents so that the subject could be broached:

I don't like to bring it up cos I'm mostly with my mum at appointments. But maybe they could say, 'Would you mind stepping out while we speak...', kind of thing.

R4 in common with others suggested that the resilience of men with DMD should not be under-estimated when it came to so-called difficult conversations:

Just bring it up and talk about it. I think we're stronger than they think we are. We deal with lots. Death's just another thing. If you get it out of the way you can put it to bed.  
[R4]

#### 4. Discussion

Our first reflection is to express some surprise that none of the men in the study – a generally ‘older’ cohort of men with Duchenne – could recount an end of life conversation. Of course it is possible that some of their clinicians may have felt that such a conversation had been had in relation to e.g. resuscitation. This may point to a mismatch in understanding what constitutes an, ‘in the round’ end of life conversation which might encompass much more than decisions about end stage medical interventions.

Our study has as mentioned, an obvious bias in that all of the participants were ready and willing to talk about end of life. We may well have not been exposed to men with DMD who had good experiences of end of life planning or who were strongly of a view that this topic should never be broached. Two issues persuade us that the data remains valuable. Firstly, our participants could all recall a time when they would not have been interested in taking part in research with this focus or talking about end of life. It suggests to us that it would not be atypical for most men with Duchenne to have times in their life when they would and they would not want to talk about end of life. The message from this research is that, given positive cues, men will decide at what point the conversation is welcomed. Secondly, we were struck by the homogeneity of the messages from participants and had, even at 15 interviews, reached data saturation. We were particularly struck by:

1. The need to be given *proactive* messages and cues that the topic of end of life could be raised and the sense that clinicians might be reluctant to do this.

2. The potential benefits to mental and physical health of having the opportunity to talk about end of life, death and dying.
3. The desire to know more about the possible causes of death and the management of end of life in terms of support and pain control and for clinicians to either initiate these conversations or give cues that the topic is not 'off-limits'.
4. The need for support (practical and emotional) to think about setting down wishes for things such as funerals, place of death, wills etc.
5. Concerns about discussing these issues with parents and an over-riding worry about how loved ones would manage in the aftermath of their death.
6. The general absence of effective emotional and psychological support.
7. The preference for end of life conversations to be shared with people who were personable, knowledgeable and sensitive.

The study has raised issues regarding the lack of integrated care across agencies as well as a potential lack of training which may have contributed to the ability and confidence in key professionals' under-taking this task. The key messages from the research point to changes that could usefully be made in the provision of services (investment in mental health support); in the nature and quality of interactions between patients and clinicians; in training opportunities to educate clinicians in the importance of embarking on good end of life conversations. Publicising some of the powerful and unassailable narratives from the men in our study may also prove useful in empowering men to speak up; give family members a greater sense of permission in discussing end of life; and encourage the wider NMD community of clinicians and support organisations to give this topic greater attention.

## **5. Concluding remarks**

Finally, a point made by many of the men in this study was that uppermost in their minds was leading a good life and getting the right support. “Let's talk about end of life then get on with living a good life”, is a useful paraphrase of this view (and one echoed by other groups of young people with ‘life-limiting’ conditions [15, 18]). To talk about end of life issues within a positive frame of reference was thought to be important:

People need to be built up in positive ways. It's like, yes, people need to have these conversations so that you can move on and actually get on with your life. The focus should be on living with Duchenne and having a good life with it. And then obviously as part of that you need to think about death and dying and be able to get into a position where you can move on from it. That's the only way it's cathartic, if you can then move on afterwards. [R1]

Our study suggests that it is possible to do both things i.e. focus on a good life but, with support and when the time is right, think about the end of life in ways which are beneficial:

I think the first step isn't talking about dying, it's to talk about living and how you live. And then when you reveal the way you live your life, it can show you how you may want to die. [R10]

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## References

- [1] Eagle M, Bourke J, Bullock R, Gibson M, Mehtaa J, Giddings D et al.(2007) Managing Duchenne Muscular Dystrophy: The Additive Effect of Spinal Surgery and Home Nocturnal Ventilation in Improving Survival. *Neuromuscular Disorders* 2007; 17: 470–475.
- [2] Abbott D, Carpenter J. ‘Wasting precious time’: young men with Duchenne muscular dystrophy negotiate the transition to adulthood. *Disability & Society* 2014; 29: 1192-1205.
- [3] Abbott D, Carpenter J, Bushby K. Transition to adulthood for young men with Duchenne muscular dystrophy: research from the UK. *Neuromuscular Disorders* 2012; 22: 445-46.
- [4] Department of Health. The end of life care strategy. London: Department of Health; 2008.
- [5] Department of Health. National end of life care programme. London: Department of Health; 2010.
- [6] Choice in End of Life Programme Board. What’s important to me: a review of choice in end of life care. London: End of Life Programme Board; 2015.
- [7] National Institute for Health Research. Better endings. Southampton: NIHR Dissemination Centre; 2015.
- [8] Temel JS, Greer JA, Muzikansky A, Gallagher ER, Admane S, Jackson VA et al. Early palliative care for patients with metastatic non–small-cell lung cancer. *New England Journal of Medicine* 2010; 363: 733-42.

- [9] Parker AE, Robb SA, Chambers J, Davidson AC, Evans K, O'Dowd J et al. Analysis of an adult Duchenne muscular dystrophy population. *QJM* 2005; 98: 729-36.
- [10] Arias R, Andrews J, Pandya S, Pettit K, Trout C, Apkon S et al. Palliative care services in families of males with Duchenne muscular dystrophy. *Muscle & Nerve* 2011; 44: 93-101.
- [11] Dawson S, Janson LJ. Mapping the journey: family carers' perceptions of issues related to end-stage care of individuals with muscular dystrophy or motor neurone disease. *Journal of Palliative Care* 2003; 19: 36-42.
- [12] Fraser LK, Aldridge J, Manning S, O'Leary S, Miller M, McCulloch R et al. Hospice provision and usage amongst young people with neuromuscular disease in the United Kingdom. *European Journal of Paediatric Neurology* 2011; 15: 326-30.
- [13] American Thoracic Society Consensus Statement. Respiratory care of the patient with Duchenne muscular dystrophy. *American Journal of Respiratory and Critical Care Medicine* 2004; 170: 456-65.
- [14] Abbott D, Prescott H, Forbes K, Majumdar, A. Exploring end of life planning issues for men with Duchenne muscular dystrophy: report of a scoping and feasibility study. Bristol: University of Bristol & UBHT; 2013.
- [15] National Council for Palliative Care. Difficult conversations: making it easier to talk about end of life issues with young adults with life-limiting conditions. London: National Council for Palliative Care; 2014.
- [16] Silverman D. Interpreting qualitative data. London: Sage; 2001.
- [17] Goodman C, Froggatt K, Mathie E. End of life care: methods review. London: NIHR School for Social Care Research; 2012.

[18] Together for Short Lives. A core care pathway for children with life limiting and life threatening conditions. Bristol: Together for Short Lives; 2013.