



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.

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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. In other countries life

expectancy may be longer or shorter depending on the norms and quality of medical intervention and support.

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 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

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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 were 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 patients' quality of life.

The literature around end of life care in neuromuscular disorders (NMD) is not extensive and, for example, we could not find any studies which explored death and dying from the point of view of men with Duchenne themselves as opposed to other family members. 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 inquired 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, and 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'. These 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.

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