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Neuromuscular Disorders 27 (2017) 115–119

www.elsevier.com/locate/nmd

Advance care discussions with young people affected by life-limiting neuromuscular diseases: A systematic literature review and narrative synthesis

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Received 16 August 2016; received in revised form 10 October 2016; accepted 15 November 2016

Abstract

End of life care policy in the UK advocates open discussions between health professionals and patients as the end of life approaches. Despite well documented understanding of the progression of life-limiting neuromuscular diseases, the majority of patients affected by such conditions die without a formal end of life plan in place. We performed a systematic review to investigate conversations regarding end of life care between healthcare professionals and younger adult patients with life-limiting neuromuscular diseases. The search strategy included terms that focused on death and dying along with other factors that could impact length of life.

The review found a very limited body of literature regarding end of life care conversations between young people affected by neuromuscular diseases and health professionals. The views and preferences of patients themselves have not been investigated. There is a shared reluctance of patients, family carers and healthcare professionals to initiate end of life care discussions. There are many factors that need to be investigated further in order to develop a consensus that would allow healthcare professionals to engage patients in end of life care conversations allowing them to face the end of their lives with appropriate plans in place.

Keywords: Neuromuscular diseases; Death; Palliative care; End of life care; Advance care plans

1. Introduction

Neuromuscular disease (NMD) is an umbrella term for a wide range of genetic and acquired conditions that are caused by an abnormality at some point in either the lower motor neurone, i.e. from the anterior horn cell, through peripheral nerves and into the neuromuscular junction, or in muscle structures and systems [1]. Onset ranges from the neonatal period to late adulthood. Severity varies greatly: the most severe forms lead to premature death in infancy and childhood [2]; others are present in early childhood and progress steadily through to teenage years, with increasing physical weaknesses and death commonly from cardiorespiratory failure in the second or third decade of life [3].

Duchenne muscular dystrophy (DMD) is the most common form of life-limiting NMD, affecting 1:3600–6000 live male births [4], with a well-documented course of progression. DMD and other rarer forms of NMD currently have no cure. Recent advances in steroid medication and in particular the use of non-invasive ventilation have seen a rise in the average life expectancy from the mid/late teens in the 1960s to the mid-20s under current management regimes [5] with some men now surviving into their 30s and 40s. Other NMDs such as spinal muscular atrophy and Ullrich congenital myopathy have a less predictable course of progression but still have a significantly shortened life expectancy.

Unlike other life-limiting diseases such as cancer or amyotrophic lateral sclerosis, these NMDs are diagnosed in childhood, and the individual is largely unaffected by the condition initially. The progression of the disease can be slow, over a period of many years and parents often struggle to discuss disease progression with their children [6]. The terminal stages may not be considered until many years after the diagnosis has been made, and may not be addressed until unexpected life-threatening crises have occurred such as respiratory infection/arrest or cardiac event.

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http://dx.doi.org/10.1016/j.nmd.2016.11.011

0960-8966/

End of Life Care (EOLC) policy in the UK advocates open discussions between health professionals and patients as the end of life approaches [7]. Primarily developed to address the needs of cancer patients, it is increasingly acknowledged that EOLC policy may not meet the needs of those with longer-term and unpredictable life limiting conditions such as NMDs. The recent Marie Curie report concerning palliative care for non-cancer long term conditions excluded neuromuscular diseases as it focused on long-term conditions that primarily affect older people [8]. NHS policy describes EOLC as applying to the last year of life, whilst acknowledging that it may cover periods from a few hours to many years [9]. Within the specialist NMD literature, the palliative and EOLC needs of people with life limiting NMDs are only briefly mentioned in condition-specific standards of care [4].

We therefore undertook a systematic review of the literature concerning conversations about EOLC between young people with life-limiting neuromuscular diseases and healthcare professionals. We use the term EOLC conversations, Advance Care Planning and Advance Decisions interchangeably in this paper, according to the terms used in publications.

1.1. Review questions

With regard to conversations between teenagers and young adults with life-limiting NMDs and healthcare professionals concerning care at the end of life:

1) Are these conversations taking place?
2) What are the preferences of NMD patients and healthcare professionals for the timing and content of these conversations?
3) What are the barriers and facilitators to these conversations?

2. Methods

A search of Medline and Embase via OvidSP and Psycinfo via EbscoHOST was undertaken with the support of a professional librarian (IK). Inclusion criteria were: publication in peer reviewed journal between 1987 and January 2015, written in English and concerning life-limiting NMDs affecting people under the age of 25. Exclusion criteria were: papers with no new empirical data (including summaries of professional meetings and opinion pieces), studies solely concerned with amyotrophic lateral sclerosis or conditions with a likely prognosis of death within the first decade of life.

The term "neuromuscular disease" incorporates a wide range of conditions, which may present with a number of complications or progressions, with varying predictability and often uncertain implications in terms of end of life events. This made creation of the search strategy challenging, since discussions concerning EOLC encompass both the period when dying is becoming increasingly likely and any disease progression associated with a potentially adverse prognosis. The search strategy therefore included terms that focused on death and dying, with others that had potential impact on length of life such as: surgery for scoliosis, insertion of gastrostomy, commencement of non-invasive ventilation or the loss of ambulation. Such events may be seen as strong reminders of the progressive and eventually terminal nature of the diagnosis and thus may lead to discussions concerning EOLC. The search strategy as run in Medline is shown in Appendix A.

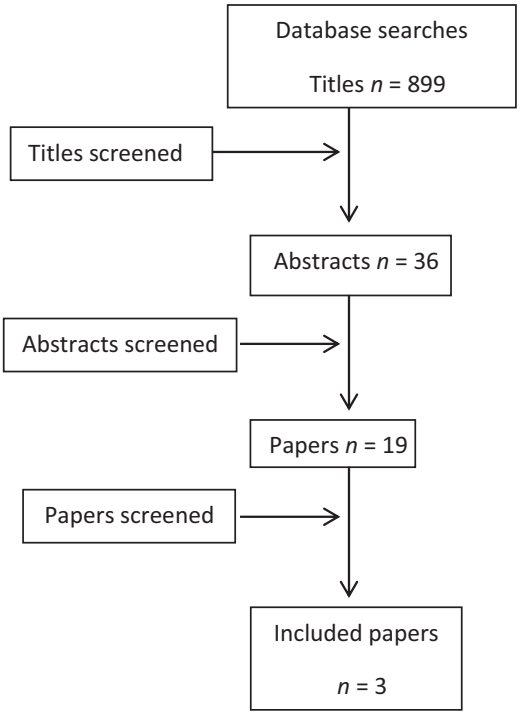

Fig. 1. Flowchart of article screening.

The initial search identified 899 titles that were screened by one researcher (AH) to exclude articles that were clearly not relevant. Two researchers (AH and SB) then reviewed the abstracts of 36 papers, from which 19 full papers were independently assessed by AH and SB: three papers met review inclusion criteria (Fig. 1). These were then independently weighted for relevance to the review questions using Gough's "Weight of Evidence" framework (Appendix B) [10]. Hand searches of four journals (Neuromuscular Disorders, European Journal of Palliative Care, Palliative Medicine, International Journal of Palliative Nursing) and citation searches of included papers identified no additional papers.

3. Results

All three papers report interviews with parents/lay carers of young people with DMD or spinal muscular atrophy. Only one [11] reports interviews with health professionals and patients. One specifically addressed the role of palliative care [12]: two included EOLC within an overview of family carers' needs and experiences rather than that of the patients towards the end of life [6,11]. Two were evaluated as "high" and one as "medium" on Gough's criteria (Appendix C) [10].

1. Are conversations taking place?

No paper gave any evidence of conversations taking place between the patient and healthcare professionals. One study [12] found one patient out of 13 had a written advance directive but gives no information of the circumstances in which that was