

Swiss physicians' practices of long-term mechanical ventilatory support of patients with Duchenne Muscular Dystrophy

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Summary

Introduction: Previous studies have shown that long-term ventilation (LTV) is still not routinely offered to or discussed with patients with Duchenne Muscular Dystrophy (DMD). This is in contrast to the increasing evidence that these interventions can successfully improve quality of life and prolong survival of affected individuals.

Aim: The aim of this survey was to describe the clinical practice and the attitudes of Swiss physicians in the respiratory follow up of DMD individuals and to understand better the current provision of respiratory services in Switzerland. Another aim was to explore the contentment of the patients about the provision of information on LTV.

Methods: Postal questionnaires requesting information on the current practice of respiratory care were sent to physicians, including pulmonologists and neurologists known or suspected to be involved in the care of children and adults with DMD. 26 of 30 (87%) physicians returned the questionnaire. A second questionnaire was sent to 90 DMD patients of whom 43 (48%) returned the questionnaire.

Results: 88% of the physicians indicated to discuss the option of LTV with their patients and families, but 50% of the surveyed physicians start such discussions only after the patient has already developed respiratory failure. Regular pulmonary function assessments are performed by the majority physicians in DMD patients, but only 50% of the physicians perform regular sleep studies to detect sleep disordered breathing.

Conclusion: The majority of Swiss physicians involved in the care of DMD patients disclose information on LTV to DMD patients, but commonly late in the disease course after respiratory problems have developed. The current routine of respiratory follow-up of DMD patients seems insufficient, at least with respect to early detection of nocturnal hypoventilation.

Key words: mechanical ventilation; physician's beliefs; Duchenne Muscular Dystrophy

Introduction

Hypercapnic respiratory failure and cardiac failure are the most frequent fatal complications in Duchenne Muscular Dystrophy (DMD) [1–3]. The majority of DMD patients (73%) with a vital capacity below 20% will succumb to respiratory failure within a few months, if left untreated [4, 5]. Several factors are implicated in the deterioration of respiratory function: progressive loss in respiratory muscle strength, altered chest wall compliance and the development of scoliosis. In addition, ineffective cough clearance of secretions, swallowing dysfunction with recurrent aspirations and loss of the ability to take spontaneous sighs result in atelectasis and recurrent respiratory tract infections [6].

For many decades long-term invasive ventilation via tracheotomy has been a controversial

option for progressive neuromuscular disorders. Ventilatory support in a terminal stage might protract death rather than extend good quality of life [7]. Since the mid-seventies, non-invasive ventilation is increasingly used to support DMD patients at an early stage of the progression of respiratory failure [8–10]. Initially, there were fears that the use of non-invasive ventilation would progress to invasive tracheotomy ventilation, thereby leading

Abbreviations

DMD	Duchenne Muscular Dystrophy
LTV	Long-Term Ventilation
NIV	Non Invasive Ventilation
PFT	Pulmonary Function Tests

to ventilator entrapment. Recent advances in the respiratory care have revolutionised the therapeutic options for chronic respiratory failure and improved the outlook of DMD patients [11–14]. These advances and a change in society's expectation of long-term disability changed the reluctance of many physicians to provide long-term ventilation (LTV) to DMD patients [15]. Nevertheless, some physicians still remain sceptical about the benefit of LTV in a progressive neuromuscular disorder and would rather not offer this option to their patients. Two recent studies found that 25–36% of all physicians involved in the care of DMD patients would never discuss the option of LTV with their patients and families [16, 17]. The most common reason was their personal belief or perception of jeopardising the dignity of a severely disabled individual by inappropriately prolonging his life. Other reasons were the lack of established cost-effectiveness and reluctance to spend public resources for this purpose. However, even for those physicians who felt LTV was an important

life-saving measure, it was still not clear when the discussion with the family and the patient should start. As a result, it is not unusual that the decision to initiate mechanical ventilatory support is made non-electively in an emergency situation during the first presentation with acute respiratory failure. This leaves little time to discuss the advantages and disadvantages of LTV with the patient and the family, and makes it highly unlikely that they can participate actively in the decision-making process.

Our study was designed to examine the attitudes and clinical practice of physicians in Switzerland involved in the care of DMD patients. It was our goal to gain information on how pulmonary function is monitored and assessed and whether the option of long-term ventilatory support in the later course of the disease is discussed with the patients or the caregivers at an early stage. Simultaneously, we aimed to gain information from the DMD patients themselves about their current awareness and knowledge of LTV.

Methods

A questionnaire was sent by mail to physicians, including pulmonologists and neurologists known or suspected to be involved in the care of children and adults with DMD. The survey was performed between January and May 2004. Participants were asked to respond anonymously to the questionnaire, which was mailed together with a covering letter explaining the purpose of the survey. The questions were developed based on a previously published questionnaire from a Canadian study [16] and a second questionnaire used in a similar study in the United Kingdom (unpublished, personal communication Prof. F. Muntoni, Dubowitz Neuromuscular Centre, Department of Paediatrics, Imperial College, London W12 OHN, UK) and adapted to Swiss clinical practice. The specific information requested from each physician in our anonymous questionnaire included:

- demographic and clinical details on DMD patients
- usual frequency to order routine follow-up examination, pulmonary function tests (PFT) and sleep studies
- personal opinion and practice to disclose information about long-term ventilation to patients and families

Concurrently, we identified 90 DMD patients through the Swiss Association of Children with Muscle Disease. We sent these patients and their families another questionnaire asking them for clinical information such as the frequency of pulmonary infection, the mode and frequency of respiratory follow-up examinations and their current awareness and knowledge of the possibility of long-term ventilation. We asked the patients to respond anonymously. The Ethics Committee of the Canton Ticino approved this study.

Results

Physician survey

Thirty physicians were identified to be primarily involved in the care of DMD patients in Switzerland and were mailed the questionnaire. Twenty-six (87%) of the 30 physicians returned the questionnaires and were included in the final analysis. Two physicians came from the Italian part of Switzerland; nine from the French part and fifteen from the German part of Switzerland. Fifteen (58%) physicians were paediatric neurologists and 11 (42%) were either adult (9) or paediatric (2) pulmonologists.

Individual practice of patient examinations and monitoring of respiratory function

The physicians stated that they scheduled DMD patients for routine clinical examinations

every 6 months (range 3–12 months). The frequency of routine examinations depended on the age of the patient. Table 1 shows the frequency of performing PFT and sleep studies with respect to age. We found that 88% of the physicians perform PFT every 9 months in DMD patients older than 10 years. Routine PFT is most commonly performed in patients older than 15 years of age. 12% (3/26) of the physicians indicated not to examine pulmonary function on a routine basis and 12% (3/26) did not answer this question. 37% (7/19) of the physicians responded that they do not perform sleep studies (overnight continuous oxygen saturation monitoring) in children less than 10 years of age. Only 53% (14/26) of the physicians perform regular sleep studies at intervals of 6–9 months in patients older than 10 years. The remaining physi-

Table 1

Frequency of performing pulmonary function tests and sleep studies in relation to the age of DMD patients.

Frequency of performing PFT					
Age of patients	Number of physicians	Never	Sometimes	Regularly	Time interval (months)*
5–9 yrs ^a	19 (73%)	2 (10%)	7 (37%)	10 (53%)	12
10–14 yrs ^a	18 (69%)		3 (17%)	15 (83%)	9
15–19 yrs ^a	18 (69%)		2 (11%)	16 (89%)	9
>20 yrs ^b	11 (42%)		1 (9%)	10 (91%)	6
Frequency of performing sleep studies					
Age of patients	Number of physicians	Never	Sometimes	Regularly	Time interval (months)*
5–9 yrs ^a	19 (73%)	7 (37%)	11 (58%)	1 (5%)	12
10–14 yrs ^a	18 (69%)	4 (22%)	6 (33%)	8 (44%)	9
15–19 yrs ^a	18 (69%)	3 (17%)	4 (22%)	11 (61%)	9
>20 yrs ^b	11 (42%)	3 (27%)	2 (18%)	6 (54%)	6

* Data are median in months; physicians who did not follow patients were excluded from this calculation

^a 6 physicians did not follow patients in this age group

^b 14 physicians did not follow patients in this age group

Table 2

Practice of surveyed physicians to disclose and discuss the options of LTV with DMD patients and family members.

	Disclose information about LTV for the first time to patients and families	Discuss the options of LTV in-depth with patients and families
Soon after diagnosis	7/25 (28%)	2/26 (8%)
After occurrence of clinical symptoms	8/25 (32%)	13/26 (50%)
After evidence of respiratory failure by PFT	9/25 (36%)	10/26 (38%)
Other reason*	1/25 (4%)	1/25 (4%)

LTV: Long-Term Ventilation

PFT: Pulmonary Function Tests

* One physician stated that his decision to discuss options of LTV depends highly on the presence of problems and issues other than clinical signs of respiratory failure

Table 3

Results from the DMD patient survey on respiratory care.

Age	N	History of respiratory tract infections	Pulmonary function test	Sleep study	Discussion about LTV	Ventilatory support
5–10 yrs	6	0	0	0	2 (33%)	0
10–15 yrs	16	3 (19%)	15 (94%)	5 (31%)	7 (44%)	4 (25%)
15–20 yrs	13	6 (46%)	13 (100%)	8 (61%)	10 (78%)	4 (31%)
>20 yrs	8	5 (62%)	8 (100%)	7 (88%)	7 (88%)	6 (75%)

DMD: Duchenne Muscular Dystrophy, LTV: Long-Term Ventilation

cians either do not routinely perform sleep studies or they did not answer this question. Six of the 26 physicians (23%) did not care for patients on LTV, sixteen physicians (65%) cared for patients requiring ventilatory support for less than 24 hours per day, and three physicians (11%) were looking after patients requiring continuous long-term ventilation. A total of 55 ventilator-dependent DMD patients were identified. The most commonly used ventilator mode for both part-time and continuous ventilator users was bi-level positive pressure ventilation via nasal mask (93%).

Individual practice of disclosing information on LTV to patients

Half of the physicians were likely to inform families and their patients about the option of long-term ventilatory support. Physicians reported always (35%), nearly always (15%) or sometimes (35%) to inform patients of this option.

Only 8% (2/26) of the physicians rarely disclosed this option and 4% (1/26) of physicians did not answer this question. Only 28% (7/26) of the physicians indicated that they would inform patients about the possibility of LTV or discuss this option soon after diagnosis. 50% of the physicians started in-depth discussions with the patients and families about LTV only when the first clinical symptoms of respiratory compromise occur, and 38% do so when respiratory failure becomes clearly evident in sleep studies or blood gas analyses (table 2).

Survey of patients (table 3)

Ninety DMD patients were identified through the Swiss Association for Muscle Disease. 43 (48%) of the patients returned the questionnaire and were included in the final analysis. Six patients were from the Italian part, 16 from the French part and 21 from the German part of Switzerland. Patients were grouped according to age (see table 1).

33% (14/43) of patients required LTV, of whom 13 patients were on non-invasive mask ventilation and one patient was ventilated via tracheostomy. Overall, 58% (25/43) of the patients responded to the question whether they were satisfied with the information on LTV provided by their physician, and 92% (23/25) indicated that they were.

The median interval for routine clinical examinations was 9 months (range 4–12 months) for the 5–10 yrs age group, 6 months (range 3–12 months) for the 10–15 yrs age group, and 6 months (range 3–12 months) for the 15–20 yrs age group and patients older than 20 years. Problems with previous respiratory tract infections increased with age.

None of the children between 5–10 years underwent a pulmonary function test or a sleep study, while older children had pulmonary function tests performed. Overnight sleep studies did not belong to the routine care in even older children, since less than a third of the children between 10–15 years and less than two thirds of the children between 15–20 years reported such examinations. Similarly, the majority of patients younger than 15 years did not report a discussion about LTV with their physician. However, older patients did report such a discussion, although many of those were already on ventilatory support.

Discussion

According to a recent consensus statement physicians have a legal and ethical responsibility to disclose treatment options, including long-term ventilation, to patients and their families. In addition, physicians should not decide to offer this type of information based on their own perceptions of quality of life [18]. In fact, more than simple disclosure is required to assist DMD patients in making a decision with respect to LTV that corresponds with their own values and preferences. Our study revealed that only 8% of the surveyed Swiss physicians choose to restrict the information about LTV and do not discuss this option with their DMD patients. Most physicians indicated that they disclose information on LTV to their DMD patients. This is in contrast with two earlier North-American studies which reported that between 25–36% of the physicians involved in DMD patient care never disclose information on LTV to their patients and families [16, 17]. These studies found that nighttime LTV has become standard practice. However, full-time invasive mechanical ventilation is not routinely discussed with all patients, because physicians varied in their opinions as to whether or not this represents a benefit or a burden for the patients. We believe that the results of our survey are most likely related to the increasing awareness of such technology and the growing evidence that full-time LTV can improve both the quality of life and the survival of DMD patients [16, 21, 22]. Another reason might be that physicians no longer control medical information on treatment options with the availability of modern communication technology and the support provided by patient associations. Nevertheless, the favourable responses by the physicians are somewhat in contrast to the responses given by the patients. A considerable number of even older patients with advanced disease indicated that they had not been informed by their physician about the option of LTV. While our survey is representative for the specialist physicians involved in the care of DMD patients in Switzerland, it does not provide

a comprehensive picture of the medical practice and attitude of DMD patient care in Switzerland.

We also wanted to find out when physicians start to inform their DMD patients and family members about the option of LTV. Only a minority of the surveyed physicians (26%) indicated that they would notify DMD patients of this option soon after diagnosis. Most surveyed physicians (88%) start in-depth information on LTV not until symptoms and signs of respiratory compromise have developed. This seems hardly appropriate given the fact that respiratory failure inevitably arises in these patients, and that the need for ventilatory support may even develop abruptly in association with a respiratory tract infection. Usually, the evolution of respiratory failure in DMD patients is a gradual one and starts with hypoventilation during REM sleep, progresses to hypoventilation during the entire sleep and ends in diurnal respiratory failure. Independent of the best timing to initiate ventilatory support, it is generally agreed that the option of LTV should be disclosed to the patients early, preferably soon after diagnosis, to enable patients and family members the opportunity to be involved in the decision-making process well before respiratory failure occurs and to establish end-of-life directives in the case of an emergency [18].

Recommendations when to start which type of ventilatory support have recently been published for DMD patients [18]. It is clear that such decisions can only be made appropriately, if the patient's respiratory status is regularly assessed and the patients have been thoroughly informed about ventilatory support options throughout the course of their disease. Our study revealed a considerable lack of uniformity in the long-term respiratory follow-up of the DMD patients in Switzerland. Repeated spirometric measurements provide a simple and relatively powerful mean of assessing disease progression. It has been demonstrated that the rate of decline of vital capacity predicts survival time and that a vital capacity below 1 L is a strong

marker of subsequent mortality in these patients (5-yr survival 8%) [19]. Nocturnal hypoventilation becomes likely if vital capacity falls below 40% [20]. Our survey indicates that pulmonary function is regularly assessed in older non-ambulant DMD patients. Current guidelines recommend performing regular pulmonary function assessments bi-annually or annually after confinement to a wheelchair, fall in vital capacity below 80% predicted, or age 12 years [18, 21, 22]. Only 42% of the surveyed physicians perform regular sleep studies in the older non-ambulant DMD patients, although it is well recognised that hypoventilation and respiratory failure will become manifest first during sleep. Although there are no randomised, controlled trials, there is growing evidence that nocturnal non-invasive positive pressure ventilation results in improved survival, improved quality of sleep, improved well-being and a slower rate of decline in pulmonary function compared with non-ventilated subjects [5, 14, 23–27]. Conversely, preventive nasal ventilation of DMD patients before the development of respiratory compromise should be avoided and does not prolong survival [28, 29]. Hence, annual evaluation for sleep-disordered breathing has been recommended in patients with DMD starting from the time they are wheelchair users and/or when clinically indicated that nocturnal ventilation can be initiated timely with the occurrence of sleep-disordered breathing [18].

One limitation of our study is the low number of participating physicians. While our results may reflect the current practice of medical specialists involved in the care of DMD patients, they do hardly give an accurate and comprehensive picture of the habit and attitude of general practitioners and other physicians involved in the care of DMD patients. Nevertheless, it seems that even specialised physicians could improve the respiratory follow-up and care of DMD patients and may not be fully aware of current international practice and recommendations. Most of the surveyed physicians are hospital based, but probably not all DMD patients are regularly followed at a specialised centre. This may explain the slightly diverging responses given by the physician and the patients with respect to information on LTV. Another limitation is the low response rate of the patients, which could lead to a bias when drawing conclusions from the patients' responses, although it is

hard to predict in which direction. In addition, the lack of a registry of DMD patients made it impossible to contact all DMD patients in Switzerland.

Today, a nihilistic approach to ventilatory support in patients with DMD is no longer acceptable. A positive attitude towards long-term ventilatory support should be encouraged regarding its benefit on morbidity and mortality in these patients. Since the average age for developing respiratory failure is 18–20 years [30, 31], paediatricians are well placed to introduce and discuss the topic of LTV early in the disease course [34]. Paediatric pulmonologists should be involved early in the respiratory follow-up of children with DMD to assess the rate of decline in pulmonary function and the occurrence of hypoventilation. Another important aspect in the care of these patients is an optimal transition from paediatric to adult care. It is crucial that patients, families and caregivers play an important role in the decision-making and are fully aware of these support modalities [32].

In conclusion, our survey found that most specialists involved in the care of DMD patients disclose information on LTV to DMD patients, but commonly late in the disease course after respiratory problems have developed. The current routine of respiratory follow-up of DMD patients seems insufficient, at least with respect to early detection of nocturnal hypoventilation. We believe that the results of our survey should strongly encourage the education of physicians involved in DMD patient care about current international standards and the development of national recommendations on the respiratory follow-up and care of children with neuromuscular disorders.

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