

Muscular dystrophy and sexual health

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Abstract

Introduction

Muscular dystrophy is a group of muscular disorders, which are hereditary with progressive muscular weakness, giving symptoms such as pain, fatigue, physical and psychological impairment. The definition of good sexual health, the individual's right to good sexual health and the relation between sexual health and general health lead to the question of how good sexual health is enabled by health professionals for persons with severe chronic disease, such as muscular dystrophy.

The aim of this review was to explore the existing literature concerning muscular dystrophy and sexual health focusing on patient information and support by health care professionals.

Materials and methods

A literature search of PubMed and PsycInfo was done for 'muscular dystrophy and sexual', 'muscular dystrophy and sexuality'. The articles that were written in English and concerned humans were included. The literature review included both quantitative and qualitative studies and covered journals, research papers, conference proceedings and theses.

Results

Only five articles were found to suit the aim of this study. Through a search of reference lists from the found articles, another three articles were found. Among those articles, only three had high relevance; the other five were too old. The three

articles that had high relevance had different scopes and methodology. One study compared levels of hypogonadism and erectile dysfunction (case-control study), another studied adult life with muscular dystrophy (cross-sectional study) and the third explored reproductive outcome and family planning after genetic counselling (combining interviews and questionnaires).

Discussion

The existing literature is scarce and there is a need for further research concerning sexual health and muscular dystrophy in order to explore how persons with muscular dystrophy experience their sexual health.

Conclusion

It is necessary to develop clinical advice for how health professionals working with persons who have muscular dystrophy can support the patient's sexual health and give relevant information to patients/partners/relatives. Despite the lack of knowledge concerning sexual health and muscular dystrophy, models for communication and openness about sexual health among health professionals could probably improve care for this group of patients.

Introduction

Sexual health is defined by the World Health Organization as: 'a state of physical, mental and social well-being in relation to sexuality. It 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'¹. According to this definition of sexual health, it should be seen in a broad perspective where the individual's right to good sexual health is an important part. Sexual

wellbeing is an integrated part of sexual health, and sexual wellbeing is related to physical and psychological health, as well as level of sexual activity². The definition of good sexual health, the individual's right to good sexual health and the relation between sexual health and general health lead to the question of how good sexual health is enabled by health professionals, for persons with severe chronic disease, such as muscular dystrophy.

Muscular dystrophy is a group of muscular disorders, which are hereditary with progressive muscular weakness. The prevalence of muscular dystrophy varies in different parts of the world but, as an example, the incidence is one in 3500 newborn males in Western Europe³. Some of the muscular dystrophies are congenital, and they often have a worse prognosis than other muscular dystrophies that develop in adults³. Despite research efforts there is no cure for muscular dystrophy⁴. Characteristic symptoms of muscular dystrophy are loss of muscle mass with subsequent muscular weakness, not due to other disease or trauma. Muscular dystrophy leads to the breakdown of muscular cells and replacement with connective tissue or fat³. If the muscle cells are replaced by fat, the shape of the muscles can have normal proportions even if the muscle capacity is severely reduced. The disease can also include endocrine disorders, cataracts and intellectual impairment. It often gives symptoms such as severe pain, fatigue, and physical and psychological disability⁴, which affect patients' whole life situation, and quality of life moreover decreases with disease duration⁵. The most common muscular dystrophies are myotonic

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dystrophy, fascioscapulohumeral muscular dystrophy, Becker's muscular dystrophy and Duchenne muscular dystrophy³. Becker's muscular dystrophy affects boys aged 5–15 years. Duchenne muscular dystrophy also affects males, but has an earlier start, often at the age of 2–3 years. Duchenne muscular dystrophy has a worse progress, despite the progress in management of the disease, the patients rarely become older than 25 years of age⁶. It is a need to change the paradigm from thinking of Duchenne muscular dystrophy as a disease in childhood and acknowledge the adulthood of patients with Duchenne muscular dystrophy and in order to do so issues of sexual health needs to be addressed⁷. As muscular dystrophy can develop at different stages of life, it might affect sexual health in different ways. Sexual health can be affected negatively in a biological, psychological and social perspective by muscular dystrophy, for example, by a changed body image, impotence, chronic pain, fatigue, risk for depression, fear of not finding a partner and worries about not being sexually desirable^{8–13}. Those with childhood onset of muscular dystrophy are at a higher risk to be single as adults than those developing the disease as adults¹⁴. However, there is limited research on how muscular dystrophy affects sexual health. Health care professionals are often reluctant to incorporate issues concerning sexual health in the care of chronically ill patients, due to reasons such as embarrassment, lack of skills and knowledge, fear of offending the patient and beliefs that the patients are too ill⁹. Persons with disabilities often experience dissatisfaction with the support from health care professionals concerning sexual health^{9,15}. It is reasonable to believe that this is true concerning health professionals working with persons with muscular dystrophy as well. Despite this, adolescents with

physical disabilities often depend on professionals for sex education¹⁶.

The aim of this review was to explore the existing literature concerning muscular dystrophy and sexual health focusing on patient information and support by health care professionals.

Materials and methods

A literature search of PubMed and PsycInfo was done for 'muscular dystrophy and sexual', 'muscular dystrophy and sexuality'. The articles that were written in English and concerned humans were included. The literature review included both quantitative and qualitative studies and covered journals, research papers, conference proceedings and theses. The bibliographies of relevant publications were checked to identify additional studies. The search was carried out in June 2013.

The identified studies were assessed using the models for critical assessment of studies recommended by the Swedish Council on Technology Assessment in Health Care. The author made an initial screening to rate the relevance of the articles included. Potentially relevant articles were reviewed independently by the author and thereafter assessed for quality and relevance. To achieve high relevance, the study should be published in the last 15 years and relate to patients with muscular dystrophy and their life, including sexual health.

Results

The literature search of PubMed and Psycinfo for 'muscular dystrophy and sexual/sexuality/intimate relationships' gave eight articles where the abstracts seemed to be covering the aim of this study (20130615). After reading the full text, only five articles were found to suit the aim of this study. Searches for 'Myotonic dystrophy and sexual/sexuality',

muscular dystrophy, 'Duchenne muscular dystrophy and sexual/sexuality', 'Fascioscapulohumeral muscular dystrophy and sexual/sexuality' and 'Becker's muscular dystrophy and sexual/sexuality' gave no further articles. The found articles were from 1977 to 2011 and were limited to refereed journal articles in English. Through a search of reference lists from the found articles, another three articles were found. Among those articles, only three had high relevance; the other five were too old. Three of the five older articles were also limited in relevance since they were not specific to muscular dystrophy but described population studies that had included one or more patients with muscular dystrophy. Both of the remaining older articles focused on male reproductive sexual health. The three articles that had high relevance had different scopes and methodology. One study compared levels of hypogonadism and erectile dysfunction (case-control study¹⁷), another studied adult life with muscular dystrophy (cross-sectional study¹⁸) and the third explored reproductive outcome and family planning after genetic counselling (combining interviews and questionnaires¹⁹). Due to the small amount of studies included and the diverse methods used, it was not relevant to perform a uniform quality test.

This is a very small number of research articles for a disease that develops among infants, children and adults, men as well as women. Due to the small amount of research material, an additional search of the internet was made using Google to find networks and associations for persons with muscular dystrophy. This search gave online sites for patients and relatives, which were mainly addressed to relatives, with very limited information about sexual health for persons with muscular dystrophy.

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Discussion

There is a very limited research on sexual health in persons with muscular dystrophy, both concerning symptoms that might decrease sexual health as well as solutions for improving sexual health. One reason might be the shortened lifespan in some of the muscular dystrophies. However, the lifespan has increased for boys with Duchenne muscular dystrophy, and the quality of life of persons with Duchenne muscular dystrophy is good in all areas except sexual health¹⁸. In this study the persons with Duchenne muscular dystrophy were described as being 'in desperate need of a love life'. The conclusion of the study was that it is important to give persons with Duchenne muscular dystrophy the possibility to develop from boy to man with competence in all areas of life, including sexuality.

As early as in 1989, another study acknowledged the importance of including intimate/sexual relationships in the care of persons with Duchenne muscular dystrophy²⁰ and, in a study concerning reproductive

sexual health, both emotional and sexual function are brought up as factors influencing family planning decisions, as well as considerations about the heredity of muscular dystrophy¹⁹. The recommendations in earlier research were counselling and support groups in order to improve sexual relations for this group of patients²⁰. For the other major muscular dystrophy diseases, there was no published research concerning sexuality or sexual health, showing a need for future research in the field. There is, today, no description of how common sexual health problems are in persons with muscular dystrophy or what problems they experience as major problems. Despite the lack of descriptions of the way in which sexual health might be affected by different muscular dystrophies, there are symptoms of the diseases that might affect sexual health (Table 1) and the impact on sexual health is probably diverse, including physical, psychological and social issues.

The age at onset of the disease is also important, since persons developing muscular dystrophy as adults

might have developed their sexual identity and their sexual problems might therefore have a very different character from those people whose disease emerges when they are adolescents, children or infants. When becoming ill as an adult, there might be a need to change a sexual role, from active to more passive, in order to perform sexual activities. For persons who develop the disease early in life, the issue is more about developing a sexual identity and finding a way to achieve a satisfying love life as adults. Here the support of relatives is important, especially since many persons with muscular dystrophy often need a great deal of assistance by their relatives in their daily living. A majority of adolescents report dissatisfaction with their body image, and it is likely that adolescents with physical disabilities are in this group²¹. The support of relatives is essential for children and youths in order to develop a good sexual health, since parents play an important role in a person's development towards independence in all areas of life, including sexuality^{22,23}. Areas of

Table 1 Symptoms of muscular dystrophy with possible negative effects on sexual health

Type of muscular dystrophy	Symptom	Possible negative effects on sexual health
Myotonic dystrophy	Difficulties closing the mouth	Difficulties in kissing and oral sex
	Delayed muscular relaxation, feeling of muscular stiffness	Difficulties with bodily positions during sexual activities
	Testicular atrophy	Negative body image
	Pregnancy complications	Affect sexual and reproductive functions
	Unclear speech	Difficulties communicating with potential partners
Fascioscapulohumeral muscular dystrophy	Mental symptoms, such as apathy and inactivity	Difficulties finding a partner and performing sexual activities
	Difficulties with mouth movements	Difficulties in kissing, performing oral sex
	Difficulties lifting the arms	Difficulties with hugging, caressing
Becker's muscular dystrophy and Duchenne muscular dystrophy	Hearing impairment	Difficulties communicating with partner/potential partner
	Decreased lower limb muscles	Functional disability, negative body image

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importance are, for example, that the family acknowledges sex as something positive, supports sexual feelings, gives possibilities for healthy sexual play with peers (flirting) and confirms that sexuality is not associated with guilt and shame.

It is necessary to see sexual health from a gender perspective, since there are differences in how sexual health problems among adults with muscular dystrophy are experienced; women experience the physical weakness as the greatest hindrance in their sexual life and erectile dysfunction is the most common difficulty for men^{17,24}. Neither has received much attention in clinical practice or research. There are also differences in perceived sexual roles that might increase the burden of muscular dystrophy, such as that 'men should be muscular and strong'.

There is research on physical and psychological disabilities in general being associated to sexual health, but this is unfortunately, probably, not sufficient for persons with muscular dystrophy due to their complex range of symptoms, including physical and psychological disability, pain and fatigue. However, specific therapy suggestions concerning single symptoms might be useful for the individual patient, for example exercise therapy to improve mobility and thereby indirectly improve sexual health or interventions directly improving sexual health such as medical treatment of erectile dysfunction. Health professionals often hesitate to communicate about sexual health with their patients, but it is still essential that they have good skills in this field^{9,25}. Knowledge and openness are the basis for good communication between health professional and patient about sexual health issues, even if health professionals can experience barriers in approaching the subject. Common barriers are the sensitivity of the subject; the health professionals being unsure as to how

to bring up the subject, being unsure of how they can support persons with muscular dystrophy who experience sexual health problems and believing that someone else in the health care team is responsible for discussing sexual health with patients²⁶⁻²⁹. The patients themselves might be reluctant to initiate communication about sexual health problems, especially if they believe that the health professionals are unavailable to offer any support or if they believe that they shouldn't be sexually active due to their disability²⁹⁻³¹. If the patient has cognitive impairment, this might increase the barrier towards communicating about sexual health and a way to overcome this barrier is to see the individual behind the disability and to acknowledge the individual's right to a sexual life^{32,33}. However, there is a greater acceptance of discussing sexual health among health professionals than among family care givers, which can limit communication for patients where family care givers are present in the clinical meeting³⁴. The heredity of muscular dystrophies might increase the difficulties of encouraging and discussing sexual health with this group of patients. If sexual activity is only seen from a reproductive perspective, heredity might be a difficult issue to approach, both for the patient and health professionals. However, sexual health deserves a broader perspective including desire, joy and satisfaction. It is therefore important to explore how care givers and relatives give support and how their attitudes are towards supporting sexual health in persons with muscular dystrophy. Even if the perspective is to improve sexual health in order to ensure sexual pleasure and satisfaction, there is probably a lack of support in this field as well. There has been a change of paradigm in sexual therapy, from a focus on sexual function to a focus on sexual satisfaction, which should be acknowledged by health

professionals². Unfortunately, this change seems to be delayed for persons with disabilities, where the main focus still is on reproduction and sexual function³⁵. The individual's choice might be a problem of how support concerning sexual health is ensured for persons in need of advice. For example, it should be the patient's choice how they wish to use their reduced muscular capacity, but it is doubtful whether patients gain understanding from health care professionals if they choose to use their energy on sexual activities instead of other activities of daily living. It should be acceptable to choose to get assistance with dressing in order to save energy for sexual life. The same choice should be given concerning medication; if there are side effects that might affect the sexual health of the patients, they should be informed in order to be able to make their own choice of what is most important to them as individuals. It would be of interest to further study how health professionals address this issue. A key word in improving sexual health and making an informed decision is information. However, considering the scarce research publications, information is probably lacking for this group of patients. It should be natural that an individual is allowed to make his or her own decisions in this field, instead of somebody else making the decision from the perspective of a health care professional or a relative. Even with the limited research available concerning sexual health in persons with muscular dystrophy, the existing information should be presented to the patients and their eventual partners. An example of information could be explaining the relation between pain and sexual function and/or sexual desire.

To improve sexual health in persons with muscular dystrophy, research concerning other chronic diseases and sexual health might

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be useful in clinical practice, since research concerning muscular dystrophy and sexual health is scarce. Decreasing symptoms of chronic disease, such as pain and fatigue, and increasing functional ability have been described to improve sexual health in persons with other chronic muscular diseases. Partner communication, cognitive therapy and physiotherapy may also be beneficial^{36,37}. Treatment of erectile dysfunction can improve quality of life and self-esteem and can be treated with medication or cognitive behavioural therapy³⁸.

A basis for improving sexual health for persons with muscular dystrophy could be to use the Recognition Model²⁶. The basis of this model is to acknowledge the person with a disability as a sexual person with sexual needs and the right to good sexual health. This is assured by all health care professionals taking their part in improving sexual health for the patient, based on their professional knowledge. As an example, the physiotherapist can be supportive in reducing pain and increasing physical function and the occupational therapist can assist in planning daily activities.

Another useful model is the PLISSIT (permission, limited information, specific suggestions, intensive therapy), a step-wise model for communication concerning sexual health³⁹. The PLISSIT can be used by different professionals to determine the appropriate level of intervention⁴⁰.

Conclusion

There is a need for further research concerning sexual health and muscular dystrophy in order to:

- Explore how persons with muscular dystrophy experience their sexual health and the importance of sexual health in their lives, in order to improve care for this group of patients.

- Explore the type of support needed from health care professionals in order to improve sexual health for persons with muscular dystrophy.

Relevance to clinical practice

It is necessary to develop clinical advice for how health professionals working with persons with muscular dystrophy can support the patient's sexual health and give relevant information to patients/partners/relatives. Despite the lack of knowledge concerning sexual health and muscular dystrophy, models for communication and openness about sexual health among health professionals could probably be useful and improve care for this group of patients.

Authors' contribution

KAJ was responsible for the study design, data collection and analysis, and the manuscript preparation.

References

1. World Health Organization. Defining sexual health. Report of a technical consultation on sexual health. In: Sexual health document series. Geneva: World Health Organisation; 2006.p35.
2. Meana M, Jones S. Developments and trends in sex therapy. *Adv Psychosom Med.* 2011 Jan;31:57–71.
3. Emery AE. The muscular dystrophies. *Lancet.* 2002 Feb;359(9307):687–95.
4. Harper PS, Van Engelen B, Eymard B, Wilcox DE, editors. *Myotonic dystrophy, present management, future therapy.* Oxford: Oxford University Press; 2004.
5. Natterlund B, Gunnarsson LG, Ahlstrom G. Disability, coping and quality of life in individuals with muscular dystrophy: a prospective study over five years. *Disabil Rehabil.* 2000 Nov;22(17):776–85.
6. Eagle M, Baudouin SV, Chandler C, Giddings DR, Bullock R, Bushby K. Survival in Duchenne muscular dystrophy: improvements in life expectancy since 1967 and the impact of home nocturnal ventilation. *Neuromuscul Disord.* 2002 Dec;12(10):926–9.
7. Schrans DG, Abbott D, Peay HL, Pangalila RF, Vroom E, Goemans N, et al.

Transition in Duchenne muscular dystrophy: an expert meeting report and description of transition needs in an emergent patient population: (Parent Project Muscular Dystrophy Transition Expert Meeting 17-18 June 2011, Amsterdam, The Netherlands). *Neuromuscul Disord.* 2013 Mar;23(3):283–6.

8. Monga TN, Tan G, Ostermann HJ, Monga U, Grabis M. Sexuality and sexual adjustment of patients with chronic pain. *Disabil Rehabil.* 1998 Sep;20(9):317–29.

9. Higgins A, Sharek D, Nolan M, Sheerin B, Flanagan P, Slaicuinait S, et al. Mixed methods evaluation of an interdisciplinary sexuality education programme for staff working with people who have an acquired physical disability. *J Adv Nurs.* 2012 Nov;68(11):2559–69.

10. Reynaert C, Zdanowicz N, Janne P, Jacques D. Depression and sexuality. *Psychiatr Danub.* 2010 Nov;22 Suppl 1: S111–3.

11. Goins LB, Markey CN, Gillen MM. Understanding men's body image in the context of their romantic relationships. *Am J Mens Health.* 2012 May;6(3):240–8.

12. Mastrogiacomo I, Bonanni G, Menegazzo E, Santarossa C, Pagani E, Gennarelli M, et al. Clinical and hormonal aspects of male hypogonadism in myotonic dystrophy. *Ital J Neurol Sci.* 1996 Feb;17(1):59–65.

13. McCabe MP, Taleporos G. Sexual esteem, sexual satisfaction, and sexual behavior among people with physical disability. *Arch Sex Behav.* 2003 Aug;32(4):359–69.

14. Bostrom K, Ahlstrom G. Quality of life in patients with muscular dystrophy and their next of kin. *Int J Rehabil Res.* 2005 Jun;28(2):103–9.

15. Milligan M, Neufeld A. The myth of asexuality: a survey of social empirical evidence. *Sex Disabil.* 2001 Jun;19(2):91–109.

16. Berman H, Harris D, Enright R, Gilpin M, Cathers T, Bukovy G. Sexuality and the adolescent with a physical disability: understandings and misunderstandings. *Issues Compr Pediatr Nurs.* 1999 Oct–Dec;22(4):183–96.

17. Antonini G, Clemenzi A, Bucci E, De Marco E, Morino S, Di Pasquale A, et al. Hypogonadism in DM1 and its relationship to erectile dysfunction. *J Neurol.* 2011 Jul;258(7):1247–53.

18. Rahbek J, Werge B, Madsen A, Marquardt J, Steffensen BF, Jeppesen J.

- Adult life with Duchenne muscular dystrophy: observations among an emerging and unforeseen patient population. *Pediatr Rehabil.* 2005 Jan-Mar;8(1):17-28.
19. Eggers S, Zatz M. How the magnitude of clinical severity and recurrence risk affects reproductive decisions in adult males with different forms of progressive muscular dystrophy. *J Med Genet.* 1998 Mar;35(3):189-95.
20. Edwards RH. Management of muscular dystrophy in adults. *Br Med Bull.* 1989 Jul;45(3):802-18.
21. Markey CN. Invited commentary: why body image is important to adolescent development. *J Youth Adolesc.* 2010 Dec;39(12):1387-91.
22. Antle BJ, Mills W, Steele C, Kalnins I, Rossen B. An exploratory study of parents' approaches to health promotion in families of adolescents with physical disabilities. *Child Care Health Dev.* 2008 Mar;34(2):185-93.
23. Nikolaisen J, Dammen Nikolaisen K. Sexuality in childhood. In: Traeen B, Lewin B, editors. *Sexology in context* ed. Oslo Univeristetsforlaget; 2008.
24. Westgren N. Attmötakvinnor med handikappisexualogiskochgynekologisk rådgivning. In: Riksförbundet för Sexuell Upplysning. 1998.
25. Kedde H, Van De Wiel HB, Schultz WC, Vanwesenbeek WM, Bender JL. Efficacy of sexual healthcare for people with chronic diseases and physical disabilities. *J Sex Marital Ther.* 2010;36(3):282-94.
26. Couldrick L, Gaynor S, Cross V. Proposing a new sexual health model of practice for disability teams: the Recognition Model. *Int J Ther Rehabil.* 2010 Jun;17(6):290-8.
27. Stausmire JM. Sexuality at the end of life. *Am J Hosp Palliat Care.* 2004 Jan-Feb;21(1):33-9.
28. Ryan S, Wylie E. An exploratory survey of the practice of rheumatology nurses addressing the sexuality of patients with rheumatoid arthritis. *Musculoskeletal Care.* 2005 Mar;3(1):44-53.
29. Bitzer J, Platano G, Tschudin S, Alder J. Sexual counseling for women in the context of physical diseases: a teaching model for physicians. *J Sex Med.* 2007 Jan;4(1):29-37.
30. McInnes RA. Chronic illness and sexuality. *Med J Aust.* 2003 Sep;179(5):263-6.
31. Eastgate G. Sex and intellectual disability--dealing with sexual health issues. *Aust Fam Physician.* 2011 Apr;40(4):188-91.
32. Löfgren-Mårtenson L. May I? About sexuality and love in the new generation with intellectual disabilities. *Sex Disabil.* 2004 Sep;22(3):197-207.
33. McGuire BE, Bayley AA. Relationships, sexuality and decision-making capacity in people with an intellectual disability. *Curr Opin Psychiatry.* 2011 Sep;24(5):398-402.
34. Evans DS, McGuire BE, Healy E, Carley SN. Sexuality and personal relationships for people with an intellectual disability. Part II: staff and family carer perspectives. *J Intellect Disabil Res.* 2009 Nov;53(11):913-21.
35. Esmail S, Darry K, Walter A, Knupp H. Attitudes and perceptions towards disability and sexuality. *Disabil Rehabil.* 2010;32(14):1148-55.
36. Josefsson KA, Gard G. Sexual health in patients with rheumatoid arthritis: experiences, needs and communication with health care professionals. *Musculoskeletal Care.* 2012 Jun;10(2):76-89.
37. Breton A, Miller CM, Fisher K. Enhancing the sexual function of women living with chronic pain: a cognitive-behavioural treatment group. *Pain Res Manag.* 2008 May-Jun;13(3):219-24.
38. Heidelbaugh JJ. Management of erectile dysfunction. *Am Fam Physician.* 2010 Feb;81(3):305-12.
39. Taylor B, Davis S. Using the extended PLISSIT model to address sexual health-care needs. *Nurs Stand.* 2006 Nov;21(11):35-40.
40. Lamont J. Female sexual health consensus clinical guidelines. *J Obstet Gynaecol Can.* 2012 Aug;34(8):769-75.