

## Perspectives on Palliative Care Among Duchenne Muscular Dystrophy Patients and Their Families in Singapore

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### Abstract

**Introduction:** With better medical care, patients with Duchenne muscular dystrophy (DMD) now live longer but face more complex medical and social needs. This study described the perceptions of DMD patients and their families of disease-specific palliative care services in Singapore. **Materials and Methods:** A multicentre, cross-sectional study involving DMD patients and their families was carried out. Structured questionnaires were administered to them to collect data on their understanding of palliative care, health services accessed and desired by them and quality of life. **Results:** A total of 30 pairs of DMD patients and their caregivers responded. Most patients were >13 years old (70%) and non-ambulant (86%). Most of them and their families (70%) were also not aware of palliative care and support services that were available to them in Singapore. Additionally, they perceived greater financial assistance and better transport services as resources that could better meet their care needs. The presence of scoliosis and need for ventilatory support were associated with lower quality of life in patients. **Conclusion:** There is a need to improve awareness and provision of palliative care services for DMD patients in Singapore where discussion of end-of-life care is often considered taboo. Prevention and correction of scoliosis and provision of appropriate ventilatory support may improve quality of life in DMD patients.

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**Key words:** Advance care planning, Palliative support services, Quality of life

### Introduction

Duchenne muscular dystrophy (DMD) is an X-linked, progressive neuromuscular disorder that causes loss of mobility and comorbidities including cardiomyopathy, contractures, respiratory insufficiency, scoliosis and eventual loss of ability to care for oneself.<sup>1,2</sup> Despite advances in medicine, DMD remains incurable.<sup>3</sup> Without non-invasive ventilation (NIV), mean age of death in DMD patients in the past was 14.4 years (95% confidence interval, 11.9–16.82 years).<sup>4</sup> However, nocturnal ventilation has improved the likelihood of survival of up to 25 years in DMD patients.<sup>4</sup>

In the late stage of the disease, DMD patients are typically wheelchair-bound, on NIV support and dependent on others for their care. Their nutritional status is also greatly compromised from progressive

swallowing dysfunction and they experience pain from scoliosis and contractures with poor sleep.<sup>5–7</sup> Beyond the direct costs incurred by DMD patients for medical intervention, the impact of the disease on their caregivers and families can be huge since their extensive health needs can cause much emotional, financial and psychological stress on their resources and coping abilities.<sup>8,9</sup> Increasingly, as DMD patients enjoy longer life expectancy, it means that the duration of nutritional, physical and social issues that significantly impact their quality of life has been extended.

Consequently, efforts to destigmatise and effect a change in the perceptions of the general population of palliative care as “giving up hope” still remains very much a work in progress. Poor awareness of “palliative care” in DMD patients and even health professionals

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has been reported.<sup>10</sup> The World Health Organization has defined palliative care for children as “the active total care of the child’s body, mind and spirit, and also involves giving support to the family. It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease. Health providers must evaluate and alleviate a child’s physical, psychological, and social distress. Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited. It can be provided in tertiary care facilities, in community health centers and even in children’s homes.”<sup>11</sup>

The shift from an adult-centric approach to a child-centric one in palliative care will help to facilitate early integration of palliative care into the care plans for children and young patients of various diseases including: 1) conditions where cure is possible but may fail (such as cancer); 2) diseases that are progressive (such as neurodegenerative disorders); 3) incurable and progressive conditions but the child may live for an extended period of time (such as DMD); and 4) non-progressive conditions but with a shorter lifespan (such as cerebral palsy).<sup>12</sup> This paradigm shift can be helpful and effective in addressing the unique needs that surface throughout the duration of life-limiting or life-threatening conditions and to provide the best quality of life and care for every child.<sup>13</sup> Early interventions had been shown to confer a positive effect on longevity and quality of life when patients and their families made changes to their care and life plans.<sup>14,15</sup>

To date, no studies in Singapore have sought to quantify the usage level and accessibility of palliative support services, identify barriers to their use and assess the degree of awareness and understanding of palliative care by DMD patients and their families. To address this gap in the literature, our study aimed to provide a situation analysis of the state and accessibility of palliative care. It also attempted to shed light on the understanding of palliative care among DMD patients and their families in Singapore.

## Materials and Methods

This was a multicentre, cross-sectional study involving outpatient male DMD patients who were identified from the multidisciplinary neuromuscular clinics in the paediatrics department of the National University Hospital (NUH) and Muscular Dystrophy Association Singapore (MDAS). Informed consent was obtained from patients and their families. Ethics

approval was granted by the Domain Specific Research Board of the National Healthcare Group, Singapore.

Patients were recruited over a 6-month period using purposive sampling. Data was collected through interviews with them and their caregivers using structured questionnaires that evaluated functional mobility, demographics, palliative services that they had accessed and desired and their understanding of palliative care. Monthly family household income was dichotomised to either above or below S\$6000 for the purpose of analysis, as it was the mean monthly household income in Singapore at the commencement of this study.<sup>16</sup>

Data on quality of life of patients and caregivers were collected using 2 validated questionnaires: the Pediatric Quality of Life Inventory™ (PedsQL™) Family Impact Module and the parental and age-appropriate child reports, PedsQL™ Neuromuscular Module (Version 3.0). For the latter, patients completed either the Young Child report (for those aged 5–7 years old), Child report (aged 8–12 years old) or Teen report (aged 13–18 years old).

No sample calculation was done. Statistical analyses were performed using SPSS Statistics for Windows, Version 20.0 (IBM Corp., Armonk, NY, USA). The relationship between service outcomes and predictive variables was derived from multivariate analysis.

## Results

A total of 33 patients were screened. Most (77.0%) of them were identified from NUH. One patient and his family declined to provide consent and another retracted their consent after they felt that the questions posed to them were too sensitive. Eventually, 30 pairs of DMD patients and their families completed the questionnaires. Table 1 shows their demographic and clinical profile. Median age of patients was 17 years (range, 7–29 years) and 21 (73.0%) of them were ≥13 years old and mostly wheelchair-bound. Eight (26.7%) of them were on NIV. Non-ambulant patients who had intact hand functions were able to manoeuvre their motorised wheelchairs.

Table 2 shows the usage frequency of health services by the patients. About 80% of them were on follow-up by the cardiologist, respiratory physician and physiotherapist that were consistent with the late stage of the disease. MDAS was their primary source of social support and 24 (80.0%) of them desired respite care. For services that were already available in Singapore, the desire for more accessible transport services topped the list for 27 (90.0%) of them while 25 (83.0%) of them indicated a desire for a case manager and home

Table 1. Demographic and Clinical Profile of Duchenne Muscular Dystrophy Patients

Variable	Patients (n = 30)
Median age in years (range)	17 (7 – 29)
At diagnosis	6 (0 – 11)
At onset of symptoms	5 (2 – 11)
Age when wheelchair was needed	10 (6 – 15)
Monthly household income ≤S\$6000 (%)	18 (60.0)
Chinese ethnicity (%)	26 (86.7)
Patients who required non-invasive ventilation (%)	8 (26.7)
Patients with scoliosis (%)	17 (56.7)
Patients with contractures (%)	20 (66.7)
Mobility (%)	
Ambulate independently	4 (13.3)
Uses manual wheelchair	7 (23.3)
Uses motorised wheelchair	19 (63.3)
Scope of care needed (%)	
Shower	24 (80.0)
Dressing	23 (76.7)
Bringing patient outdoors	21 (70.0)
Transfers	20 (66.7)
Feeding	15 (50.0)
Turning during sleep	15 (50.0)

respiratory care services. Finally, 14 (50.0%) of them wanted pain management service.

Patients resided in public flats with a mean family size of 5 members (range, 3–8 individuals). Among them, 18 (60.0%) had monthly household income ≤S\$6000. After medical subsidies given by the Singapore government, 14 (46.7%) patients and their families still required financial assistance. Consequently, greater financial support was listed as a desired service by them (76.7%).

However, only 9 (30.0%) patients and their families were aware of palliative care and 22 (73.0%) of them had never heard of Advance Medical Directive (AMD). After they viewed a simple description of AMD provided in the questionnaire, 19 (63.3%) caregivers agreed to support patient's decision to either accept or refuse medical intervention upon turning 21 years old.

Table 3 shows significant differences in reports between patients and their parents pertaining to the dimension on “about my neuromuscular disease” in the neuromuscular module ( $P = 0.041$ ). Parents believed

Table 2. Utilisation of Health Services in Singapore by Duchenne Muscular Dystrophy Patients

Service	Patients, n = 30 (%)
Current access to	
Cardiologist	26 (86.7)
Physiotherapy	25 (83.3)
Respiratory physician	24 (80.0)
Neurologist	23 (76.7)
Social support group in MDAS	23 (76.7)
Orthopaedic surgeon	16 (53.5)
Financial support (through medical social worker)	14 (46.7)
Transport service	13 (43.3)
Occupational therapy	12 (40.0)
Pastoral or religious support	8 (26.7)
Respiratory therapy service	6 (20.0)
Speech therapy	5 (16.7)
Home nursing support	4 (13.3)
Desired access to	
Enhanced transport service	27 (90.0)
Case manager*	25 (83.3)
Home respiratory support*	25 (83.3)
Respite care	24 (80.0)
Greater financial support	23 (76.7)
Pain management	15 (50.0)

MDAS: Muscular Dystrophy Association of Singapore

\*Not available in Singapore.

that the disease was causing more physical problems in patients than what was perceived by the latter. Our study did not explore the reasons for this difference.

Independent factors that affected health-related quality of life (HRQoL) were identified from multivariate analysis. Presence of scoliosis and lower family income were associated with low HRQoL (Table 4). Patients and their parents also attributed low HRQoL to the need for ventilatory support. However, only caregivers reported that the presence of scoliosis was linked to low HRQoL in patients.

## Discussion

In our study, DMD patients (30%) had poor knowledge of “palliative care”. This finding could be attributed to the likelihood that palliative care is still perceived

Table 3. Parental and Child Reports on Pediatric Quality of Life Inventory™ Neuromuscular Module

Dimension	Median Parental Report (Range)	Median Child Report (Range)	P Value
About my neuromuscular disease	62.5 (14.7 – 86.8)	67.65 (16.2 – 97.1)	0.041
Communication	66.67 (0 – 100)	58.33 (16.7 – 100)	>0.05
About our family resources	55 (20.0 – 100)	62.5 (20.0 – 100)	>0.05
Total score	61 (17.0 – 88.5)	66.83 (21.0 – 87.0)	

as terminal care in conditions such as cancer than in life-limiting disorders like DMD. Although AMD is available for close to 2 decades, its awareness and use remain low.<sup>17</sup> In our study, less than one-third of patients and their families were aware of AMD. Eventually, over half of them reported that they would support the patient's choice after AMD was made known to them.

Our study did not examine knowledge of advance care planning (ACP) in DMD patients and their families. Nevertheless, the need for ACP to better prepare families and provide a continuum of care to support them as the disease progresses—and at the same time provide optimal control of physical symptoms in patients—should be emphasised.<sup>14</sup> Previous studies had identified barriers that precluded consideration of ACP and AMD including lack of knowledge of palliative care, lack of access to supportive services and cultural aversion to the taboo subject of end-of-life care (especially in an Asian country like Singapore).<sup>18,19</sup> However, it is anticipated that as more DMD patients live longer into adulthood, the need to educate them and their families on ACP and AMD will become greater.

Our study found that 50% of patients and their families who were aware of palliative care felt that existing support services were inadequate, suggesting that such

services remain suboptimal in the country. Since DMD patients make repeat visits to various health providers as the disease progresses, there is a need to increase accessibility to various palliative support services that are offered in the community.

Most families (83.3%) also indicated a desire for a case manager who can render personalised and consistent continuity of care to patients with complex medical conditions. In Singapore, this role is currently undertaken by the physician who attends to the patient on a regular basis. In the long run, it is not a feasible arrangement in view of the high patient load that every physician sees in public hospitals throughout the country. Currently, primary care practitioners also play a limited role in end-of-life care.<sup>20</sup> To address this situation and improve the delivery of palliative care services to DMD patients, case managers and nurses can be trained to provide specialised and holistic care to this group of patients.<sup>21</sup>

In our patients, utilisation of medical care was high and this could be attributed to selection bias of patients who attended the multidisciplinary neuromuscular clinics. Most (73%) of them were older non-ambulant boys ( $\geq 13$  years old). Loss of independent ambulation was associated with nocturnal hypoventilation and eventual NIV support, even in asymptomatic children with neuromuscular disorders.<sup>22,23</sup> About 25% of our patients were on NIV support, yet only 6 (20%) of them were recipients of respiratory therapy. Not surprisingly, 25 (83.3%) patients desired to be offered home respiratory care services.

Pain has been reported by non-ambulant DMD patients.<sup>24</sup> Although most of our non-ambulant patients had intact hand functions that allowed them to manoeuvre their motorised wheelchairs, they still required assistance in turning to relieve them of the pain from remaining in the same position over long periods of time. Consequently, our finding that nearly 50% of them desired pain management service is hardly surprising.

Half of our DMD patients required assistance in activities of daily living such as feeding, shower and transfers, and 16 caregivers had helpers to mitigate

Table 4. Factors Affecting Health-related Quality of Life

Variable	$\beta$ (95% CI)
PedsQL™ Family Impact Module	
Presence of scoliosis	-21.9 (-36.4 – -7.4)
Monthly household income $\leq$ S\$6000	-16.7 (-27.7 – -5.8)
PedsQL™ Neuromuscular Module	
Requires ventilatory support	
Parent proxy	-12.6 (-24.0 – -1.2)
Child	-18.7 (-31.5 – -5.8)
Presence of scoliosis	
Parent proxy	-13.2 (-23.2 – -3.2)

$\beta$ : Beta coefficient; CI: Confidence interval; PedsQL™: Pediatric Quality of Life Inventory™

caregiver exhaustion. Nevertheless, most (80%) of them required respite care. Although respite care can alleviate the caregiving burden of individuals caring for patients with chronic needs, it is not part of the standard of care.<sup>25,26</sup> In MDAS, caregivers are offered short-term breaks in a designated room while others tend to their sons who are participating in its programmes. Star PALS, a paediatric palliative care service in Singapore, also offers home and respite care. The provision of accessible and flexible family-centered, home-based respite care to support families in caring for children with complex needs should form part of the standard of care.

Although Singapore is a small city-state and medical care is within easy reach of her residents, this study found that the safe transport of non-ambulant DMD patients was a valid concern. Although the country has invested heavily to upgrade and develop its public transport infrastructure to accommodate individuals with disabilities, non-ambulant DMD patients still struggle to use public transport since their motorised wheelchairs are often too bulky and heavy and they do not fit into a regular taxi or sedan. For reasons of safety and comfort, it is important for them to use a vehicle that is fitted out with a wheelchair retention system. In our patients, 27 (90.0%) of them were vocal in their wish to have more transport options made available to them. Consequently, our institution has partnered various charities to fund regular transport services to fetch patients for their hospital appointments and return them home when completed. However, a more innovative and sustainable funding model is needed to meet the need of this group of patients.

Another key finding of our study was the economic cost and burden of disease DMD has on families and society which are magnified as the disease progresses.<sup>8</sup> In our study, about 60.0% of patients and families had monthly household income  $\leq$  S\$6000 (or S\$72,000 per annum) that was comparable to the mean household income in Singapore. With a complex disease like DMD, its impact on the finances of patients and their families is substantial. Consequently, 23 patients and their families indicated that they would like to be awarded more financial assistance. With longer life expectancy, there is greater need for financial support by DMD patients to meet their increasing medical costs from hospitalisation, medications, clinic visits and purchase of assistive devices. Additionally, the economic burden on the household and society—in terms of lost productivity—will be felt when their caregivers are forced to cut back on their work hours or to cease employment altogether to care for them. Clearly, there is a need to formulate robust policies and programmes

that can help to ease the financial hardship faced by DMD patients and their families.

Parents of the patients reported that the presence of scoliosis was associated with low quality of life in their children. With loss of ambulation, neuromuscular scoliosis from pelvic imbalance sets in.<sup>6</sup> The discomfort a patient experiences from sitting over a long period of time—which worsens as the patient deteriorates gradually—necessitates the use of fitting orthotic devices and appropriate wheelchairs and early evaluation for surgical stabilisation of the spine. Although the presence of pain was not specifically asked in our study, it may be an important parameter that affected quality of life in our patients. Consequently, pain assessment should be included as part of screening and care assessment.

Both parents and patients reported that the need for ventilatory support was associated with low quality of life. Some studies had reported that the need for NIV did not correlate with poorer quality of life, and that home ventilation could lead to improvement in quality of life.<sup>27,28</sup> However, in their study of 80 ventilator-assisted DMD patients, Bach et al found that 12.5% of them reported dissatisfaction with their life.<sup>29</sup> Moreover, Baiardini et al reported that the use of wheelchairs and ventilators were significantly associated with lower quality of life.<sup>30</sup>

## Conclusion

As the epidemiologic profile of DMD patients changes, palliative care is important to address the complexity of their evolving needs and to ensure good quality of life for them and their families. More effort is needed to improve awareness of palliative care and for it to begin early in DMD patients. There is a need to develop robust and sustainable palliative care services including ACP and respite care, and more specialised, wheelchair-friendly public transport facilities are needed too. As DMD patients live longer, greater financial support is helpful to cushion the financial burden of the disease on patients and their families. Medical interventions such as prevention or early treatment of scoliosis and proper ventilatory support can help improve the quality of life of DMD patients. Although DMD remains incurable, there is much that can be done to help patients and their families enjoy a reasonable quality of life.

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