



National Hospice and Palliative Care Organization
Palliative Care Resource Series

'WHO YOU GONNA CALL?'
MEN WITH DUCHENNE MUSCULAR
DYSTROPHY DISCUSS END-OF-LIFE
PLANNING

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INTRODUCTION

A research study carried out in the UK in 2015-16 asked young men with Duchenne muscular dystrophy (DMD) about their views and experiences of having conversations about end-of-life planning, death and dying.

DMD is an inherited neuromuscular disease which affects mostly males and which leads to loss of mobility, curvature of the spine, respiratory difficulties and cardiac failure. In the UK the mean age of death without specialised treatment was 19 but during the last few years there have been significant improvements in the ways DMD is managed. Teenagers who have been using nocturnal home ventilation to support their breathing can expect to live to around 27 years (Eagle et al. 2007). The mean age of death is continuing to rise as more effective medical interventions impact the current generation of men who are reaching stages of adulthood that were, at the time of their childhood, largely unexpected and unplanned. Advances in medical technology have meant that a whole generation of men with DMD are alive, who simply were not expected to be.

The study outlined here came about in part after a national survey and round of qualitative interviews with boys and young men with DMD aged 15+, their family members and clinicians (Abbott & Carpenter, 2014, Abbott, Carpenter & Bushby, 2012). In this study, it became clear that open conversations about the nature of disease progression, prognosis, questions and concerns about death and dying were extremely difficult to have.

- Parents did not always know what their sons did or didn't know.
- Boys and young men were reluctant to ask questions for fear of upsetting family members.
- Clinicians sometimes found it difficult to bring the topic up – especially with the rapid changes in life expectancy.

This article focuses on end-of-life conversations with young and older men with DMD. Following is an excerpt from one interview which eloquently encapsulates important issues the study identified.

"I went to a friend's funeral and it makes you think about what you would like at end of life and how you would like it organised. I was thinking about my funeral, how to make it easier for my family when I go as well, cos if I didn't make any plan, then I die, then they wouldn't really know my wishes. But if I share what I want then it's a bit more stable for them really.

Cos to me death is upsetting but it's a natural thing. 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. I don't want to waste my life crying about the fact that I'm going to die. 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.

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?**

I think a good doctor who wanted to have these conversations would be like my respiratory consultant – he's really calm, he waits for you to say something but you feel that he's actually listening to what you're saying, that he's emotionally invested with you. If you can talk about how you live and the way 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.**"

The text above in bold focuses on a key message. Talking about death and dying can be regarded as taboo and research shows not everyone agrees that teens and men with DMD should be encouraged to think or talk about end of life (Abbott & Carpenter, 2010, chapter 6; Abbott, 2012; Gibson et al., 2009; Mitchell, 2012). However, the interviewee's response above highlights the importance of being given a choice; being proactively given a cue that it's okay to talk about it – or not; and that the key to a successful conversation will be the quality of the relationship between the clinician and the person with DMD. If no one makes time, space or gives permission for fears and concerns to be aired then, as our speaker says, "Who you gonna call?"

THE RESEARCH

This study focused on the views and preferences of men with DMD and how they could best be supported to approach end-of-life planning. The hypothesis was that despite it being a highly sensitive topic to address, men with DMD may want to engage with their fears, concerns and knowledge gaps about end of life. This hypothesis was borne out to a small extent by data in Abbott & Carpenter's study (2012, 2014) which focused on the experiences of transition to adulthood for young men with DMD. We know anecdotally from a clinical perspective that men with DMD may want to discuss this topic but at the same time 'protect' their families in a way which can prove challenging for clinicians, patients and family members to negotiate.

This was a small scale, qualitative study conducted with 15 men with DMD in the UK. The study was promoted by a user led organisation of teens and men with Duchenne called 'DMD Pathfinders' on their closed Facebook page and those who expressed an interest were interviewed in the participant's homes and with permission, digitally recorded. Research data was transcribed and then analysed using established qualitative thematic analysis. The study was given ethical approval by the University of Bristol ethics committee. Participants were between 20 and 45 years of age and were a mix of those living with family members and those living independently with their own support. Eleven participants identified as White British and 4 as Asian British. Further details about the study can be found in Abbott et al., 2017.

KEY MESSAGES

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 adult health services. Most had experienced conversations initiated largely by respiratory clinicians which were solely about views on resuscitation i.e. in what circumstances would the person want to be resuscitated, or not. In the absence of more holistic discussions about end of life, most men did the thinking on their own from time to time. One man who was interested in what it would be like, "...when you get near to that time..." and was asked if he had those conversations with anyone, "Just with myself, really."

"Someone to share it with would make it a little easier because it's a lot to think about. You're not supposed to think about it until you're old, in your 70s isn't it. But you have to do it early. If there was someone else to talk to or just email or something and take a bit of that off you, that sort of help would be good. Some of it. Even just half of it. At least it's let out."

This is a biased sample of men who were willing to talk about end-of-life issues with us, so it is assumed that they felt there would be positive reasons to address the topic:

"It would lift the weight. You can just get on with it then. You don't want to dwell on it but you do want to get it out of the way."

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 up the issue.

"They must be afraid to ask those sort of questions, or don't want to offend or upset you. But me, I wouldn't mind any question. "

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

What men wanted to know about in adulthood varied to an extent, but a common factor was 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 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? How do you want things done? Those kind of things. I've asked that, about what it would be like in the end and they say, 'Don't know. Can't tell you.' Which doesn't really help.

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?"

Thinking about practicalities such as funerals and wills.

While all of the men had thought about it, none of the men said that they had made wills or written anything formal about their wishes about place of death or funeral arrangements. 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 they could ask or who could help. One of the things that worried some of the men was they would not be properly represented by their funerals – too religious or not religious enough, the wrong music, the right/wrong way to be buried, cremated etc.

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.”

How do these issues get talked about amongst family and friends?

Only one of the men said he talked openly with his family about end-of-life issues including death; for everyone else the lack of discussion was the fear of causing upset.

“I don't really talk about it with my parents because I don't think they want or like to hear it. It's quite a sensitive thing isn't it.”

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 one man.

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. There's probably a lot of misperception on both sides. Really everybody wants the same thing but it's that initial barrier, getting over that really.”

All of the men said that they valued social media, the internet, and Facebook 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. And sometimes you use it to vent which helps.”

What would good end-of-life conversations look like?

There was a lot of consensus about what a good end-of-life conversation would look like. 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.
- 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.
- 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'.

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. One man 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." Most men 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."

DISCUSSION

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.

Despite this being a small scale, qualitative study, the homogeneity of the messages from participants stuck out. In particular:

- the need to be given proactive messages and cues that the topic of end of life could be raised and the sense that many clinicians were reluctant to do this;
- the potential benefits to mental and physical health of having the opportunity to talk about end of life, death and dying;
- 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;
- the need for support – practical and emotional – to think about setting down wishes for things such as funerals, place of death, wills etc;
- concerns about discussing these issues with parents and an over-riding worry about how loved ones would manage in the aftermath of their death;
- the preference for end-of-life conversations to be shared with people who were personable, knowledgeable and sensitive.

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. For many men, inadequate support felt more life shortening than Duchenne itself.

"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."

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ADDITIONAL INFORMATION

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ACKNOWLEDGMENTS

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