

Palliative Care in Duchenne Muscular Dystrophy: A Study on Parents' Understanding

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Abstract

Introduction: Duchenne muscular dystrophy (DMD) is a neuromuscular disease of childhood, which has clear progression. The international standardized care guidelines for DMD suggest that palliative care is essential for the affected children. **Objective:** To explore the parent's understanding of palliative care services available for children with DMD and the challenges faced by them in utilizing the same. **Methods:** A cross-sectional qualitative exploratory study was conducted among six families of boys diagnosed with DMD. A semi-structured interview guide with prompts was used to conduct in-depth interviews which lasted for an average of 1 h. Thematic analysis was done to identify the pattern or themes. **Results:** The major themes identified were "palliative care, living with DMD, awareness about palliative care services and challenges." *Awareness about palliative care services* is the dominant theme identified as influencing rest of the experiences narrated by the parents of children with DMD. **Discussion:** Integration of palliative care services from an early stage of the illness can help the child to make transition from one stage to another stage of the illness. To ensure the utilization of the available palliative care services, there is a need to create awareness about it among the general public. **Conclusion:** Introducing the concept of palliation of symptoms and ensuring quality of life of the child with DMD by accessing the available services can aid the parents to reach out for help for their child.

Keywords: Children with duchenne muscular dystrophy, paediatric palliative care, parents

INTRODUCTION

Recent advances in medicine have improved survival among children with any life-limiting illness. However, approximately 25% of these children would still die of their disease even after many years of medical care.^[1] Care requires a holistic approach to the child's and families' physical, emotional, psychological, social, financial and spiritual needs. The shift in focus from cure to long term care has been advocated worldwide for many chronic conditions. Emphasis on providing palliative care interventions to ensure optimal care, improving the quality of life of the person suffering from the condition as well as their families are of the focus of intervention for many life-limiting conditions. Palliative care for adults is now a recognized specialty worldwide, but the same is not true for paediatric palliative care.^[1] Only recently, palliative care for children began to receive the attention of the health care providers.

The societal stigma associated with the belief that "children are not supposed to die," prevents the discussion about the concept of palliation among the needy group.^[2]

Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family. It begins when the illness is diagnosed and continues regardless of whether or not a child receives treatment directed at the disease. Effective palliative care requires a broad multidisciplinary approach that includes

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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 centres and even in children's homes.^[3]

Paediatric palliative care in India evolved in the mid-1980s. Multiple stakeholders are involved in the process of easing the suffering of children at various stages of their illness. Family and care professionals work side by side with the active treatment team. In India, along with the parents, extended family members play a vital role in the care of a child with a chronic condition. The child has been seen as an extension of self by most of the parents. Hence, a search for a cure for a child's chronic health condition is always the priority. Family is never prepared for the death of their child or even the discussion about providing palliative care interventions.

Duchenne muscular dystrophy (DMD), one of the nine types of muscular dystrophy is a rare genetic disorder characterized by progressive muscle degeneration and weakness. It generally manifests in children in early childhood, usually between ages three and five. As the path of progression is clear for DMD, discussions about the suitable interventions can be initiated at the early stages of the illness.^[4] The condition requires highly specialized care, especially during the later stage.^[5] Treatment decisions can become complicated if the patient and the family are not made aware of the condition, possible difficulties and the help available.

Studies conducted among Indian cohort of parents of DMD children found that parents deny the child's condition for longer period and they often experience chronic guilt and helplessness.^[6] They have inadequate understanding of the disease, experience moderate burden, rely more on religion, focus on venting of emotions, and instrumental and emotional social support for coping.^[7] The treatment guidelines for DMD emphasize multidisciplinary care.^[8] As per the international standardized care guidelines, palliative care forms a major intervention in this multidisciplinary care for the patients with DMD.^[9] The studies focusing on the implementation of palliative care guidelines for children with DMD from Indian context is hardly found. The current study explored the parents' understanding of palliative care services for children with DMD, the services available and accessed by them and the challenges in utilizing them in Indian context.

METHODS

Design

It is a qualitative exploratory study with a cross-sectional design.

Tool

A semi-structured interview guide was prepared for this study. This was developed through the literature search and the interview with the experts working with children with DMD. Later the interview guide was validated by five experts from the multidisciplinary team. The schedule contains

open-ended questions. The format allows use of probe questions and prompts as per the statements and responses given by the parents during the course of the interview.

Participants

Through purposive sampling, six parents (either father or mother) of a child diagnosed with DMD receiving treatment from the national tertiary referral center for Neurological disorder were recruited for the study. The participants were the primary caregivers of the children with DMD, who had given consent for audio recording and a home visit.

Procedure

The study received ethical clearance from the Institute Ethics Board of National Institute of Mental Health and Neuro Sciences (NIMHANS), Bengaluru. The children diagnosed with DMD and their families undergoing treatment at neuro muscular clinic at NIMHANS are referred for routine psychosocial intervention. Among those families, the first author approached the follow-up cases, explained the study and obtained written informed consent for interviewing in their respective homes. The interview was scheduled as per the convenience of the respondents. An in depth interview was conducted with the parents by using the semi-structured interview schedule prepared for this study. Parents were requested to narrate their understanding of palliative care services for children, services used by them and the barriers for the same. Interviews lasted for an average of 1 h. The interviews were participant-led, thus ensuring rigor. Probe questions and prompts were used only when necessary. These interviews were audio-recorded and transcribed. Two interviews were in English, and four were in Malayalam. Interview in regional languages was transcribed into English and back-translated by a regional language speaking person, in order to ensure the conformability of data.

Analysis

The interviews were transcribed, and the identifying information was removed, and new alphanumeric codes were given to each respondent. Transcripts were read and re-read multiple times. Thematic analysis was done manually according to the guidelines given by Braun and Clarke.^[10] Initial codes were made from the transcripts by the first author (AS), it was reviewed by second author (MW) and corresponding author (PTT) and revised after discussion with the research team. Peer debriefing about the themes was done as part of ensuring the rigor. The codes derived were audited by the research guide (PTT) before finalizing them. Once the codes were finalized, overarching themes were constructed. Mapping of themes was attempted in order to bring out the interconnection between the themes that reflects the parents' understanding about pediatric palliative care.

RESULTS

Sociodemographic details of the parents and clinical details of the children

Table 1 shows the relevant Sociodemographic details of

the parents and clinical details of the children with DMD. Among the six participants who gave consent to be part of in depth interview; there were four mothers and two fathers. Mothers were primarily homemakers and fathers were working. Five children were full time wheelchair users and in two families both children were affected. Caregiving is equally shared among parents.

Mothers were observed to have greater difficulty responding to the questions in the initial phases, but they shared as many experiences as their male counterparts with regard to the concept of palliative care, accessibility and barriers. The significant findings from the analysis are mentioned below.

The term palliative care

The participants were encouraged to narrate their thoughts and views about palliative care. They reported that the term palliative care is associated with *ambiguity*. Most of them associate the term “palliative care” to “death.” This association causes an internal stigma for the parents to think about palliative care services.

“It is all about death, and I can’t think about my child’s death” (C2).

“Never want to think about child’s death, so no discussion about palliative care” (C5).

The family with two children affected with DMD, had prior experience of the course and prognosis of this condition. They were aware of the necessity of palliative care, hence were not finding it difficult with the term “palliative care.”

“My first child died, and the second child is also affected. Now I feel our child require palliative care.” (C4).

The internal stigma against the term palliative care can influence the care of the child with DMD. Parents’ efforts to avoid thoughts about “death or loss” of their child, lead to a situation where the ideal care for their child is compromised.

Living with illness

Under this theme, parents of DMD children narrated how they deal with the *Relentless Progression*. Each participant was aware of the progression because all the children in the study were above 12 years and usually disease worsens by then.^[7] They could feel that the progression is occurring at

a faster pace, but the parents were not prepared to discuss this with their children.

“Never wish to talk about the progression of illness with child” (C5).

Parents reported that by the time they reach homeostasis about child’s difficulties, as the progression of illness is a continuous process, another difficulty would have started. Shift from one stage to further stages of functional limitations created imbalance for the family.

“Transitions are a most difficult process– each stages we breakdown” (C5).

“When we come with some solutions for his difficulties, he would have started with some new difficulty” (C3).

Another major experience narrated by the parents as their child’s difficulty in accepting the change occurring to him. When the parents try to modify the routine of the child according to the functional limitation caused by the progression of the symptoms, the child is never able to adjust to the change. He feels his freedom is curbed, and parents are trying to exert control over him. Parents feel helpless to make the child accept his health condition and move on with his life.

“I know his symptoms are drastically progressing since last 6 months. And he is never ready to accept this and creating numerous issues at home” (C2).

“He was very reluctant to lie down, he feels, he will fall sick and completely bedridden” (C6).

Along with the challenges caused by the symptom deterioration, handling their own emotional issues and that of their children becomes a challenging task for the parents living with children with DMD.

Awareness about palliative care services

Most of the participants are aware of the term palliative care and the aim of palliative care services. But palliative care pertaining to children, they were not much clear. The moment they hear about palliative care, they could assume the services for older people, but not for the children.

“Heard about it, but don’t have much understanding about it” (C3).

Table 1: Sociodemographic details of the parents and children with Duchene Muscular Dystrophy and their brief illness profile

Age	Class	DOI	Mobility	Sibling affected	Caregiver	Interview done with	Occupation
16	12	12	Nonambulatory	No	Both parents	Mother (C1)	Home maker
15	11	10	Nonambulatory	No	Mother	Mother (C2)	Home maker
13	7	8	Nonambulatory	No	Both parents	Father (C3)	Government employee
17	12	13	Nonambulatory	Yes	Both parents	Father (C4)	Private company
12	6	7	Nonambulatory	Yes	Mother	Mother (C5)	Home maker
15	10	9	Ambulatory	No	Mother	Mother (C6)	Self employed

DOI: Duration of illness

"I know about adult palliative care services, but is there something to address children's needs as well?" (C1).

In a condition like DMD, which is chronic in nature, the parents were not sure as to when the child require palliative care services. They were unaware of the indicators for palliative care interventions.

"When does the child require palliative care services?" (C5).

"What are the child palliative care needs?" (C2).

They are seeing their children for a few years in this particular condition and have witnessed them progressing as well. However, what all issues can be addressed through palliative care was not clear for many of the parents. At what stage of the illness, to approach the palliative care team, what symptoms of the child do get benefited from palliative care interventions, were a grey area for the parents.

Challenges

Under this theme, the *barriers* felt by the parents in being the caregiver to their child and also the challenges they felt in availing and accessing the palliative care services are described.

While living with the child with DMD, parents are met with challenges which they have to handle on a day to day basis. Lack of sensitivity and awareness from the general public and even from medical fraternity is a painful experience narrated by many of the parents.

"General Doctors and nurses are not aware of this condition, and they are not equipped to provide adequate support" (C4).

Many treat Child with DMD as someone who is going to die despite whatever intervention being done. Furthermore, hence, they find it futile to invest their time and energy for the child diagnosed with DMD. The knowledge about the fatality of the condition causes a block to people who are involved in the care of the child with DMD.

"When we consult the hospitals with some medical emergencies, most of the people respond negatively, and sometimes they ask on face why you brought your child here knowing this condition" (C3).

"We have informed you earlier also, getting him admitted here cannot solve the problem. Why do you want to bring him again and again here" (C6).

Another challenge narrated by some parents was about their difficulty accepting that their child's condition will have a poor prognosis. They have knowledge about the condition, but emotional acceptance of the problem is lacking.

"Even after seeing the children with the same problem, I have difficulty to think that my child will have similar problems" (C4).

Dealing with the child's needs causes another major challenge for the parents. Lack of open communication

between the parents and their children might have posed this challenge for the parents.

"We really do not know what the child thinks... or wants at this stage" (C3).

Parents know their children, they are very close to them as well, but what emotional stage the child goes through, especially during the transition into another stage of illness is not very clear for the parents.

Parents reported that for availing the palliative care services, they have to cross many barriers. The logistic issues to reach the centre are the primary difficulty.

"There are palliative care services available, but reaching to the centres are really difficult" (C3).

"Hours of the journey to service centres and that really consume our daily wages" (C2).

The opportunistic costs involved in availing the palliative care services, the expenses incurred by them, loss of wages of the parent accompanying the child, creates barriers for them to access palliative care services.

Mapping of themes

The final stage of thematic analysis, mapping of the themes^[10] was attempted for the themes. This helped the authors to find the interconnection among the themes emerged. Awareness about palliative care services is the dominant theme identified as influencing rest of the experiences narrated by the parents of children with DMD. The awareness about the palliative care services removes the misconceptions and stigma about the term "palliative care." This encourages parents in availing the palliative care services, which smoothens the transition phases across the illness trajectory. The attitude of the general public which becomes a challenge for optimal care of the child with DMD can be addressed by adequate awareness about the palliative care among general public [Figure 1].

DISCUSSION

The current study found that parents have difficulty to accept word palliative care, and most of the family connects this word with death. As reported by Wong and Baker in 1998, "children are not supposed to die" is imbibed in the minds of most of the parents. Parents' association of the word palliative care to "death" and "ultimate loss" may be a probable reason why most of them are never ready to accept the suggestion of palliative care for their children. Non-acceptance of palliative care on similar grounds was also observed in earlier studies in adults.^[11,12] Awareness about children's palliative care services is limited for the respondents of the current study.

There is a lack of awareness about the care to be provided for children with DMD. This lack of awareness about the palliative care benefit even among the medical fraternity, as

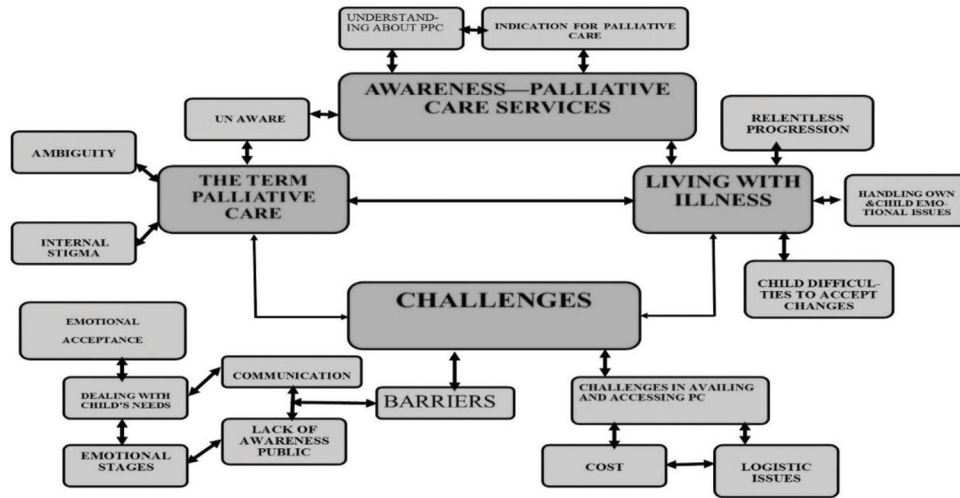


Figure 1: Thematic map

being reported in the current study, is the major challenge for the families in taking care of the children with DMD. As highlighted in the earlier studies,^[4] providing appropriate training to the general medical practitioners and creating adequate awareness among the caregivers may help deal with the barrier to paediatric palliative care.

One of the major challenges reported by the parents in the current study is a lack of clarity in children needs. They are not able to identify the emotional stage of the child, especially during the transition stages. There are different views about the child’s emotional state and experiences about their life-limiting condition. Some studies reported that until the age of ten, children are not aware of what is happening to them, and they do not experience or express anxiety about their death^[13] whereas, some other studies^[14] reported although children do not express their fears and anxieties directly, given a chance, they may do so indirectly. These findings point towards the need for conversations between parent and child may be assisted by the palliative care professionals, to deal with the anxieties related to the child’s death. Easing the emotional challenges surrounding this discussion could be of great support for the parents of children with DMD. At par with the earlier study findings,^[5,15] current study suggests introducing palliative care services early in the disease trajectory, to ensure optimal care for children with DMD, especially during the terminal stage.

Mapping of the themes identified “Awareness about palliative care services” as the major theme influencing other themes emerged from the study. The awareness about palliative care services gives the families an indication to avail of those services. It is the lack of awareness which creates ambiguity and internal stigma against the term “palliative care.” This attitude of the parents towards the term palliative care may make them reluctant to access the relevant services, and thereby becomes a major block in availing the services. The attitude of the general public and the health care professionals towards the term palliative care also makes it a barrier for the parents

of children with DMD. Though there are logistic issues to avail those services, the attitude towards it seems to be a primary factor in availing those facilities. The parent’s experience of living with the illness is also being influenced by their general attitude to the term palliative care. Proper awareness about the palliative care services helps in decreasing stigma associated with those services. The emotional difficulties faced by the parents in dealing with the child with DMD could have been a little eased through availing appropriate support services. Better care for the child with DMD can be ensured through the welcoming the palliative care interventions. The central theme of the current study gives direction for the practitioners to intervene, to provide a positive experience for the parents of children with DMD.

The implication of the study

The current study is an addition to the existing body of literature from the Indian context. It points towards the need for creating awareness about palliative care services and initiating palliative care discussions in the initial stage of routine clinical neurological interventions. This can prepare the parents and the child with DMD with adequate information about palliative care services and prepare them emotionally to avail those services.

Limitation

Resistance from the participants to open up about the topic as it was the first-time discussion surrounding this topic posed a major challenge for the researcher. The small sample size is one of the limitations of the current study. The proximity of the families for a home visit made it difficult to recruit more cases. Gender of the respondents was not kept homogeneous. Hence to make generalizations about the difference between the experiences of father and mother could not be made. As most of the children whose parents were recruited for the current study were wheelchair-bound, due to the increased care needs, only one parent could participate in the interview. Hence the comprehensive views of the family about the palliative care for their child could have missed.

CONCLUSION

The current study points towards the need to create awareness among the public about paediatric palliative care services, especially for the neuromuscular conditions. Early intervention in the disease trajectory could ease the associated emotional issues during the transitional stages. For the effective paediatric care interventions, there should be an integration of palliative care and neuromuscular disease care at the earlier stages. Different stakeholders should be involved in ensuring optimal care for children with a life-limiting condition like DMD.

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Conflicts of interest

There are no conflicts of interest.

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