



The Importance of Sexual Health Counseling in the Care of Patients with Duchenne Muscular Dystrophy

Alyssa Rohan¹, Ellis Barrera² and Nora E Renthal^{2*}

¹Department of Neurology, Boston Children's Hospital, Boston, MA, USA

²Department of Pediatrics, Division of Endocrinology, Boston Children's Hospital and Harvard Medical School, Boston, MA, USA

Corresponding author: Nora E Renthal, Department of Pediatrics, Division of Endocrinology, Boston Children's Hospital and Harvard Medical School, Boston, MA, USA.

Received Date: August 02, 2023

Published Date: August 15, 2023

Abstract

The purpose of this review is to summarize the clinical literature available on the role of sexual health counseling in the medical and social management of patients with Duchenne Muscular Dystrophy (DMD). As treatment options slowing disease progression continue to emerge, patients with DMD are anticipated to experience longer life expectancies, and there is a growing need for increased support for patients transitioning through adolescence and into adulthood. While the physical impact of DMD is well-documented, there is a growing recognition of the psychosocial aspects associated with the condition. Adolescence and young adulthood mark a critical period for individuals with DMD as they navigate the challenges of transitioning from pediatric to adult care. Sexual health counseling provides accurate information, education, and support related to sexual development, relationships, intimacy, contraception, and reproductive choices. By incorporating sexual health counseling into the comprehensive care of individuals with DMD, healthcare providers can help empower patients, foster healthy sexual development, and improve their overall quality of life. As the medical community continues to recognize the importance of holistic care for individuals with DMD, the integration of sexual health counseling into routine care becomes essential.

Key words: Duchenne Muscular Dystrophy; Sexual health; Mental health; Counseling; Disability

Abbreviations: DMD-Duchenne Muscular Dystrophy

Introduction

Duchenne Muscular Dystrophy (DMD) is a degenerative neuromuscular disease caused by a genetic mutation in the dystrophin gene, resulting in a deficiency or the complete absence of the dystrophin protein. DMD is an X-linked disease and affects approximately 20 per 100,000 live male births [1]. Mutations in dystrophin lead to the progressive deterioration of skeletal, smooth, and cardiac muscle [2]. Historically, patients with DMD require the use of a wheelchair in their early teenage years, and then experience cardiac and pulmonary complications resulting in death in their late teens or early twenties [3]. Without appropriate medical intervention, the mean age of survival for a patient diagnosed with

DMD is 19 years of age [4], however, clinical advancements and the regular use of corticosteroids have improved the prognosis and life expectancy for patients [5]. Indeed, the median life expectancy for DMD patients who receive respiratory support is 32 years [6,7]. Due to the increased life expectancy, other aspects of living with DMD, such as emotional wellbeing and support during the transition to adulthood, have become increasingly important [8].

The World Health Organization defines sexual health as "a state of physical, emotional, mental and social well-being in relation to sexuality; it is not merely the absence of disease, dysfunction or infirmity. Sexual health requires a positive and respectful approach

to sexuality and sexual relationships, as well as the possibility of having pleasurable and safe sexual experiences, free of coercion, discrimination and violence. For sexual health to be attained and maintained, the sexual rights of all persons must be respected, protected, and fulfilled." (Sexual health (who.int)). Furthermore, sexual health is recognized as a right, and all individuals should be able to pursue sexual exploration and further their sexual health.

Sexual health education consists of information regarding the biological, sociocultural and relational angles of sexuality [9]. However, students with disabilities frequently receive little to no sexual health education that is relevant to their disability [9]. A literature review performed by Michielsen and Brockschmidt in 2021 investigating sexual education for youth with disabilities, revealed that most of the published studies related to sexual education and disabilities focus on individuals with intellectual and cognitive disabilities, with far fewer studies on sexual education for youth with physical disabilities [10]. Sexual health counseling is an essential component of delivering comprehensive care and compassion to every patient, particularly when medical interventions, like testosterone treatment for delayed puberty, could potentially influence sexual health and reproductive abilities. Individuals with disabilities face significant difficulties meeting their intimate and sexual needs, not only because of their reduced mobility, but also because of the stigma they face. Indeed, individuals with disabilities are often infantilized by their caregivers and/or viewed as being asexual [11].

In their guidance for the treatment of DMD patients, Birnkrant et al. listed dating, intimacy and sexuality as high priority topics among patients with DMD [8]. However, there are many factors that complicate addressing such issues with DMD patients in the clinical care setting. At the ages when individuals typically seek independence and autonomy from their caregivers, DMD patients rely more on their caregivers for physical support due to their functional decline. The increased physical dependence on caregivers can complicate the already awkward household discussions related to sexual wellbeing, masturbation, dating, and relationships [8]. Furthermore, in an interview study with parents of patients with disabilities conducted by East and Orchard, parents reported not feeling adequately equipped to discuss these topics with their children [12]. In their considerations and recommendations for care of patients with DMD, Birnkrant et al. suggest that medical providers should initiate discussions related to masturbation, having sex, and becoming a parent. However, research into the occurrence and potential benefits of such discussions is scarce.

Discussion

Sexual health is an essential aspect of overall well-being and quality of life, yet individuals with DMD have been largely underserved in this area. The limited published research and educational materials on sexual health for individuals with DMD have resulted in a significant knowledge gap. Consequently, individuals with DMD may not have received the necessary support and information to navigate sexual relationships, intimacy, and reproductive health.

In the past decade, there has been a shift in attitudes towards sexual health in the context of DMD. Researchers and healthcare providers have started to acknowledge that addressing sexual health is crucial to enhance the overall well-being and life satisfaction of individuals with DMD. Several papers have explored the unique sexual health needs and concerns of this population, shedding light on the challenges they face in establishing healthy relationships and understanding their sexual identities [8,13]. Despite this increased appreciation for the importance of sexual health, there remains a paucity of research exploring the implementation of sexual health guidelines in clinical practice for individuals with DMD. Few studies have specifically delved into how healthcare providers can effectively integrate discussions about sexual health into routine care for their patients with DMD. This gap in research poses a significant barrier to ensuring comprehensive and patient-centered care for this population.

Conclusion

The new landscape of genetic therapy for DMD has brought about groundbreaking advancements in the field. Approaches such as gene editing and gene therapies have shown promising results in preclinical and clinical trials, offering hope for disease-modifying treatments. As novel genetic therapies give hope for improved patient survival and longer lifespans, new questions arise concerning the implications for sexual health in individuals with DMD [14].

In this setting, sexual health remains an underrepresented and understudied aspect of care for individuals with DMD. While there has been a growing appreciation for its significance in recent years, the lack of research exploring the implementation of sexual health guidelines at the clinical level remains a significant concern. Addressing this gap is vital, especially with the emergence of genetic therapies for DMD, as it presents new opportunities to investigate the impact of these treatments on sexual health. Prioritizing research in this area will lead to a better understanding of the sexual health needs of individuals with DMD, ultimately improving their overall quality of life and well-being.

To achieve progress, healthcare providers must recognize the critical role of sexual health counseling in the medical and social management of patients with DMD. Allocating additional resources to better understand the most common and distressing sexual health challenges faced by DMD patients is imperative. By addressing the unique needs of individuals with DMD in the realm of sexual health, healthcare providers can enhance their quality of life, support their transition to adult care, and promote overall well-being. Integration of sexual health counseling into routine care is crucial to ensuring comprehensive and patient-centered management of individuals with DMD.

The need for more research is evident, particularly regarding the impact of social isolation on sexual health. Studies have shown that social isolation is a significant concern for individuals with DMD, and it directly affects their sexual well-being [15]. By emphasizing the importance of social interaction and support,

healthcare providers can point to tools and interventions that can help alleviate the negative effects of isolation on sexual health.

Moreover, as treatment for DMD improves, patients are living longer, leading to the need for more research addressing issues related to lifespan health. Given the rapid advancements in the management of DMD, the early age of onset and rapid progression of DMD, and the isolating nature of the disease with a reliance on caregivers, gender considerations become crucial in understanding sexual health challenges. Tailored research specific to DMD patients is necessary to cater to their unique circumstances effectively.

Looking ahead, future research in sexual health should eventually lead to the development of better tools and interventions to ameliorate the impact of DMD on sexual well-being. Taking advantage of the onset of better technology and social media platforms can be a way to address social isolation, connecting individuals with DMD to support networks and providing access to valuable resources and information.

In summary, addressing the underrepresentation and lack of research on sexual health in individuals with DMD is of utmost importance. Healthcare providers must prioritize research in this area and integrate sexual health counseling into routine care to comprehensively support the well-being and quality of life of DMD patients. By emphasizing the need for more research, highlighting the impact of social isolation, and considering the unique challenges faced by DMD patients in the context of gene therapies and gender considerations, we can pave the way for better tools and interventions to enhance their overall health and happiness.

Acknowledgement

None.

Conflict of Interest

The authors declare no conflict of interest.

References

- David J Birnkrant, Katharine Bushby, Carla M Bann, Susan D Apkon, Angela Blackwell et al. (2018) "Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and neuromuscular, rehabilitation, endocrine, and gastrointestinal and nutritional management". *Lancet Neurol* 17(3): 251–267.
- David J Birnkrant, Katharine Bushby, Carla M Bann, Benjamin A Alman, Susan D Apkon et al. (2018) "Diagnosis and management of Duchenne muscular dystrophy, part 2: respiratory, cardiac, bone health, and orthopaedic management". *Lancet Neurol* 17(4): 347–361.
- D Duan, N Goemans, S Takeda, E Mercuri, A Aartsma Rus (2021) "Duchenne muscular dystrophy". *Nat Rev Dis Primer* 7(1): 1–19.
- Katharine Bushby, Richard Finkel, David J Birnkrant, Laura E Case, Paula R Clemens, et al. (2010) "Diagnosis and management of Duchenne muscular dystrophy, part 1: diagnosis, and pharmacological and psychosocial management". *Lancet Neurol* 9(1): 77–93.
- Michela Guglieri, Kate Bushby, Michael P McDermott, Kimberly A Hart, Rabi Tawil et al. (2022) "Effect of Different Corticosteroid Dosing Regimens on Clinical Outcomes in Boys With Duchenne Muscular Dystrophy". *JAMA* 327(15): 1456–1468.
- E Landfeldt, R Thompson, T Sejersen, HJ McMillan, J Kirschner, et al. (2020) "Life expectancy at birth in Duchenne muscular dystrophy: a systematic review and meta-analysis". *Eur J Epidemiol* 35(7): 643–653.
- J Broomfield, M Hill, M Guglieri, M Crowther, K Abrams (2021) "Life Expectancy in Duchenne Muscular Dystrophy: Reproduced Individual Patient Data Meta-analysis". *Neurology* 97(23): e2304–e2314.
- DJ Birnkrant et al. (2018) "Diagnosis and management of Duchenne muscular dystrophy, part 3: primary care, emergency management, psychosocial care, and transitions of care across the lifespan". *Lancet Neurol* 17(5): 445–455.
- A Kriofske Mainella, S Smedema (2022) "Sexual Health Education and Life Satisfaction for People with Congenital Neurological Disabilities". *Rehabil Couns Bull*.
- K Michielsen, L Brockschmidt (2021) "Barriers to sexuality education for children and young people with disabilities in the WHO European region: a scoping review". *Sex Educ* 21(6): 674–692.
- R Addlakha, J Price, S Heidari (2017) "Disability and sexuality: claiming sexual and reproductive rights". *Reprod Health Matters* 25(50): 4–9.
- LJ East, TR Orchard (2014) "Somebody Else's Job: Experiences of Sex Education among Health Professionals, Parents and Adolescents with Physical Disabilities in Southwestern Ontario". *Sex Disabil* 32(3): 335–350.
- K Areskoug Josefsson (2013) "Muscular dystrophy and sexual health". *OA Musculoskelet Med* 1(2).
- RJ Fairclough, MJ Wood, KE Davies (2013) "Therapy for Duchenne muscular dystrophy: renewed optimism from genetic approaches". *Nat Rev Genet* 14(6): 373–378.
- PS Dreyer, BF Steffensen, BD Pedersen (2010) "Living with severe physical impairment, Duchenne's muscular dystrophy and home mechanical ventilation". *Int J Qual Stud Health Well-Being* 5(3).